

CANCER SEMINAR

FALL, 1967, Vol. IV, No. 1

PENROSE CANCER HOSPITAL «» COLORADO SPRINGS, COLORADO
Sisters of Charity

CANCER SEMINAR

VOLUME FOUR

AUGUST, 1968

NUMBER ONE

JUAN A. DEL REGATO, M.D., *Editor*

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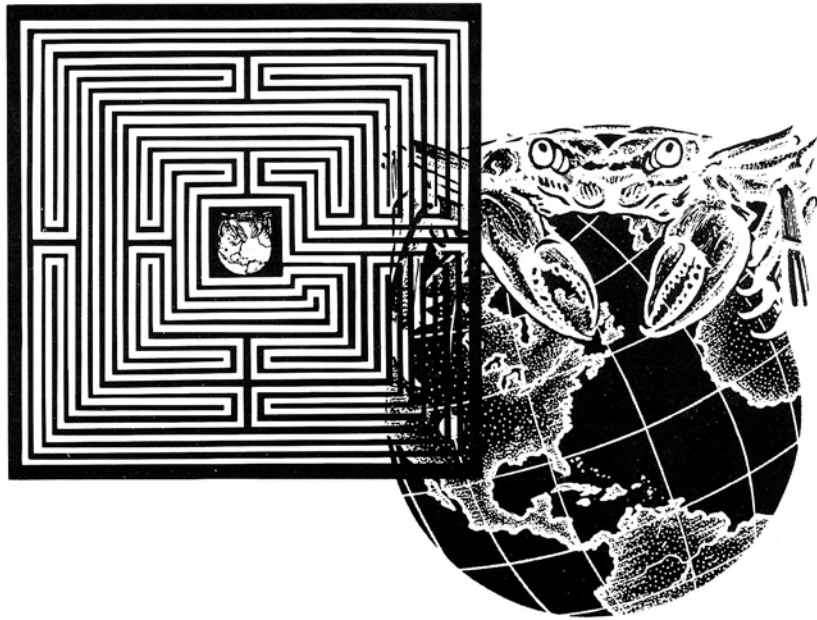
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2215 North Cascade Avenue, Colorado Springs, Colorado 80907

The CANCER SEMINAR is published annually by Graphic Services and edited by the Penrose Cancer Hospital of Colorado Springs, Colorado. J. A. del Regato, M.D., editor. Subscription rate: \$3.00 annually in the United States, Canada and Mexico. Address all correspondence to the editorial office.

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BONE TUMORS

THIS WAS OUR Nineteenth Annual **CANCER SEMINAR** and the third devoted to bone tumors; the previous seminars on this subject were held in 1950 and in 1958. No other subject is as capable of commanding the attention of radiologists, pathologists and surgeons with equal intensity.

On this occasion it was our privilege to present to our select audience two eminent representatives of the younger generation: Dr. Jack Edeiken, Professor of Radiology, Jefferson Medical College, and Dr. Harlan J. Spjut, Professor of Pathology, Baylor University College of Medicine. They are newly acknowledged experts on bone tumors in the United States; their acuity and competence will be evident to the readers of these proceedings. Dr. C. Howard Hatcher, Professor Emeritus of Orthopedic Surgery, Stanford University School of Medicine, contributed to the

discussion from his rich baggage of experience in this field.

This **CANCER SEMINAR**, held on Saturday, October 28th, 1967, produced a record attendance of 560 radiologists, pathologists and surgeons and a variety of other interested practitioners of medicine. The edited proceedings have been enriched by the excellent quality of the photomicrographs, and the careful chosen captions, for which we are all indebted to Dr. Spjut.

As always, we wish to express our sincere thanks to all those who in one way or another have contributed to these **CANCER SEMINARS** and to their success.

J. A. del REGATO, M. D.
Colorado Springs, Colorado
August, 1968

1. Synovial Sarcoma of the Knee

Contributed by **J. F. Kuzma, M.D. and R. Kascht, M.D.**, Milwaukee, Wisconsin

THE PATIENT was a 65-year old man in May, 1966, when he complained of pain above the left knee. Examination revealed a tumefaction overlying the epiphysis of the left tibia.

Dr. Edeiken: The roentgenograms reveal a large eccentric osteolytic lesion in the lateral aspect of the upper end of the left tibia. The lesion extends to the joint surface, the overlying cortical bone is thin and appears perforated in several areas. The tumor is well delineated. There is a soft tissue mass extending toward the fibula and at the proximal end of the fibula there is an overlying single calcification.

Metastatic disease and myeloma must always be considered first in this age group. It would be unusual to find soft tissue mass from either of these two lesions. The presence of calcification overlying the fibula in the soft tissue mass excludes the diagnosis of metastatic disease or multiple myeloma.

Tumors which contain giant cells often occur in the end of a long bone, are eccentric and tend to extend to the joint surface. They are often mistaken for true giant cell tumors when they may, in fact, be fibrosarcoma, chondroblastoma, chondromyxoid fibroma, chondrosarcoma, non-osteogenic fibroma, the Brown tumor of hyperparathyroidism, pigmented villonodular synovitis, aneurysmal bone cyst, and synovial sarcoma. Most of these lesions can be excluded on the basis of age. The presence of the small calcification should exclude pigmented villonodular synovitis, giant cell tumor, fibrosarcoma, and aneurysmal bone cyst. Because of the calcification, one must consider seriously chondrosarcoma or other cartilaginous tumors. In the first two primary bone tumors mentioned, one would expect more calcification within the osteolytic area of the bone. Synovial sarcomas

frequently calcify. At times the soft tissue element invades the bone and the lesion may appear as a primary bone tumor, such as this one does.

Dr. Edeiken's impression: 1) SYNOVIAL SARCOMA.

Roentgenologic Impressions Submitted by Mail

Giant-cell tumor	52
Cartilaginous tumor	17
Aneurysmal bone cyst	8
Others	19

Dr. Edeiken: A true giant cell tumor will never contain calcification; if you use that as a general rule you will be safe. This could well be a cartilaginous tumor but in that case I would expect calcification throughout; an aneurysmal bone cyst should not be accompanied by calcification.

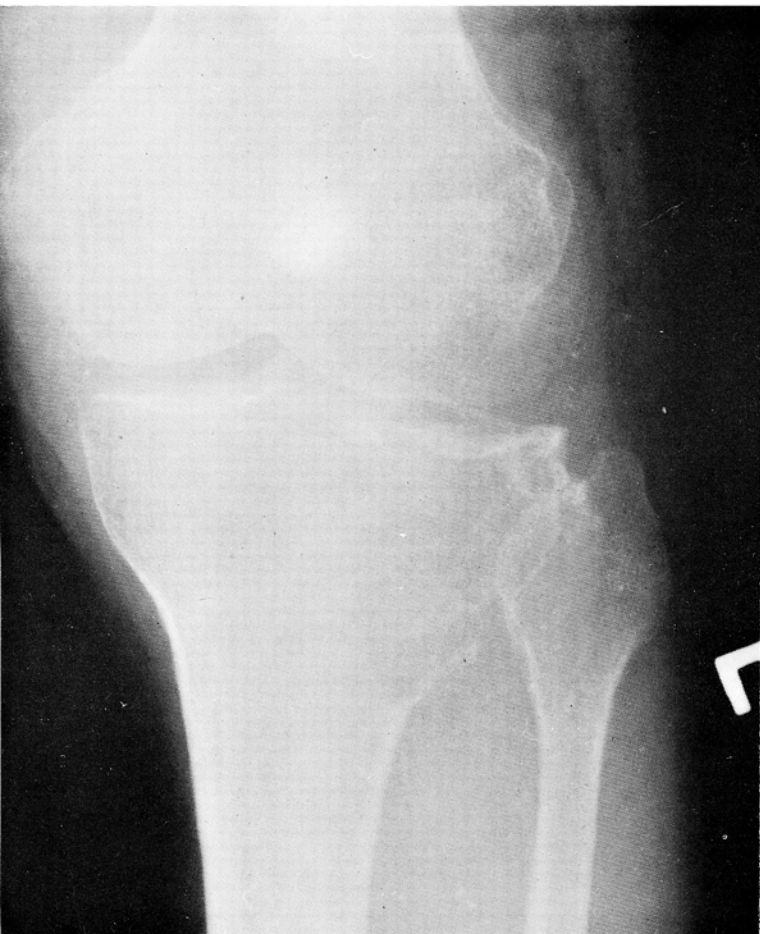
Dr. Regato: Dr. J. W. Travis, of Topeka, offered an impression of giant-cell tumor. Dr. B. L. Pear, of Denver, preferred reticulum-cell sarcoma and Dr. A. Ritch, of Roswell, New Mexico, fibrosarcoma.

Operative findings: In May, 1966, an amputation was done 17.5 cm above the knee. A non-encapsulated tumor was found on section of the specimen with lobulated margins, anteriorly situated to the upper end of the fibula.

Dr. Spjut: At first glance, the histological sections of this case might suggest the possibility of metastatic carcinoma to the tibia. Well-formed glands that have a resemblance to an adenocarcinoma of the large bowel or some other well-differentiated adenocarcinoma might be considered. However, the intervening stroma suggests that some other lesion should be considered. The stroma is extremely cellular and seems to be an intimate part of the lesion. This seems to be more than a stroma that has been provoked by a metastatic carcinoma.

Fig. 1—Osteolytic lesion of the lateral aspect of the upper end of the tibia.

Fig. 2—Section of the surgical specimen showing non-encapsulated, anteriorly situated tumor.



Another lesion that would come to mind is the adamantinoma of the tibia. This lesion could present with the radiographic findings that have been demonstrated. Histologically, this lesion usually presents areas that resemble ameloblastoma of the mandible, that is, an epithelial component resembling squamous epithelium. We don't have the squamous epithelial component in this lesion. A vascular origin has been suggested for the adamantinoma, but the spaces in this lesion do not appear to be vascular. Another lesion that may be considered in the differential diagnosis is villonodular synovitis. This lesion may cause destruction of bone. However, in this case, we do not see a villous pattern nor do we have the accumulation of foamy macrophages, hemosiderin and multinucleated giant cells so commonly seen in this disease.

We see well-formed gland structures in which there are some papillary forms; the cells are columnar to cuboidal. Very few mitoses are found and most appear normal; calcification is not seen but if present may be helpful in the diagnosis of synovial sarcoma. The combination of glandular structures plus the very cellular stroma suggest that this is a well-differentiated synovial sarcoma. Special stains in the case of synovial sarcoma generally are not of any specific help although often the mucin and PAS stains are positive. Both the mucicarmine and PAS stains were positive in this case. Positive material is demonstrable intracellularly in the gland lumens. In a recently reported large series of synovial sarcoma, the lesions were divided into three histological forms: (1) two distinct cellular components, the pseudoepithelial and spindle cells are seen throughout (2) the spindle cells predominate (3) pseudoepithelial component is dominant. The latter two groups are the commonest types of synovial sarcomas. The well-differentiated form illustrated by this case is the least common. There seems to be no prognostic significance attached to the differentiation. Formerly, it was considered that the prognosis of synovial sarcoma was extremely poor. But in two recent reviews a survival rate of 51% and 25% for 5 years has been reported. Patients still continue to die of the tumor 5 years and 10 years after treatment.

Dr. Spjut's diagnosis: SYNOVIAL SARCOMA.

Histopathologic diagnoses submitted by Mail

Adamantinoma	61
Mestastatic adenocarcinoma	45
from thyroid.....15	
Synovial sarcoma	36
Others	3

Dr. Spjut: I don't believe that we have the histological features to support the diagnosis of adamantinoma; pathologists who submitted this diagnosis probably thought that these gland-like spaces were actually vascular and went along with the view that these are tumors of vascular origin. A diagnosis of metastatic adenocarcinoma is one that has to be considered strongly because of these gland-like structures; but then, doing that is to ignore the stroma or to explain the stroma as provoked by a carcinoma.

Dr. Regato: Dr. F. Schajowicz, of Buenos Aires, submitted a diagnosis of synovial sarcoma on account of the fibroblastic stroma; Dr. C. P. Schwinn, of Los Angeles, also submitted synovial sarcoma. Dr. M. McGavran, of Saint Louis, and Dr. R. Schultz, of Sioux Falls, preferred adamantinoma. Dr. W. R. Platt, of Saint Louis, and Dr. H. L. McGaffey, of Idaho Falls, offered metastatic adenocarcinoma. Dr. R. Marcial-Rojas, of San Juan, thought it to be a metastasis from an adenocarcinoma of the thyroid.

Subsequent history: In September, 1966, extensive pulmonary metastases were found; the patient was given several chemotherapeutic drugs and, after some initial response, the tumor continued to develop slowly. He was reported symptomless but gradually failing in September, 1967.

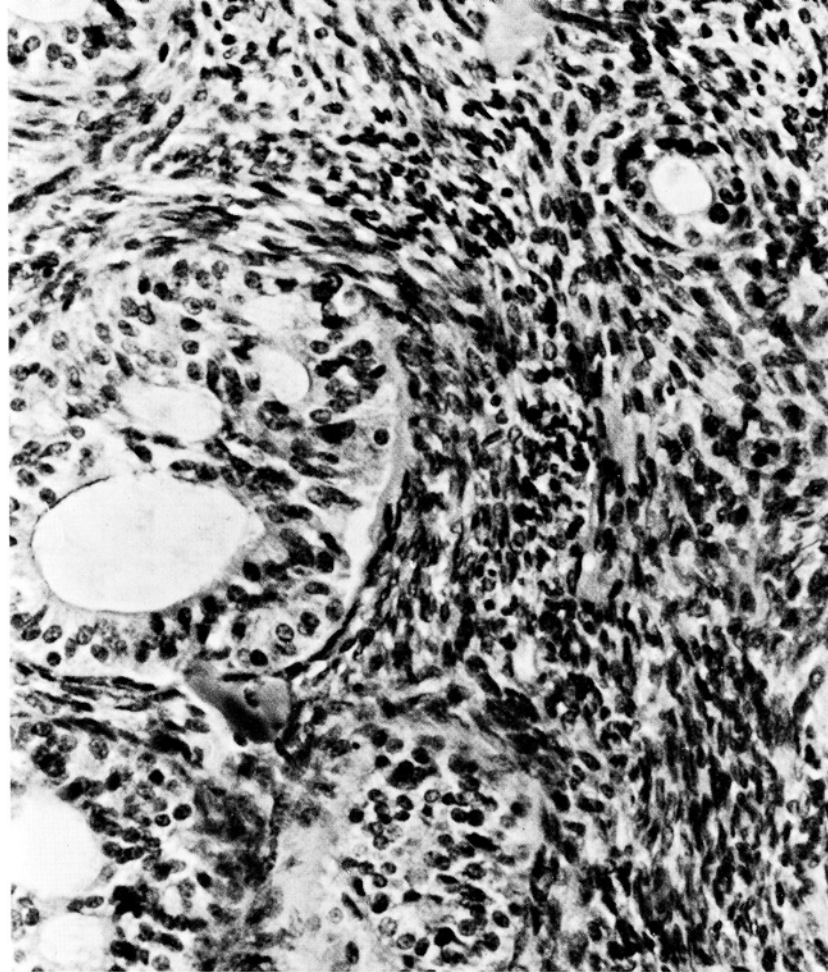


Fig. 3—Illustrated is the combination of a cellular stroma and glandular structures. Both are well differentiated and characteristic of a synovial sarcoma. Hematoxylin-eosin 300X.

Dr. Hatcher: I missed this one completely. I thought almost surely that this would be a metastatic carcinoma. I must admit that the thought of synovial sarcoma didn't cross my mind. A lateral roentgenogram might have shown a posterior mass which would suggest synovial sarcoma in that region, invading bone from the outside. The treatment of synovial sarcoma is one that is primarily surgical: complete removal of the tumor-bearing area and the regional lymph nodes above the lesion. The survivals, in my experience, have been very few indeed.

J. Kuzma, M.D., Milwaukee, Wisconsin: The patient continues with his disease in a sort of status quo. Dr. Spjut, would you comment on interosseous development of synovial sarcoma. Is there such?

Dr. Spjut: I don't know of interosseous development of synovial sarcoma; I haven't ever heard of or seen one. Synovial sarcomas will develop in rather unusual places; we have seen a couple of cases that were actually intra-abdominal; presumably related to the vertebrae, but this was never proved.

Dr. Regato: One interesting point about synovial sarcomas is that we have equally authoritative and controversial opinions about their radiosensitivity described in the medical literature.

F. Buschke, M.D., San Francisco, California: I don't like the term "radioresistant" but, in my experience, radiation therapy of synoviomas has not accomplished very much.

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2. Fibrosarcoma of the Head of the Femur

Contributed by **W. J. Frable, M.D.**, Richmond, Virginia

THE PATIENT was a 36-year old woman in October, 1966, when she complained of pain in the right hip which had been increasing for three years. In 1962, a lesion of the femoral neck, causing pain, had been curetted but the pain recurred a year later. Examination failed to reveal any physical abnormality except for limitation of motion of the right hip.

Dr. Edeiken: The films reveal marked deossification in the head and neck of the right femur with beginning deossification of the shaft of the femur and atrophy of the soft tissues suggesting that this patient has had considerable pain for a period of time. There is calcific debris in the soft tissues lateral to the right iliac crest which may be the donor site of the previous surgery. There is a well defined lytic lesion in the femoral neck, proximal to the intertrochanteric line, with considerable sclerotic bone with the confines. It must be emphasized that there was previous surgery and bone chips are in place accounting for the increased density.

The history suggests that at surgery a lesion was found which did not cause much concern and conservative surgery was performed. This might have been a chondroma or fibrosarcoma. Since it is a slow growing lesion the most likely possibility is fibrosarcoma.

This could be a chondrosarcoma, but I would expect that with recurrence there would be some evidence of soft tissue calcification.

Other differential diagnoses might include chondroblastoma, fibrosarcoma, pigmented villonodular synovitis, reticulum cell sarcoma, and lymphoma.

Dr. Edeiken's impression: FIBROSARCOMA.

Roentgenologic Impressions Submitted by Mail

Chondrosarcoma	37
Osteosarcoma	14
Chondroblastoma	10
Fibrosarcoma	10
Others	33

Dr. Edeiken: I think chondrosarcoma is a likely diagnosis. Osteosarcoma is unlikely because the lesion is well defined in the neck of the femur; it appears to be growing slowly, and there is no evidence of calcification. There is a chance that it is an osteosarcoma. Chondroblastomas occur in the epiphysis; they can occur in the greater or lesser trochanter. They usually start also before the epiphysis is closed and this is a 36-year old woman; this central lesion seems to be in the neck of the femur and metaphyseal area.

Dr. Regato: Dr. J. C. Lemon, of Denver, also suggested fibrosarcoma. Dr. J. D. Cox, of Colorado Springs, offered chondroblastoma and Dr. E. Salzman, of Denver, chondrosarcoma.

Operative findings: On October 27, 1966, the patient was operated upon: a Jewett nailing, bone graft and biopsy were done. A diagnosis of fibrosarcoma was rendered and the patient was taken again to the operating room in November, 1966; a right hemipelvectomy was performed.

Dr. Spjut: This lesion has histological features, mature bone and a fairly cellular stroma, that at first glance might suggest parosteal osteosarcoma. However, the radiographic

Fig. 1—Marked deossification of the head and neck of the femur.



Fig. 2—Section of surgical specimen showing tumor of the head and neck of femur extending outside the cortex.

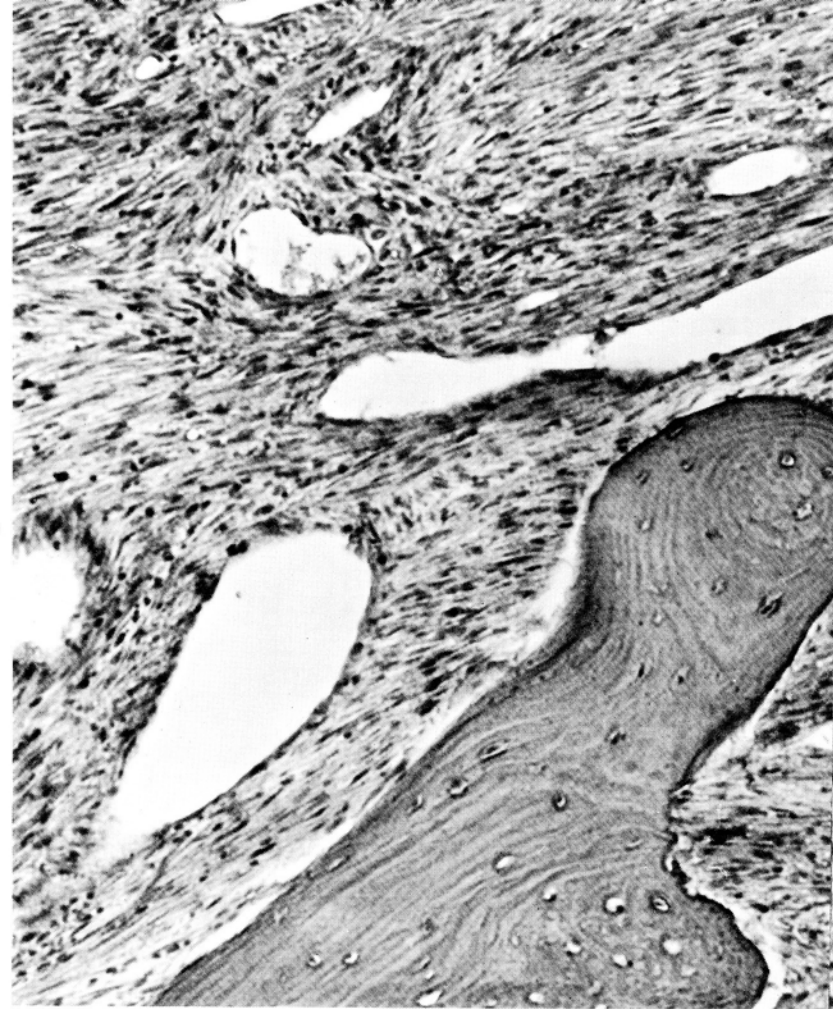


Fig. 3—The spindly fairly cellular stroma of the fibrosarcoma. The bone is residual and not neoplastic. Hematoxylin-eosin 180X.

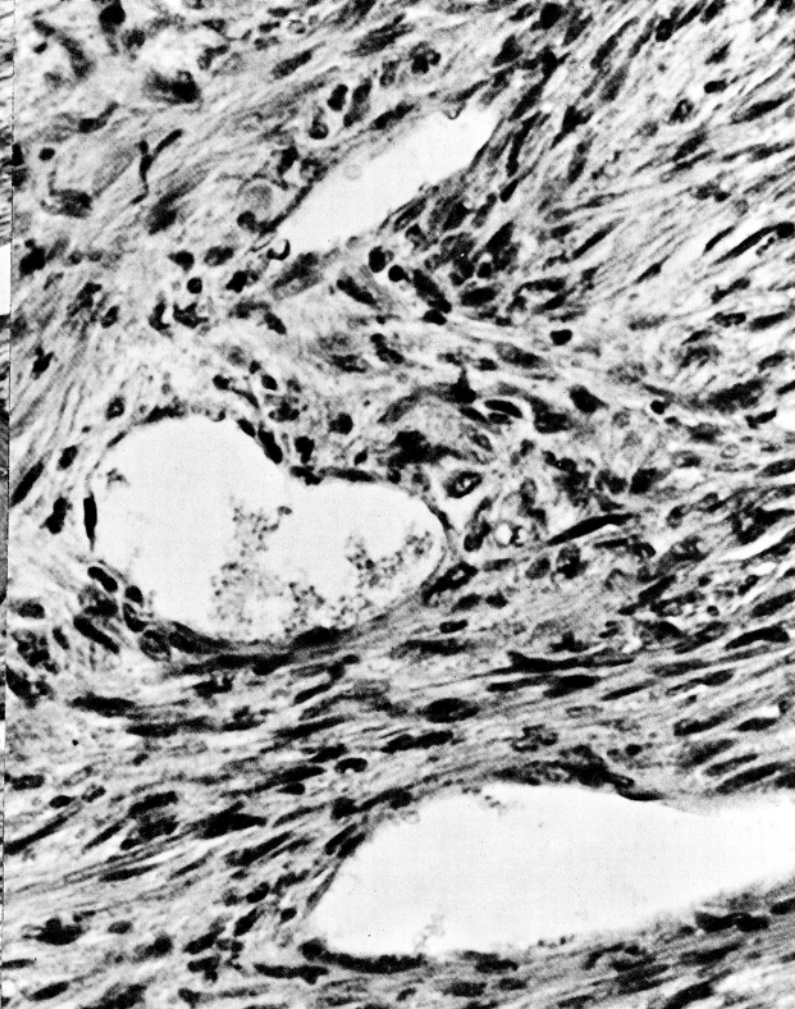


Fig. 4—Under higher magnification the good differentiation of the lesion is appreciated. Hematoxylin-eosin 370X.

findings do not support this diagnosis. What are the alternatives? One has to consider fibrous dysplasia, desmoplastic fibroma, osteosarcoma and fibrosarcoma. Fibrous dysplasia, I believe, can be ruled out as the lesion does not have metaplastic bone formed in the stroma. The bone appears to be lamellar and not of the woven variety. The ossifying fibroma generally does not have the cellularity of the stroma that we see in this case; however, mature bone and bone surrounded by osteoblasts may very well be seen in the ossifying fibroma. Some of the bone may be woven bone; also, this is a rather uncommon lesion in long bones. Desmoplastic fibroma is a tumor that deserves high consideration; we know that it is capable of destroying bone. However, this case differs from the desmoplastic fibroma in that the latter lesion usually has a more mature stroma with well-defined collagen fibers making up at least part of the stroma. As far as osteosarcoma is concerned, we have no particular evidence to support this in that there is no evident malignant bone or osteoid. The stroma in the tumor of this case is orderly and well-differentiated; however, it is more cellular than one would expect for most benign lesions. The nuclei and the cells are spindled, and the nuclei are larger than one would expect for fibrocytes. Occasional normal appearing mitotic figures are seen. How do we account for the mature bone that seems to form part of this lesion? Ordinarily, if one demonstrates malignant tumor bone or osteoid formation in a lesion such as this it should be classified as an osteosarcoma. In this lesion the bone appears to be mature and gives the impression that the tumor is invading and surrounding bony trabeculae. Whether this is an accurate interpretation or not cannot be determined from one slide.

This tumor would fall into the category of a fibrosarcoma, the cellular stroma being the key in this diagnosis. Fibrosarcoma tends to occur in the long bones, especially the tibia and femur and occurs at a later age than does osteosarcoma. There is practical importance in trying to distinguish fibrosarcoma from an osteosarcoma in that, in most series, the prognosis is somewhat better. The five year survival rate for fibrosarcoma ranges up to 31 per cent.

Since tissue from the 1962 operative procedure was not available for review, one can only speculate as to its nature. It is conceivable that there was a pre-existent benign lesion such as fibrous dysplasia. At present we have no histological evidence for this. The interval of four years is longer than the usual period of symptoms for patients with fibrosarcoma. I suppose that it is conceivable that fibrosarcoma was present four years ago.

Dr. Spjut's diagnosis: FIBROSARCOMA.

Histopathologic Diagnoses Submitted by Mail

Fibrosarcoma	72
Leiomyosarcoma	25
Fibrous dysplasia	15
Desmoplastic fibroma	9
Osteosarcoma	6
Others	21

Dr. Spjut: Histologically we have what appears to be mature bone. There has been one case reported of leiomyoma of bone from England and the photomicrographs suggested that possibility but we can't know whether it is fibrous or not so that if one is going to say leiomyoma of bone we would have to presume that this is secondary and

invasive. We don't have what is a strict histological definition of fibrous dysplasia.

Dr. Regato: Dr. L. Lowbeer, of Tulsa, offered a diagnosis of monostotic fibrous dysplasia with focal fibrosarcomatous transformation. Dr. A. O. Severance, of San Antonio, offered leiomyosarcoma.

Subsequent history: In September, 1967, the patient presented pulmonary metastases and was receiving chemotherapy.

Dr. Hatcher: With a lesion of these characteristics in this location the first diagnosis to be considered should be chondrosarcoma. The history of three years of pain before the final surgery was applied is suggestive of chondrosarcoma. Synovial sarcoma may occur in this region about the hip and produce the same roentgen appearance; a synovial sarcoma may be predominantly a fibrous tissue tumor and not show the cleft and glands that the classical synovial sarcoma does. In regard to treatment of this patient, the hemipelvectomy, of course, is the most definitive treatment. It would be possible to have done a regional resection of the tumor, removing the entire hip joint, capsule and acetabulum, en-bloc, and then restore function by bringing the stump of the femur under the ischium and putting a bone transplant between the ischium and the stump of the ileum. Such surgery can be just as curative as hindquarter removal.

I think it might have been possible here. . . A simple disarticulation would not have been wise because this tumor inevitably grows out of the neck and into the hip joint so that the entire hip joint has to be extirpated.

W. J. Frable, M.D., Richmond, Va.: The original diagnosis on frozen section was osteogenic sarcoma; the subsequent diagnosis on the permanent section was fibrous dysplasia. The patient had recurrence and pain for the next 3 years, finally coming to another biopsy prior to her hemipelvectomy. Now she is living and reasonably comfortable. She has metastases present in her skull, ribs and parenchyma of the lung that are being reasonably controlled with Vincristine and steroid therapy.

L. Lowbeer, M.D., Tulsa, Oklahoma: I admit that there was no woven bone in my slide. From the history and from the presence of bone, I think this may be one of the cases in which fibrosarcoma originated on the basis of fibrous dysplasia.

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3. Polyostotic Fibrous Dysplasia

Contributed by **J. F. Kuzma, M.D.** and **J. E. Bell, M.D.**, Milwaukee, Wisconsin

THE PATIENT was a 32-year old man in January, 1962, when he gave a history of frontal boss since childhood and of an automobile accident 7 years previously: several ribs had been crushed and the skull had also been injured. On examination there was obviously deformity of the frontal region and of the rib cage; there was also proptosis of both eyes and divergent strabismus. Serum calcium was 9.7%, phosphorus 3.0 mgm% and alkaline phosphatase 34 K.A. units.

Dr. Edeiken: The chest roentgenogram reveals many expanded ribs with "ground glass" density. These changes characterize fibrous dysplasia. The skull roentgenogram reveals prominent frontal bossing with increased density extending throughout the paranasal sinuses. Scattered throughout are radiolucent areas.

Fibrous dysplasia is the only condition which will produce this appearance.

Elevation of the alkaline phosphatase is not unusual in fibrous dysplasia of the skull and may be a sign of superimposed osteosarcoma. The radiolucent lesions within the skull density are probably fibrous tissue and part of the fibrous dysplasia. However, one must also consider the unlikely possibility of sarcomatous degeneration.

Dr. Edeiken's impression: FIBROUS DYSPLASIA.

Roentgenologic Impressions Submitted by Mail

Fibrous dysplasia	45
Paget's disease	16
Neurogenous tumor	10
Van Buchen's disease	5
Others	23

Dr. Edeiken: This is a 32-year old patient, somewhat young for Paget's. Neurofibromatosis very infrequently involves the bone from within and commonly will show notching of the ribs on the inferior margins; they may show single or multiple interosseous lesions but never the picture we have here. And Van Buchen's disease—I heard that diagnosis last night.

Dr. Regato: Dr. M. Bischoff, of Denver, Dr. J. W. Barber of Cheyenne, and Dr. L. O. Martinez, of Miami, also submitted an impression of fibrous dysplasia. Dr. R. Henschel, of Saginaw, offered Paget's disease.

Operative findings: In December, 1966, biopsies of the skull and ribs were done; the fragments were whitish in color, and rubbery, as well as bony, in consistency.

Dr. Spjut: This lesion appears to fall in the category of the fibro-oseous lesions of bone; those most likely are fibrous dysplasia, ossifying fibroma and fibro-osteoma. Since this patient apparently has multiple bony lesions, it would follow that this is not an ossifying fibroma of bone as these are considered to be monostotic lesions. Histologically, the case at hand demonstrates a fairly characteristic pattern of fibrous dysplasia, that is, evidence of metaplastic bone forming from a fibrous stroma. If one polarizes the section it is possible to demonstrate the rather disorganized pattern of woven bone as compared to the contoured pattern seen in lamellar bone. Ordinarily, one does not expect to see osteoblastic rimming in fibrous dysplasia and this is used as a point in the differential diagnosis between fibrous dysplasia and ossifying fibroma. Lamellar bone and at times residual woven bone are seen in an ossifying fibroma. Fibro-osteomas are dominantly lamellar bone. Osteoblastic rim-

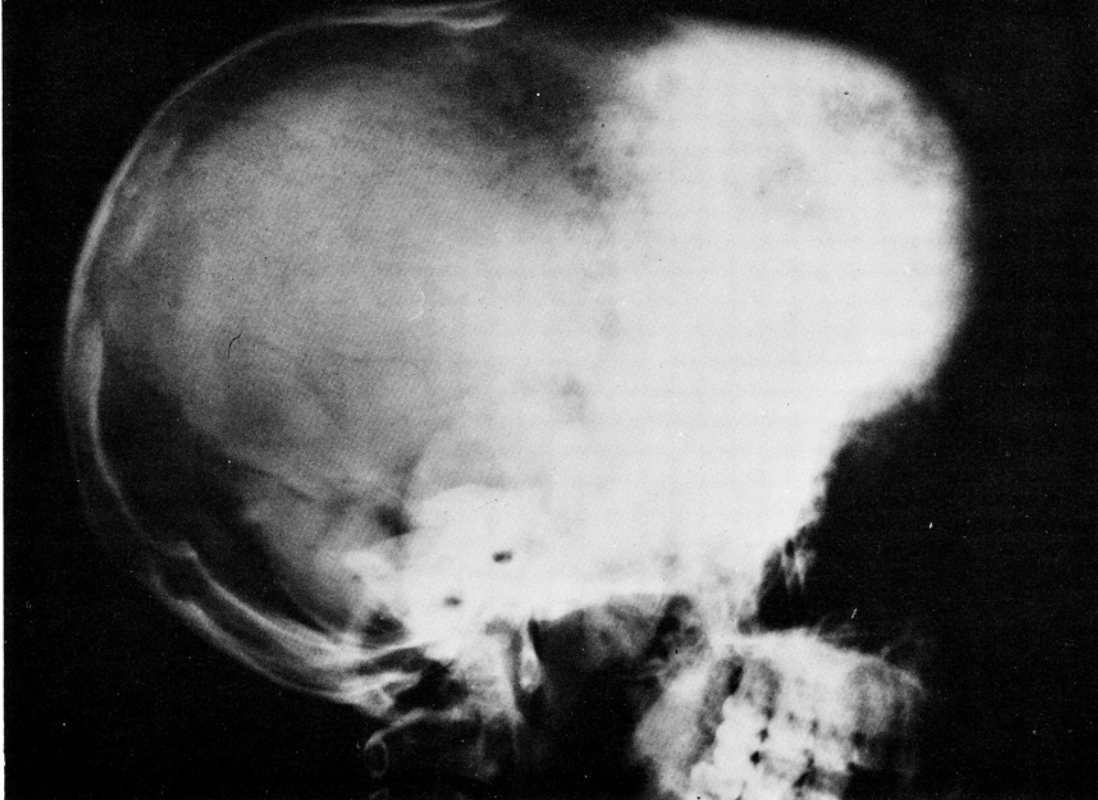
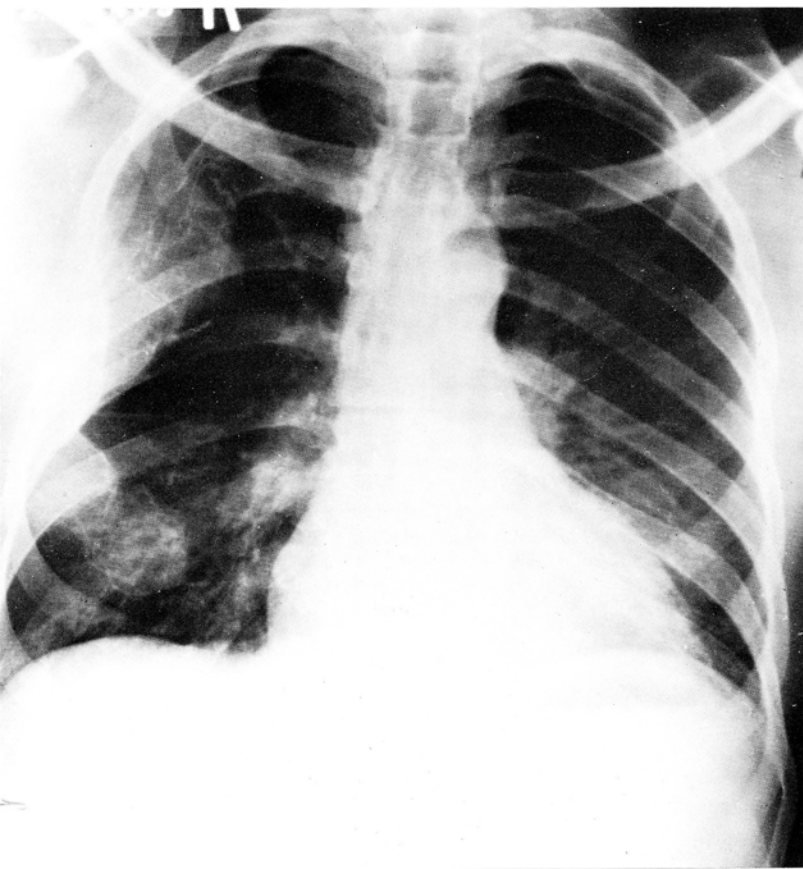


Fig. 1—Prominent frontal boss with increased bone density.

ming is seen in some areas of this lesion and may very well indicate that there has been some injury to the area from which this biopsy was made. However, we have seen other cases of polyostotic fibrous dysplasia in which osteoblastic rimming was demonstrated. We do not feel that the presence of this rules out the possibility of fibrous dysplasia particularly when one is dealing with multiple lesions. It is worthwhile to have a reasonably strict histological definition of fibrous dysplasia, otherwise such lesions as osteomas, ossify-

Fig. 2—Expanded ribs with "ground glass" density.



ing fibromas and non-ossifying fibromas, even unicameral solitary bone cysts, will be found among lesions designated as fibrous dysplasias. This merely serves to clutter up the diagnosis of this lesion. In addition there are clinical implications attached to fibrous dysplasia that require elucidation.

Dr. Spjut's diagnosis: FIBROUS DYSPLASIA.

Histopathologic Diagnoses Submitted by Mail

Fibrous dysplasia	94
Ossifying fibroma	9
Paget's disease	8
Others	24

Dr. Spjut: A fibrous dysplasia is just another form of ossifying fibroma; in fact, the osteoma of bone is considered the end-stage of a fibrous dysplasia. Histologically, there are some differences; I believe that probably we should adhere to these differences rather than just dumping all fibrous lesions with a little bit of bone into the designation of fibrous dysplasia. The ossifying fibroma in this case can be ruled out because we don't have osteoblastic rimming. In some cases of fibrous dysplasia, one will find lamellar bone, and not only lamellar bone but may find osteoblastic rimming. Lamellar bone may be formed in fibrous dysplasia, particularly in lesions that may have been traumatized or fractured through. The healing of the fracture will produce lamellar bone in cases of fibrous dysplasia. I don't see any histological features here that would lead me to say that this was Paget's disease.

Dr. Regato: The experts were almost unanimous in their diagnosis of fibrous dysplasia.

Subsequent history: This patient has been living without additional difficulties except for occasional syncopes requiring hospitalization. His serology is reported negative. He was last seen in June, 1967.

Dr. Hatcher: The clinical history does not state anything about possible skin pigmentations. About one-third of the patients with fibrous dysplasia will have also skin pigmentation. This is not a necessary part of the disorder. This in-

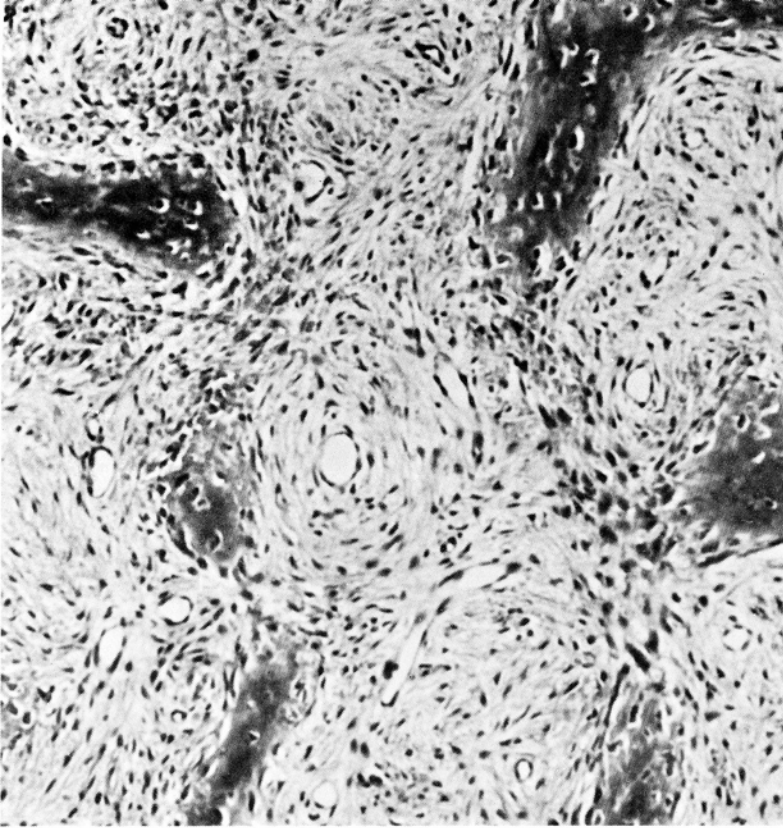


Fig. 3—The woven bone spicules of the fibrous dysplasia. The bone appears to spring from the fibrous stroma. Osteoblastic rimming of the trabeculae is inconspicuous. Hematoxylin-eosin 180X.

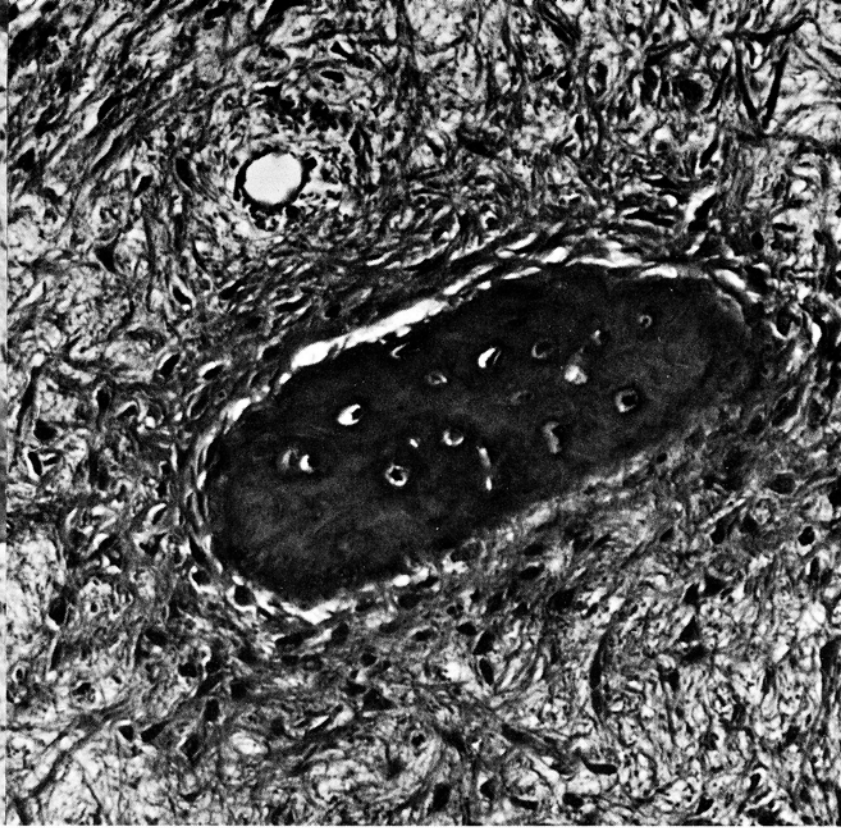


Fig. 4—Higher magnification demonstrates the relationship of the osteoid trabeculum to the stroma. Hematoxylin-eosin 300X.

involvement of the skull is quite characteristic, of course, and I am sure would lead most to make the diagnosis simply from seeing the man on the street. His alkaline phosphatase being elevated in middle age indicates that disease is probably still active. The complications from fibrous dysplasia can be quite serious. Anosmia is quite common, and progressive blindness could occur from gradual obliteration of the optic foramina. This man should have periodic visual field examinations to be sure that his optic nerves are not being depressed and if so a neurosurgeon can unroof the optic nerves and sometimes retard the onset of total blindness. A plastic surgeon might be tempted to pare off this large bony mass from the forehead. If he did, I am sure he would regret it because this can be extremely bloody and the possibility of producing any great improvement in his appearance would not be great. It might reduce his head size but it would not relieve the proptosis. Another possible complication, but a relatively rare one, is the development of sarcoma. Most of the instances of sarcoma that have appeared in areas of fibrous dysplasia, have been in those patients who, many years ago, were irradiated in an attempt to control the progression of the fibrous dysplasia. I don't

think anyone nowadays would irradiate such a lesion; it is very fortunate indeed that this particular disease involves mainly the outer table of the skull and not the inner table. It grows outward instead of inward so that cranial symptoms other than those from the cranial nerves are not common.

J. F. Kuzma, M.D., Milwaukee, Wisc.: This patient is colored so that the question of pigmentation can not be raised. He does have right optic nerve atrophy and beginning changes in his visual fields. He has had many admissions to the hospital primarily for alcoholism and trauma. The background of his blackouts and syncopal attacks has not been established.

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4. Alveolar Soft Parts Sarcoma of the Forearm

Contributed by **J. F. Kuzma, M.D.** and **J. E. Bell, M.D.**, Milwaukee, Wisconsin

THE PATIENT was a 70-year old lady in March, 1967, when she presented an ulcerated swelling of the right thumb accompanied by tender swelling of the forearm; there was a history of fracture and dislocation of the right ulna, fifteen months previously with open reduction and wiring. On examination there was an extensive tumefaction of the distal two-thirds of the forearm and of the lateral aspect of the hand; there was evidence of important super-

ficial collateral circulation; and a tender movable axillary adenopathy. The hemoglobin was 9.6 gm%; the urine showed 3+ sugar.

Dr. Edeiken: There is marked deossification of both forearm bones with an area of destruction in the mid portion of the radius. The soft tissues show a large mass and there is a phlebolith present in the soft tissues of the mid forearm. This is the appearance of a very vascular lesion as

indicated by the marked deossification of the bone and the presence of a phlebolith. The destruction of a portion of the radius would suggest that this is a malignant tumor, although it is possible that massive osteolysis could cause destruction without there being malignancy present. This could be a rhabdomyosarcoma, a fibrosarcoma, or a liposarcoma; the presence of the phlebolith suggests that it is a vascular tumor with disappearing bone or destruction by the malignant tumor; hemangiosarcoma is most likely.

Dr. Edeiken's impression: HEMANGIOSARCOMA.

Roentgenologic Impressions Submitted by Mail	
Osteomyelitis	34
Hemangiosarcoma	15
Osteosarcoma	11
Reticulum-cell sarcoma	10
Others	21

Dr. Edeiken: I can't see any reason to call this an osteosarcoma. Reticulum-cell sarcoma is a possibility and here we can have a very large soft tissue tumor mass with invasion of bone. However, I would not expect the mass to be so generalized throughout the forearm and I would not expect to see the evidence of vasculature on the film.

Dr. Regato: Drs. R. P. Spurr, of Denver, and J. W. Barber, of Cheyenne, also submitted angiosarcoma.

Operative findings: On March 20, 1967, an amputation was done above the elbow. A large mass, 8 cm in diameter, was found over the distal portion of the radius; the overlying skin was freely movable. On section, the mass was yellow-white in color with focal areas of hemorrhage and several satellites, 1 to 3 cm nodules. The middle third of the radius was completely destroyed.

Dr. Spjut: This tremendously destructive lesion presents somewhat of a histological problem. There are a number of possibilities that have to be considered. First, this does not appear to be a primary malignant tumor of bone, at least,

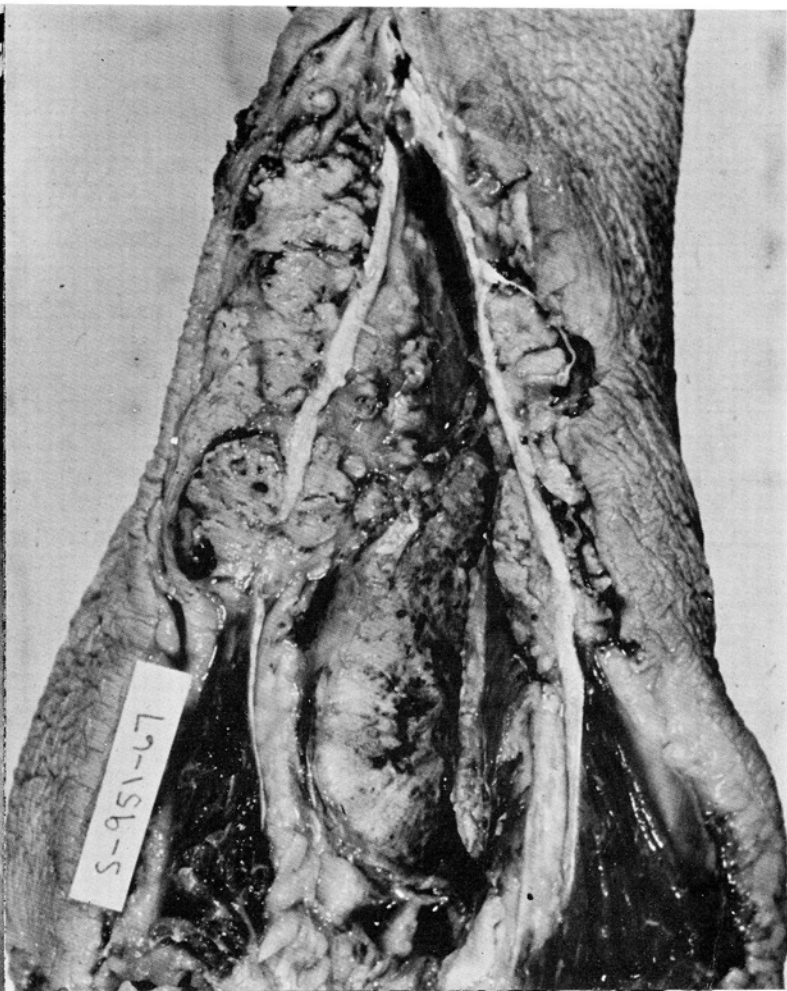
Fig. 1—Marked deossification of the bones of the forearm with an area of destruction in the mid portion of the radius.



not one that I have ever seen. One is left with the possibility of a metastatic or invasive malignancy. Of the metastatic lesions that seem possible with the histological pattern are renal cell carcinoma and adrenal cortical carcinoma. Another carcinoma that could very well be associated with the extensive bony destruction of this case is carcinoma of the thyroid but this does not, to me, have the histological features of a thyroid carcinoma. Of the invasive lesions, alveolar soft part sarcoma and the alveolar rhabdomyosarcoma seem to be the most reasonable.

The histological pattern (circumscribed nodules of tumor with intervening thin-walled blood vessels) suggested the possibility of an endocrine lesion. Because of the dense, acidophilic, granular cytoplasm of many of the cells, adrenal cortical carcinoma does metastasize to bone in approximately 17% of the cases. Renal cell carcinoma frequently metastasizes to and destroys bone. Histologically, the lesion is not entirely incompatible with either. However, the neat compartmentation makes one wonder. The alveolar rhabdomyosarcoma is a lesion that could possibly destroy bone but the lesions described under this title usually are fairly pleomorphic and one is ordinarily able to find strap cells with dense acidophilic cytoplasm. Search for these cells and cells with cross striations has been fruitless. The alveolar soft part sarcoma is an enticing possibility. The tumors may very well have the pattern demonstrated here. In addition, they do bear a resemblance to renal cell carcinoma and have been mistakenly designated as such. Special stains on the slides of this case have been non-rewarding. The oil-red-O stains were negative, the PAS stains did demonstrate some questionable crystalline structures that have been described in the alveolar soft part sarcoma. Crystals have been demonstrated electromicroscopically, but their significance is not known. A recent review of some 53 cases of alveolar soft parts sarcoma from Memorial Hospital, in New York,

Fig. 2—Surgical specimen showing tumor nodules.



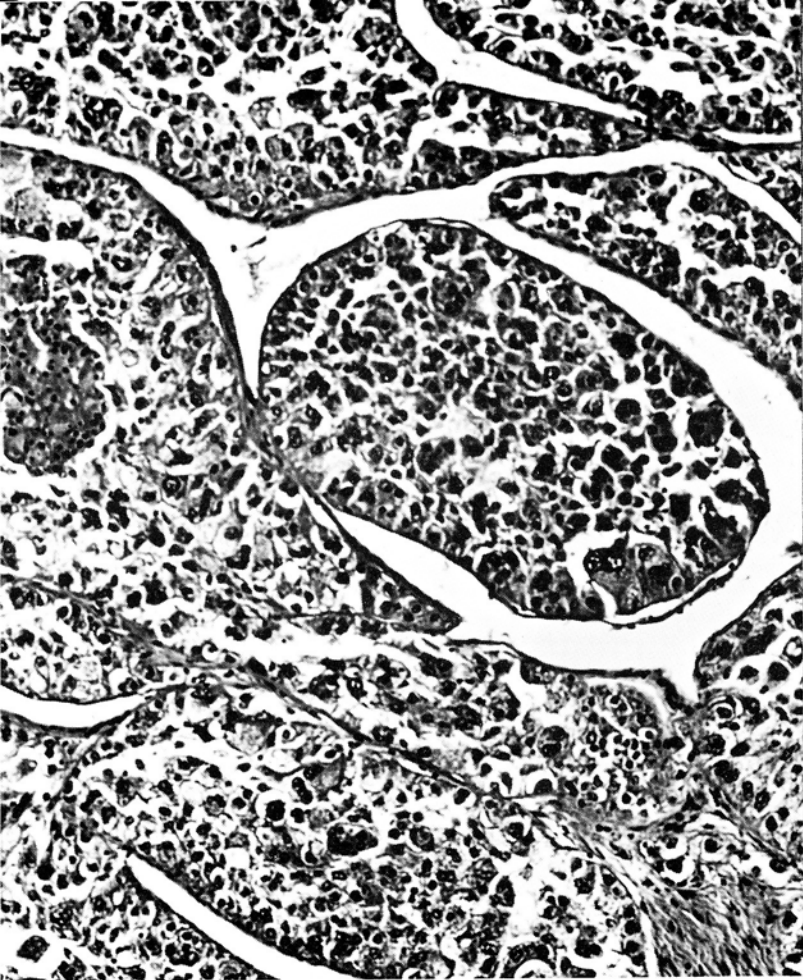


Fig. 3—The alveolar pattern of alveolar soft-part sarcoma. The vascularity is evident. Hematoxylin-eosin 125X.

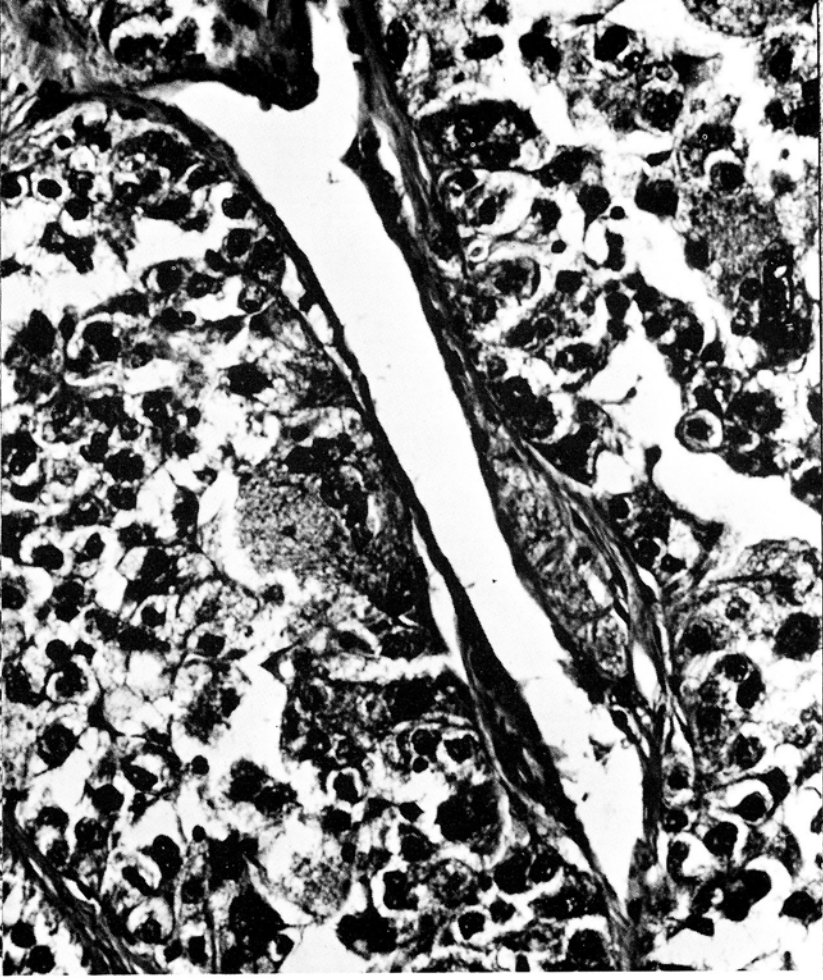


Fig. 4—Higher magnification demonstrating the resemblance to renal cell carcinoma. Hematoxylin-eosin 300X.

indicates that these lesions may occur in the forearm although they are more common in the lower extremities; the oldest patient was 56 years of age and the average age for women was 20. However, since the histologic features strongly suggest this lesion, one would have to consider this as the first possibility in spite of age. The published reports indicate that this lesion is slowly progressive but lethal; a five year survival rate of around 59% can be expected. Metastases may occur many years after the primary tumor has been removed.

One item mentioned in the history of this patient that raises some question is the isolated bit of information that the patient has a 3 plus sugar in the urine. There are many explanations, but there are reports that indicate that a number of neoplasms may be associated with disturbances of carbohydrate metabolism. Among them are: soft tissue and bone tumors and many different carcinomas.

Dr. Spjut's diagnosis: Probable ALVEOLAR SOFT-PARTS SARCOMA.

Histopathologic Diagnoses Submitted by Mail

Metastatic carcinoma (renal 26)	53
Alveolar soft tissue sarcoma (+ rhabdo).....	51
Angiosarcoma	12
Synovial sarcoma	9
Malignant granular-cell myoblastoma	8
Others	17

Dr. Spjut: The diagnoses given by the pathologists point up the problem in this case: trying to differentiate a metastatic carcinoma, renal in particular, from the alveolar soft

part sarcoma, the rhabdomyosarcoma. If we look at these vascular spaces, we see that these cells are not endothelial cells; they are on the outside of the vessels. We don't have a malignant endothelium, which one would have to have in order to diagnose angiosarcoma. We don't have the features of synovial sarcoma that we pointed out in Case No. 1. Some have considered that the alveolar soft part sarcoma was a form of granular cell myoblastoma of the malignant variety; perhaps in that way these lesions might be related, but they are not so considered in more recent writings.

Dr. Regato: Dr. M. R. Abell, of Ann Arbor, and Dr. A. M. Ginzler, of Warren, Ohio, also submitted a diagnosis of alveolar soft parts sarcoma. Dr. W. J. Holaday, of Galveston, and Dr. H. Rodriguez, of San Antonio, preferred metastatic carcinoma. Dr. R. Boyer and Dr. R. M. Sherwin, of Colorado Springs, suggested malignant granular-cell myoblastoma. Dr. M. H. McGavran, of Saint Louis, offered angiosarcoma.

Subsequent history: In June, 1967, the patient was doing well except for a purulent draining sinus.

Dr. Hatcher: From the history given and the roentgenograms, I thought this might well be a lymphangiosarcoma; I wonder if in her earlier years she might not have had a radical mastectomy with chronic edema in that arm in which an angiosarcoma might have developed. In regard to treatment, it seems obvious that ablation of the extremity is the only possible means of removing the tumor. The history mentions palpable lymph nodes in the axilla; these could be due to reactive nodes. One might make sure by removing all of the axillary nodes and doing a fore-quarter resection.

J. F. Kuzma, M.D., Milwaukee, Wisconsin: The arm was fractured in a fall and for this she had surgical correction and no tumor was noted in that extremity. While this extremity was being treated for the fracture, a medical student found a bruit and he told about it to the attending physician who said: "Now look, we are treating the fracture, we know about this case". Later on when the lesion bled, the patient came back. She has not permitted any additional work-up or therapy. Our own diagnosis was metastatic renal cell carcinoma. Preliminary pyelograms suggested a lesion in the kidney, but on follow-up by tomograms, the lesion disappeared so that we don't know if she has something in her kidneys or not.

W. O. Brown, M.D., Scottsbluff, Nebraska: About 1932, Fred Stewart reported in the *American Journal of Cancer* a series of primary liposarcomas of bone; others subsequently decided that they were not but they had a morphology very similar to the alveolar soft part tumors that we see today.

Dr. Spjut: There are acceptable cases of liposarcoma primary in bone; those that I have seen were recognizable as liposarcomas as we ordinarily define them histologically in the soft tissues, but were centrally destructive lesions of bone which I suppose could radiologically fit with this lesion.

L. Lowbeer, M.D., Tulsa, Oklahoma: Wouldn't the reticulum pattern of this tumor be more or less decisive in order to find out whether this is an epithelial neoplasm or not. What is the reticulum pattern? Is it, or is it not, epithelial?

Dr. Spjut: We didn't do a reticulum stain. We did other stains trying to find some residual fat which we didn't find. The reticulum stains could very well help to decide whether this is epithelial or not.

A. M. Ginzler, M.D., Warren, Ohio: Dr. Spjut, do you consider these tumors identical with those that Smetana described as malignant non-chromaffin paraganglioma?

Dr. Spjut: These have been considered to be identical by many observers; my experience with this lesion is confined to 2 cases; one that I thought was metastatic renal cell carcinoma; but wasn't, and then this one.

R. G. Vernon, M.D., Dubuque, Iowa: I was interested in the apparent vascularity of this tumor and yet as I studied it over a long period of time I failed to find a single red blood cell in any of the spaces that were so obviously vascular. I wonder if there is any significance to this.

Dr. Spjut: I think these are vascular channels and they happen to be empty and I believe this is something we see pretty often on lesions that are vascular. You see hemangiomas that are empty; most of the sinusoids in liver biopsies are empty of red cells. I don't really know how to explain this. One might ask, are these lymphatic channels rather than blood vascular channels?

J. Kuzma, M.D., Milwaukee, Wisconsin: I believe there are two reasons why these vascular spaces are empty. One is, much of the blood was on the floor of the house when the patient had this massive bleeding, and the second is that the extremity was perfused after operation in an attempt to get an angiographic picture of the lesion.

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5. Osteosarcoma of the Sacroiliac Region

Contributed by: **E. M. Bricker, M.D.**, St. Louis, Missouri; **A. D. Maddox, M.D.**, Las Cruces, New Mexico and **O. R. Ramos, M.D.**, El Paso, Texas

THE PATIENT was a 43-year old woman in May, 1967, when she presented a recurrent mass in the right lower abdominal quadrant; first, in 1959, and again in 1965, a similar mass had been resected from the right iliac bone; in the latter occasion she was given chemotherapy and was offered a hemipelvectomy. On examination there was a palpable mass, 6 cm in diameter, adherent to the medial aspect of the iliac crest; there was no pain and the general condition was perfect.

Dr. Edeiken: This patient had a resection of a mass from her right iliac bone in 1959. These roentgenograms are not available. The first roentgenograms available were obtained in 1963 and reveal only a slight increase in density in the iliac bone adjacent to the sacro-iliac joint which is either post-operative or due to recurrent tumor. By 1965 the lesion had grown considerably with an obvious soft tissue mass with calcification extending superiorly. The densities within the soft tissue mass are homogenous and suggest the presence of calcified osteoid tissue and therefore an osteoid producing tumor.

The presence of soft tissue mass outside of bone indicates that this is a malignant process. It would be unusual for a benign osteoid producing tumor (such as an osteoblastoma) to extend outside of the bone. This patient was treated with chemotherapy and by 1967 we can still see the calcified mass. One must assume that there is a malignant osteoid producing tumor. The only two tumors that we need consider in our differential diagnosis under these circumstances are parosteal sarcoma and osteosarcoma.

The long history (8 years) would be unusual for a central osteosarcoma, but it may occur. It is not unusual for patients with parosteal sarcoma to have a long history.

Dr. Edeiken's impression: PAROSTEAL OSTEOSARCOMA.

Roentgenologic Impressions Submitted by Mail

Chondrosarcoma	46
Osteosarcoma	18
Others	23

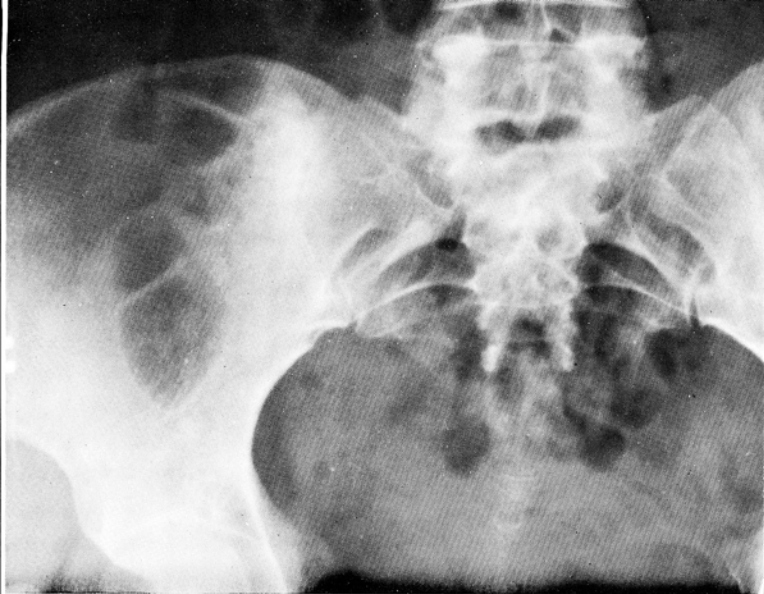


Fig. 1—Roentgenogram taken in 1963 showing only a slight increase in bone density in the right sacro-iliac area.



Fig. 2—Roentgenogram taken in 1967 showing considerable increase in the calcified mass.

Dr. Edeiken: This is the commonest site of chondrosarcoma and should be our first thought perhaps. The osteosarcoma, I would accept if they put in front of it perosteal. It's almost inconceivable that an osteosarcoma will grow so slowly over a period of 8 years.

Dr. Regato: Dr. B. L. Pear, of Denver, and Dr. J. W. Barber, of Cheyenne, offered an impression of chondrosarcoma. Dr. E. Salzman, of Denver, preferred parosteal osteosarcoma. Dr. J. W. Travis, of Topeka, wavered between the two.

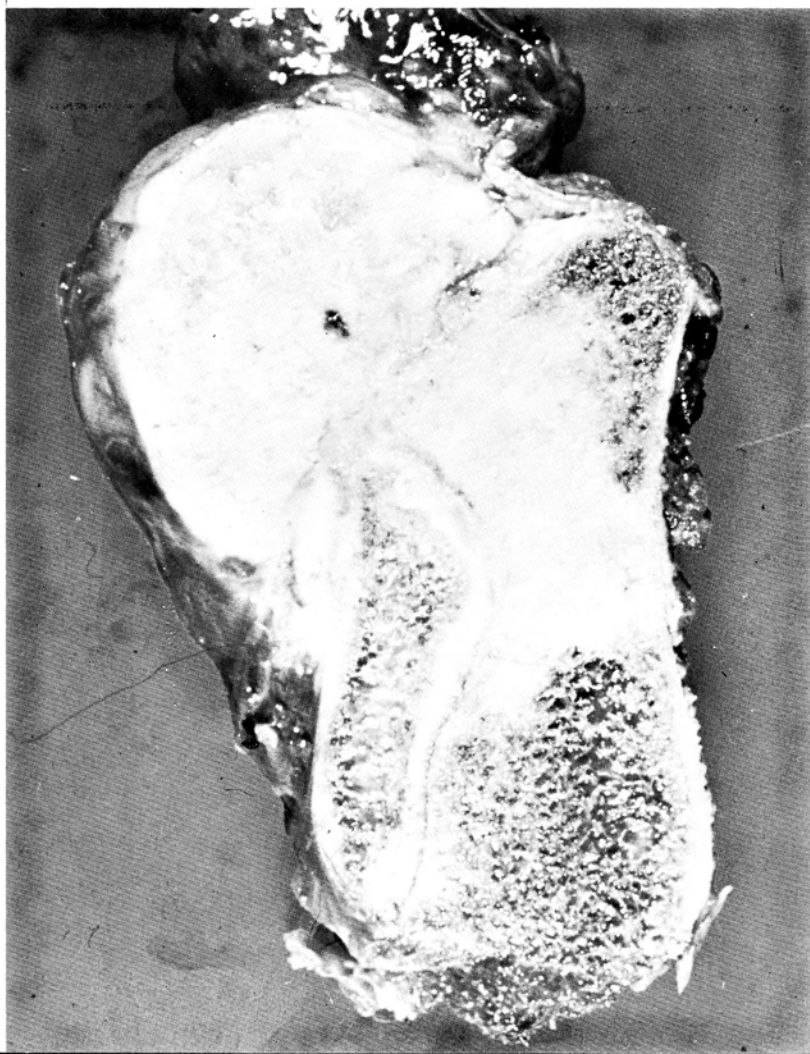
Operative findings: On May 29, 1967, a partial removal of the right iliac wing and sacrum was done. A bone graft was applied to replace the defect. The specimen measured 15 x 10 x 6 centimeters. The tumor bulged from the medial aspect of the ilium and measured 6 cm in height; there appeared to be no involvement of the sacro-iliac joint. Margins appeared free of tumor.

Dr. Spjut: Histologically, this lesion presents a rather difficult differential diagnosis. Could this be a parosteal osteosarcoma or is it an osteosarcoma of the fibroblastic type? Is this a chondrosarcoma, or even, considering the history, a benign reactive lesion? Because of the history of relentless recurrence, histological findings of bizarre osteoid, mitoses, and extremely cellular fibrous stroma, I do not feel that this is a benign lesion. Which of the malignant tumors could this be? Since there are only a few specks of cartilage in this lesion and it is dominantly fibrous and bone producing, it would seem unlikely that this is a chondrosarcoma. Because of the well-differentiated nature of the lesion, particularly the fibrous stroma and the reasonably well-formed bone and osteoid, parosteal osteosarcoma has to be considered. One feature that might be against this lesion is that apparently parosteal sarcoma has not been described as involving the pelvic bones. These tumors have been dominantly a lesion of the long bones, with an occasional one involving the scapula. Radiographically, this lesion does not fulfill the features of parosteal osteosarcoma.

What about osteosarcoma? The history is of a rather long duration. The question might be raised, is this history compatible with an osteosarcoma, particularly of the pelvic bones? One has to recognize, however, that patients with osteosarcoma are curable and that some of the lesions have unusual courses. Drs. Ackerman, Edeiken, Farrell, and I

have reviewed the case of a man who survived 35 years after treatment for osteosarcoma. Since we do not have sections of the previous specimens, we might speculate about a pre-existing parosteal osteosarcoma. The history might suggest this, but now the only recognizable element is osteosarcoma. Histologically, this lesion appears to be well-

Fig. 3—Surgical specimen of tumor bulging from medial aspect of iliac bone.



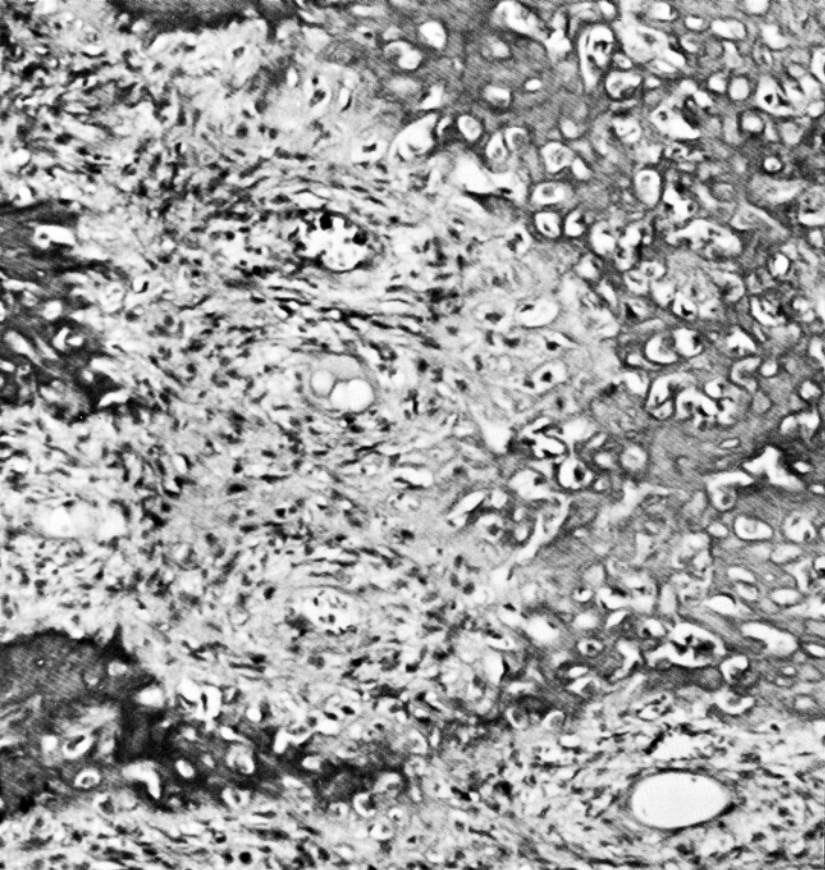


Fig. 4—Malignant fibrous stroma and cartilage. Hematoxylin-eosin 180X.

differentiated. The stroma is not particularly pleomorphic although cellular; a number of mitoses can be found. The osteoid is bizarre and appears malignant. The lesion is invasive of the fat at the periphery. Study of the periphery of this tumor shows that it is immature, an observation that is helpful in ruling out a reactive lesion, particularly one of this duration. My diagnosis in this case is osteosarcoma of the fibroblastic type, perhaps with an antecedent parosteal osteosarcoma.

Generally, the 5 year survival from osteosarcoma is 15%-20%. Different sites vary as to expected survival, e.g. tibial lesions are related to a better survival than are femoral or pelvic osteosarcomas. The histological pattern also influences survival with fibroblastic having a better rate than chondroblastic or osteoblastic.

Dr. Spjut's diagnosis: OSTEOSARCOMA, dominantly fibrous.

Histopathologic Diagnoses Submitted by Mail

Osteosarcoma	52
Parosteal osteosarcoma	33
Cartilaginous tumor	14
Juxtacortical osteosarcoma	9
Ossifying fibroma	5
Myositis ossificans	5
Others	27

Dr. Spjut: Histologically I don't believe we have any evidence to support a malignant or benign cartilaginous tumor as there are only a few bits of cartilage in this lesion. Some of the very poorly differentiated chondrosarcomas may be dominantly spindle-celled, but still one finds recognizable cartilage; in this lesion we have a combination of malignant cartilage but a great deal of malignant bone and osteoid plus the malignant fibrous stroma so I don't believe we can support that diagnosis. Ossifying fibroma can't be supported due to the fact that one would have to agree that

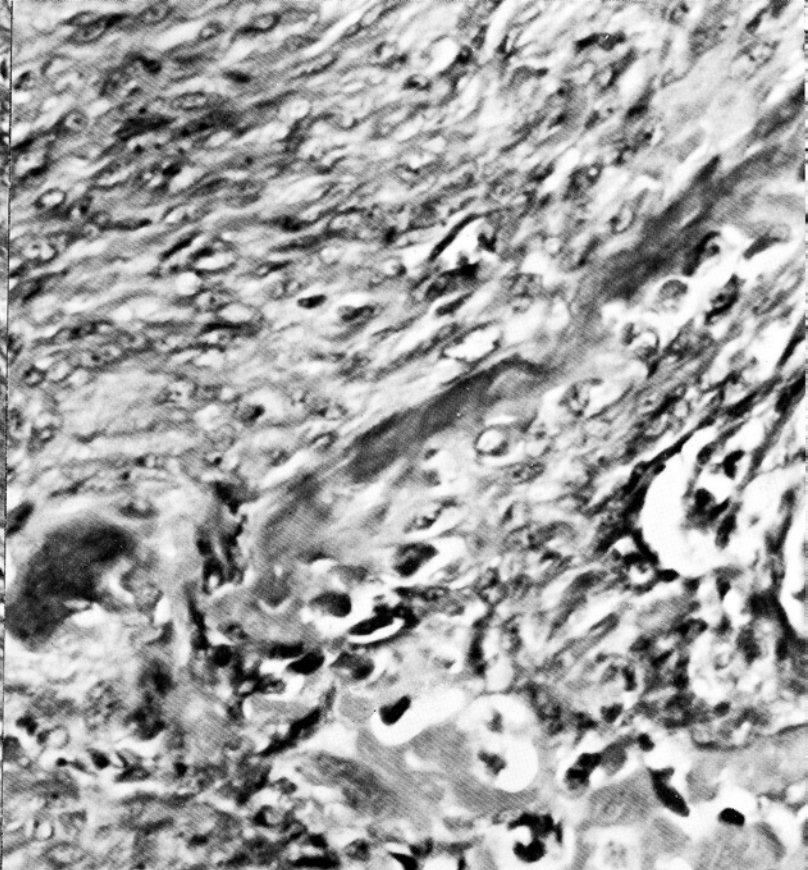


Fig. 5—Malignant fibrous stroma and osteoid. Hematoxylin-eosin 370X.

we have not only malignant stroma but malignant bone. Most ossifying fibromas occur in the head and neck area. The histological guide-post in making a diagnosis of myositis ossificans would be a study of the periphery of the lesion searching for evidences of maturation which in this case were lacking.

Dr. Regato: Dr. J. B. Frerichs, of El Paso, and Dr. B. Sharma, of Portland, made a diagnosis of low grade osteosarcoma. Dr. M. R. Abell, of Ann Arbor, and Dr. T. H. McConnell, of Dallas, designated it as parosteal sarcoma.

Subsequent history: In October, 1967, the patient is well but has kept a foot drop and muscular weakness of the right lower extremities, requiring a brace; no significant pain was reported and there are no signs of tumor.

Dr. Hatcher: The lesions in this location are extremely difficult for successful surgical treatment. The usual osteosarcoma in this region if it extends medially, will grow over the anterior part of the vertebral bodies and the sacrum. I am glad to see that a regional resection was done rather than a hindquarter resection; tumors so located can be just as adequately treated by regional resection as by removal of the entire lower extremity and pelvis. The complication of a foot drop is a quite common one in trying to get tumors out of this region because the first sacral nerve and the 5th lumbar come across that area and are apt to get either stretched or traumatized. I think this is a highly successful result in a highly malignant tumor with a peculiar long history.

Editor's note: In June, 1968, this patient was reported to have had a lobectomy for treatment of a solitary pulmonary lesion; there was no evidence of local recurrence.

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6. Myxopapillary Ependymoma of the Sacral Region

Contributed by J. M. Pawlowski, M.D., Sacramento, California

THE PATIENT was a 40-year old man in December, 1966, when he gave a history of persistent pain, of fifteen years' duration, in the right lumbosacral region radiating to the knee; for about ten years there was also numbness of the right gluteal region and, recently, he had become impotent. On examination there were marked varicose veins of both lower extremities, saddle anesthesia of the right side and absence of both deep ankle reflexes. The spinal fluid was mildly xanthochromic with 15.4 mg% proteins and no leukocytes; calcium was 9.5 mgm% and phosphorus 3.8 mgm%.

Dr. Edeiken: The roentgenograms reveal a large expanding lesion of the posterior elements of the 5th lumbar and 1st sacral segments. A myelogram shows a complete obstruction at the mid position of L-4 centra with expansion of the column and considerable irregularity at the inferior margin of the column. This is obviously a long standing tumor and probably benign. One should consider chordoma, ependymoma, meningioma, neurofibroma, teratoma, epidermoid, and dermoid.

Chordomas will almost invariably protrude anterior to the vertebral column. They begin in the center of the centra and will erode the body of the vertebra from within. This tumor appears to have begun in the area of the posterior elements of the spinal canal and therefore chordoma is excluded.

It is possible that any of the other above mentioned tumors could cause this type of bone destruction, but the tumors which grow the largest are the embryonic type of tumors which would include ependymoma, epidermoid, and dermoid.

Dr. Edeiken's impression: EMBRYONIC TUMOR (ependymoma, teratoma, or epidermoid).

Roentgenologic Impressions Submitted by Mail

Chordoma	49
Neurofibroma	12
Ependymoma	10
Osteoid osteoma	7
Others	19

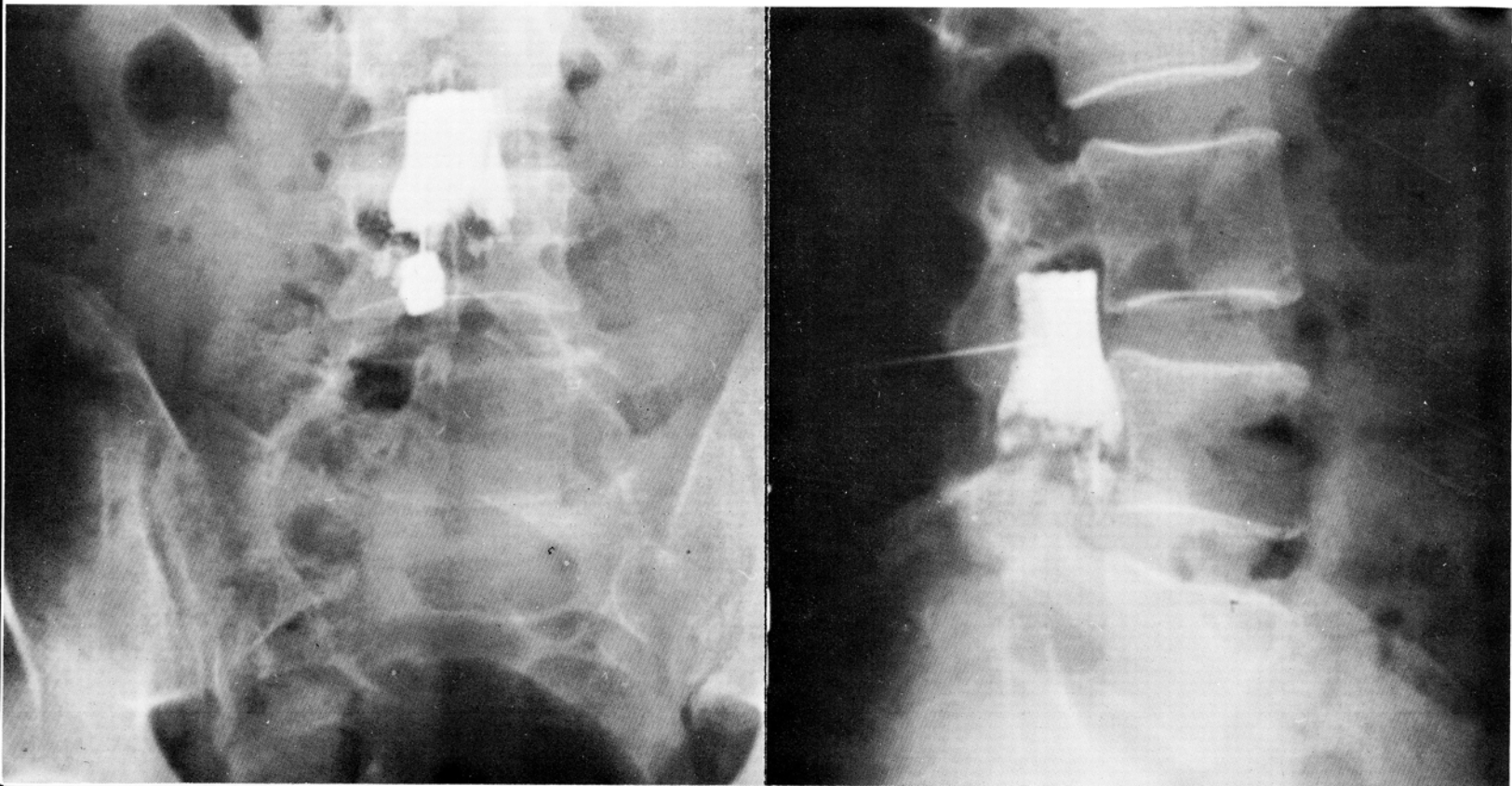
Dr. Edeiken: I'd like to say that this can't be chordoma. The neurofibroma is certainly possible; I haven't seen an intramedullary fibroma nor one so large. The ependymoma I think should be put right near the top. Osteoid osteoma? I don't see how.

Dr. Regato: Dr. P. A. Riemenschneider, of Santa Barbara, offered an impression of chordoma. Dr. C. H. Taggart, of Colorado Springs, preferred hemangioma and Dr. B. L. Pear, of Denver, ependymoma.

Operative findings: In December, 1966, a lumbo-sacral incision was done and a partially encapsulated bluish tumor was found; it had eroded the upper three segments of the sacrum and displaced the muscles laterally. The tumor extended from the lumbosacral disc and left side of the dural sac at L4. The capsule was incised and most of the tumor removed by suction. The capsule and remaining tumor were left in place. The material removed was reddish-gray in color and weighed 32 grams; it had mucoid appearance with areas of firmness and of hemorrhage.

Dr. Spjut: The histological features of this case present a rather intriguing differential diagnosis. The differential diagnosis would include chordoma, angiosarcoma, and ependymoma. At first glance, many areas in this lesion appear

Fig. 1 and Fig. 2—Myelogram showing complete obstruction at the mid point of L-4 centra due to large expanding lesion at the level of the fifth lumbar and first sacral segments.



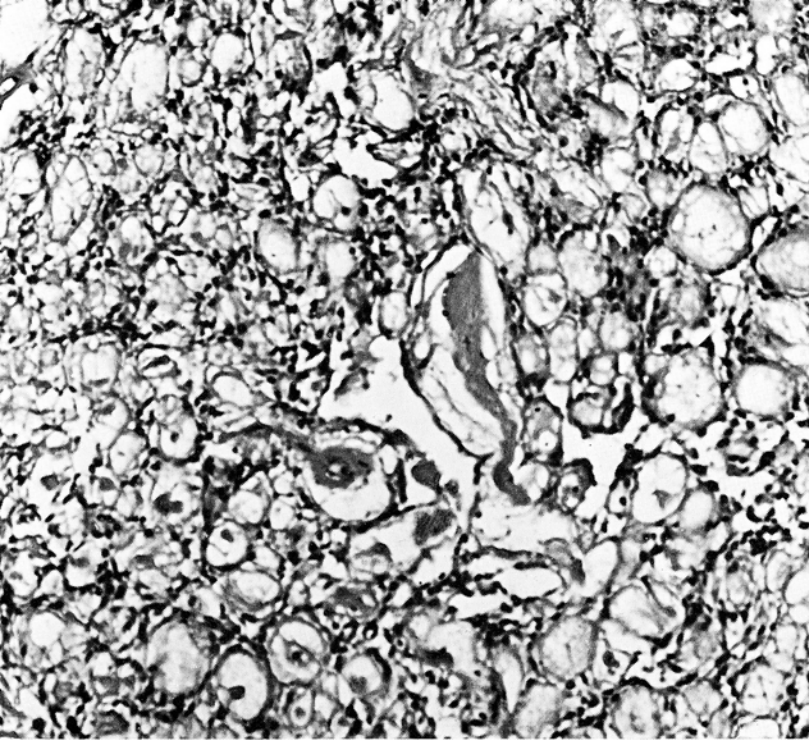


Fig. 3—An area of myxopapillary ependymoma that superficially suggests chordoma. However, the structures are papillary and surrounded by cells rather than being physaliphorous cells. Hematoxylin-eosin 100X.

to be physaliphorous cells. But as one searches around this section it becomes apparent that there are papillary structures. In addition, there are areas that appear epithelial. Other portions of the lesion appear to be vascular and one can at least imagine an endothelial component, suggesting the possibility of angiosarcoma. Careful examination of the large, rounded structures that appear to be physaliphorous cells, indicates that these are papillary structures and surrounded by cells. Physaliphorous cells usually have a nucleus and are large vacuolated cells and would not be expected to be surrounded by cells as seen in this case. Consequently, I believe that this lesion is not a chordoma and falls into the category of myxopapillary ependymoma. Ependymomas are the most common tumor of glial origin of the spinal cord, comprising 50% of the neoplasms of the filum terminale. The long history in this case is compatible with ependymoma. In the few cases of this particular type reported, bony destruction has been described. Most cases have presented as a subcutaneous mass, often mistaken for a

Fig. 5—An area in which the cells bear a close resemblance to ependymal cells. Hematoxylin-eosin 300X.

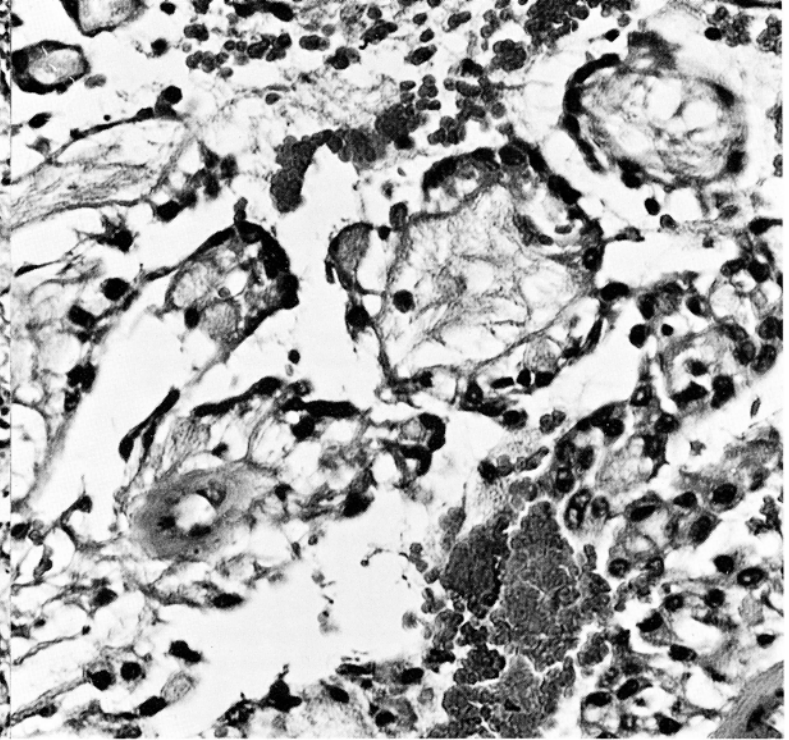
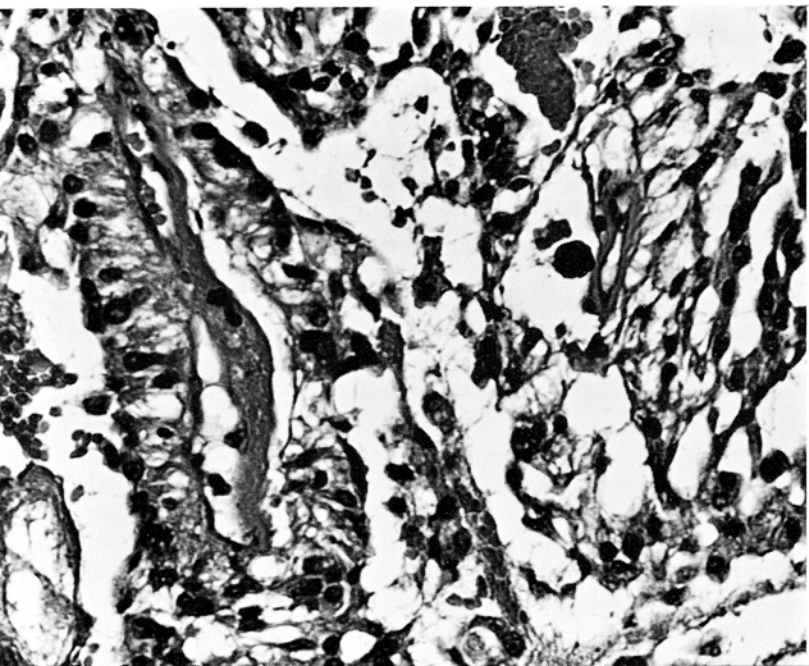


Fig. 4—Higher magnification of the papillary structures illustrating the cells and the congregation around blood vessels. Hematoxylin-eosin 300X.

pilonidal sinus. Histologically, the mucinous material in these lesions is positive by mucicarmine and alcian blue stains. There are no histochemical studies which will specifically identify this lesion. The long-term prognosis for this lesion is considered poor, but death or recurrence may not occur for years.

Dr. Spjut's diagnosis: MYXOPAPILLARY EPENDY-MOMA.

Histopathologic Diagnoses Submitted by Mail

Chordoma	109
Ependymoma	19
Angiosarcoma	6
Others	2

Dr. Spjut: We can say that this is not a chordoma because these are not physaliphorous cells. Those regions that appear to be angiomatous are really the papillary portion of this ependymoma. If one looks at these little areas carefully one will find that these are ependymal cells that project into papillary regions. Despite the majority, I will have to stick with myxopapillary ependymoma. This is not a well known lesion. It wasn't known to me until about a year ago. This is the first case I've ever seen but I guessed at it anyway.

Dr. Regato: Dr. C. P. Schwinn, of Los Angeles, also made a diagnosis of myxopapillary ependymoma of the filum terminale. Drs. Magda and John Kepes, of Kansas City, concurred in an emphatic diagnosis of ependymoma. Dr. C. O. Burdick, of El Paso, and Dr. H. K. Giffen, of Omaha, preferred chordoma.

Subsequent history: Following operation there was regression of pain but the saddle area of anesthesia persisted. In October, 1967, he was reported doing pretty well, needing two capsules of Darvon a day; he had some remaining sphincter problems but no motor deficit of lower extremities.

Dr. Hatcher: This is one of the cases I felt absolutely certain of; I thought it was surely a chordoma. In regard to the treatment, I suppose the only thing that can be done is to keep the spinal canal well decompressed and remove what tumor is possible to spare the nerve roots. I would like to ask if radiation has any effect on this type of tumor.

F. Buschke, M.D., San Francisco, California: I'm not aware of any publications that mention radiotherapy for this particular type of lesion in this location. I want to ask Dr. Spjut what is the relationship of this type of ependymoma with the intracranial ependymoma which we treat quite frequently: those respond and are controlled by radiation therapy alone.

Dr. Spjut: These lesions are ependymomas and this sub-classification is an attempt to explain their gross features: the rather abundant mucoid material; they are related to the intracranial ependymomas. Since they arise posteriorly some think that they arise from heterotopic ependymal cells; I think that it has been demonstrated that there are ependymal cells in the **filum terminale** which is presumed to be the origin of the ependymomas in this region. It is also of interest that ependymomas are the most common of the spinal cord tumors.

Dr. Regato: In respect to chordomas as well as ependymomas, the error has been committed of referring to them as radio-resistant because they do not react promptly to radiations but this is a concept that needs to be verified. Some of these tumors regress slowly but if they receive an adequate irradiation in time and quantity, the tumor may regress completely. Ependymomas are among the curable tumors of the central nervous system and spinal canal. There

are numerous cases reported in the medical literature. I have irradiated a few chordomas, at either end of the spine, simply because they were beyond surgical control or had recurred after surgery; some of them were rather voluminous yet did respond by a rather slow regression to the point of apparent cure but most of these cases recurred subsequently. I am not certain that chordomas are radiocurable but certainly they are radioresponsive. Ependymomas are curable by means of radiotherapy depending on their location and extent and the adequacy of the irradiation, a factor not often considered by those who speak about these lesions.

J. Kepes, M.D., Kansas City, Kansas: I like to think of **filum terminale** as part of the spinal cord, even though it is mixed in with the fibers of the **cauda**. The ependymal cells in the normal filum probably are not heterotopic; they make up a good portion of the filum because that is the portion of the spinal cord where everything else has gone, the nerve roots, and the nerve cells, and all that is left is a framework that still has portions of a central canal. However, we have had an occasional true heterotopic ependymoma that is pre-sacral.

References

Anderson, M. S. Myxopapillary ependymomas presenting in the soft tissue over the sacrococcygeal region. *Cancer*, 19: 585-590, 1966.

7. Aneurysmal Cysts of the Fibula

Contributed by **F. S. Parrish, M.D.** and **J. A. Murray, M.D.**, Houston, Texas

THE PATIENT was a 12-year old boy in November, 1966, when he complained of pain in the left knee which had been present for three months following trauma. On physical examination there was a firm, fixed mass on the lateral aspect of the left leg, just below the knee.

Dr. Edeiken: The roentgenograms reveal a rather large lesion in the proximal portion of the fibula which is confined by the epiphyseal line. There is a sudden transition between the tumor and the host bone both inferiorly and superiorly. The lateral cortex shows slight deformity indicating a pathologic fracture; the medial margin cannot be discerned.

The well defined margins would suggest that this is a slow growing tumor and probably benign. There is expansion of bone, a suggestion of a thin bone density bordering the medial portion of this lesion. The differential diagnosis should be made with a benign cartilaginous tumor. However, the expansile character strongly suggests aneurysmal bone cyst. Many aneurysmal bone cysts are secondary to an underlying process either benign or malignant and may be chondromyxoid fibromas or other benign tumors and osteosarcoma. For this reason, even though the appearance strongly suggests aneurysmal bone cyst, the possibility of osteosarcoma would not be completely surprising.

Dr. Edeiken's impression: ANEURYSMAL BONE CYST.

Roentgenologic Impressions Submitted by Mail

Osteosarcoma	31
Ewing's sarcoma	30
Aneurysmal bone cyst	13
Others	22

Dr. Edeiken: I think the primary diagnosis should be aneurysmal bone cyst.

Dr. Regato: Dr. A. Ritch, of Roswell, offered an impression of osteosarcoma. Dr. J. C. Lemon, of Denver, offered Ewing's sarcoma. Dr. J. W. Barber, of Cheyenne, and Dr. R. Henschel, of Saginaw, aneurysmal bone cyst.

Operative findings: On November 9, 1966, an excision of the proximal half of the left fibula was done without breaking into the tumor. The specimen measured 14.5 cm; it presented a fusiform enlargement at the metaphysis. The tumor itself measured 5.9 x 4.3 cm; on cross section it was firm and homogeneous, gray color with focal hemorrhages; the cortex was thin and apparently penetrated at one point.

Dr. Spjut: Is this an aneurysmal bone cyst? Is this an osteosarcoma with an outstanding vascular component, or is this a cystic giant cell tumor? This case was seen a year or so ago and, at that time, the problem presented was that of osteosarcoma versus aneurysmal bone cyst. First, how can we rule out the possibility of giant cell tumor? This patient is 12 years old and ordinarily one does not make a diagnosis of giant cell tumor in a patient this age unless the situation is perfect. Also, we note that radiographically this is a metaphyseal lesion; the epiphysis does not appear to be involved; this in itself would be against a giant cell tumor, although there have been rare giant cell tumors that were entirely metaphyseal in location.

In order to diagnose osteosarcoma, one has to have a malignant stroma and an associated malignant osteoid or bone. In areas of this lesion, there are extremely cellular stromal foci, plus there is a bony component that appears to be woven bone. If one studies this bone and the osteoid carefully, and particularly the surrounding osteoblasts, it



Fig. 1.—Expansile lesion of the proximal end of the fibula, confined by the epiphyseal line.

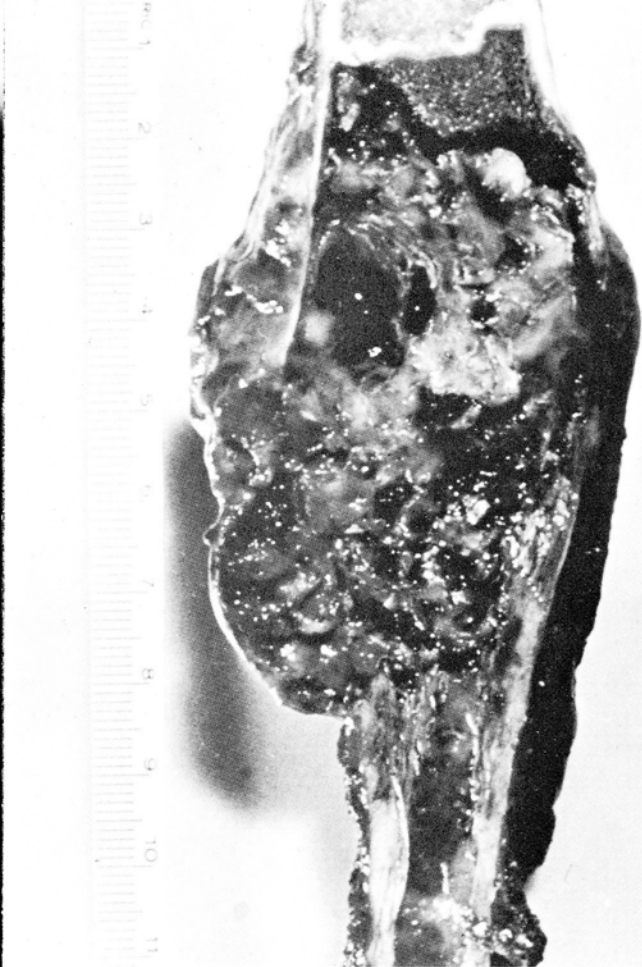


Fig. 2.—Section of surgical specimen showing homogenous lesion within bone.

becomes apparent that this is not malignant: it is reactive. Since this patient gives a history of trauma to this area, it might be surmised that a pre-existing lesion was traumatized, resulting in fracture and stimulation of reactive bone that we have in the lesion. The stroma is cellular and mitotic figures are present, but it does not come up to the standards of the malignant stroma. Numerous foci of multinucleated giant cells are present, but these are a rather common component of aneurysmal bone cysts. The large vascular spaces are reasonably characteristic of an aneurysmal bone cyst in that in some areas we find the spaces lined by multinucleated giant cells and in others there appears to be osteoid in the wall. We have seen bone lesions in which histological mistakes have been made both ways, i.e. aneurysmal bone cyst mistaken for osteosarcoma and osteosarcoma diagnosed as aneurysmal bone cyst. To avoid this, one needs good radiographic studies, a well-taken biopsy, and careful histological examination of the stroma and osteoid.

Aneurysmal bone cysts most commonly involve the long bones or the small tubular bones, although in early reports it was a fairly common lesion in the vertebral column. The lesion has been described as involving nearly all the bones of the body. What is the pathogenesis of this lesion? I do not have any specific answer to these questions. It has also been considered by some that aneurysmal bone cysts represent a vascular alteration of a pre-existing tumor, for example a giant cell tumor. Some suggest a vascular malformation, others believe that trauma plays a role and others that the hemangioma serves as the focus of origin. As one sees more and more bone tumors, it becomes apparent that there is a mixture of lesions. For example, we have seen a characteristic chondroblastoma with a small area of typical

aneurysmal bone cyst. On the other hand, we have seen a radiographically and histologically characteristic aneurysmal bone cyst with small foci that resemble chondroblastoma; there seems to be a mingling of types in many of the tumor and tumor-like lesions of the bone.

Dr. Spjut's diagnosis: ANEURYSMAL BONE CYST.

Histopathologic Diagnoses Submitted by Mail

Aneurysmal bone cyst	81
Bone cyst	25
Osteosarcoma	8
Others	29

Dr. Spjut: In order to make the diagnosis of bone cyst one almost has to have a non-specific histological slide, that is, one with a few bone wisps or fibrous tissue on it. Some solitary bone cysts may give us the appearance that we have seen today; those that have had previous curettage and bone chips placed in them may give bizarre patterns and that might be why those making this diagnosis thought that this had been previously treated. The osteosarcoma is one of the differential problems in an aneurysmal bone cyst in which there is a tremendous fibrous and osteoid reaction.

Dr. Regato: Dr. A. O. Severance, of San Antonio, and Dr. G. Simon, of New Haven, also diagnosed an aneurysmal bone cyst. Dr. M. H. McGavran, of Saint Louis, offered angiectatic osteosarcoma, whereas Dr. H. L. McGaffey, of Idaho Falls, preferred fibroblastic osteosarcoma.

Subsequent history: The patient was reported well in June, 1967.

Dr. Hatcher: Aneurysmal bone cysts of the upper end of the fibula are quite common in young children. I thoroughly approve of the excisional biopsy done. One might

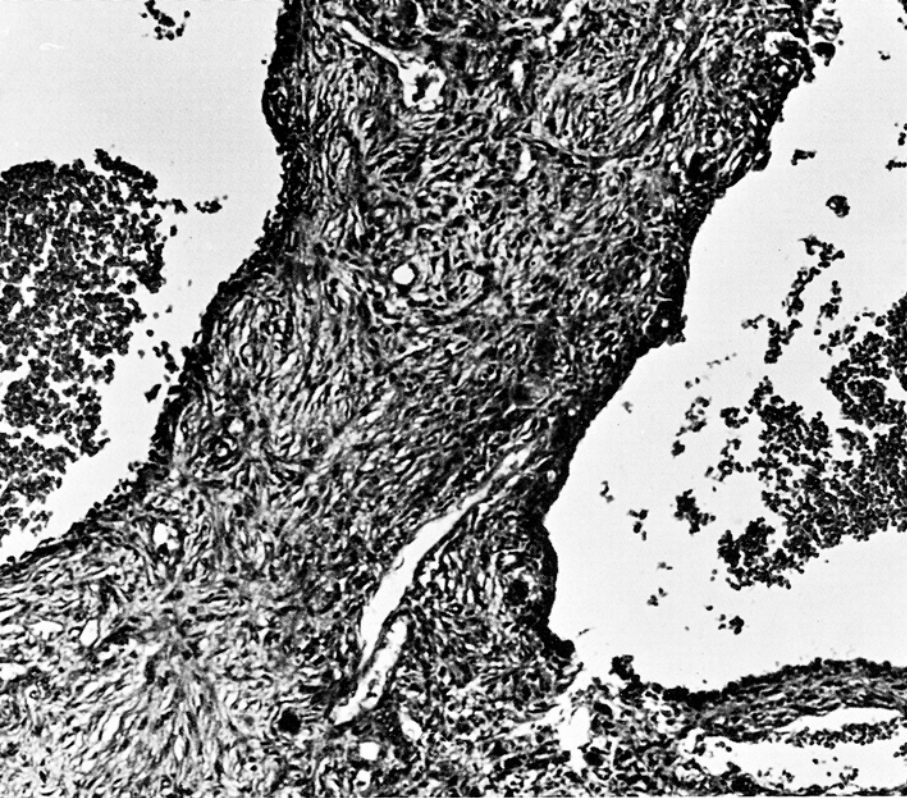


Fig. 3—Vascular spaces of the aneurysmal bone cyst. The fibrous trabeculum is fairly cellular. Hematoxylin-eosin 170X.



Fig. 4—Abundant calcified osteoid is present, but it does not have the features of malignancy. Hematoxylin-eosin 180X.

just as well take out the entire upper end of the fibula. It leaves no disability for the child providing the peroneal nerve is protected and it gives the pathologist such a nice specimen. The child is cured by the excisional biopsy. The aneurysmal bone cyst may be treated by simple curettage at times requiring preliminary tissue examination. These lesions will heal following a small dose of radiations as little as 800 rads. They also will heal sometimes, I am sure, spontaneously. Once in a while an aneurysmal bone cyst will give real trouble; it may be curetted repeatedly and packed with bone repeatedly and still fail to heal. Usually those have a narrow zone of this typical juicy tissue and

they fill up with blood. They may require regional resection of the entire involved area of the skeleton. Fortunately, they usually heal by surgical interference.

Dr. Regato: Dr. Hatcher, these lesions used to be diagnosed as giant cell tumors years ago, before they were recognized as being different entity, isn't that right?

Dr. Hatcher: Yes, and particularly is that true of the lesions appearing in the spine. I think many lesions have been reported as giant cell tumor of the spine that were in reality aneurysmal bone cysts.

References

Lichenstein, L.: Aneurysmal bone cysts. *Cancer*, 6: 1228-1237, 1953.

8. Myositis Ossificans of the Leg

Contributed by **A. O. Severance, M.D.** and **H. F. Elmendorf, Jr., M.D.**,
San Antonio, Texas

THE PATIENT was a 34-year old man in October, 1966, when he complained of a mass in the left leg; he was an aircraft mechanic who had been kicked by a cow two months previously. On examination there was a non-tender, palpable mass, on the lateral aspect of the middle third of the left leg.

Dr. Edeiken: The roentgenograms reveal an irregular densely calcified mass on the lateral aspect of the fibula at the junction of the upper and middle thirds. There is an irregular soft tissue mass associated. Below the main calcified mass there is a smaller calcification.

Since this lesion developed after trauma, the most outstanding consideration is myositis ossificans. However, one would expect the myositis to show a regular calcification throughout the entire lesion. The spotty calcification below the main density suggests a tumor rather than myositis. There is no erosion of bone or thickening of the cortex as

one would expect with periosteal chondroma. Parosteal sarcomas tend to surround the bone but may be eccentric early. One can often find a cleavage between the densely calcified parosteal sarcoma and the host bone.

Whereas this lesion appears to be a parosteal sarcoma, for the reasons listed above, it would not be surprising to find myositis ossificans.

Dr. Edeiken's impression: PAROSTEAL OSTEOSARCOMA.

Roentgenologic Impressions Submitted by Mail

Parosteal sarcoma	34
Myositis ossificans	21
Calcified hematoma	16
Osteomyelitis	6
Others	15

Dr. Edeiken: I don't really think that you can come to an absolute conclusion on this case. I think we can include



Fig. 1—Irregular and densely calcified mass in lateral aspect of the fibula without erosion of bone or thickening of the cortex.

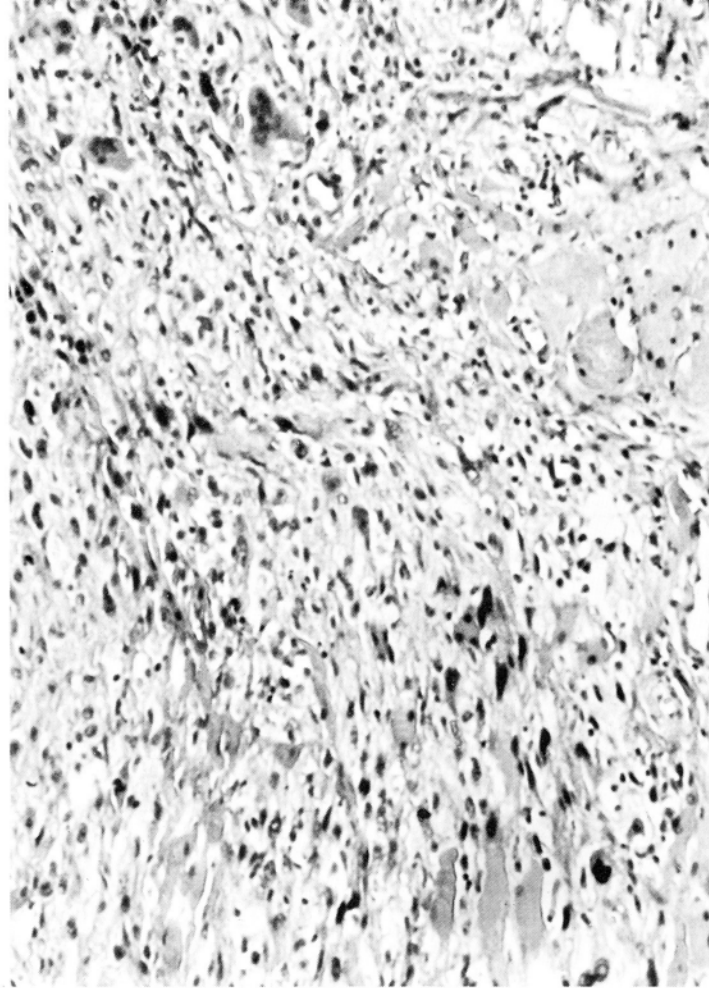


Fig. 2—Skeletal muscle fibers at the periphery of the lesion. The reaction of the injured fibers is to be noted. Multinucleated giant cells are present. Hematoxylin-eosin 180X.

calcified hematoma with the myositis ossificans. There is nothing here that goes along with an active bone infection: no stimulation of the periosteum, no osteolytic areas in the host bone, so I would not consider the diagnosis of osteomyelitis.

Dr. Regato: Drs. E. Salzman and M. Bischoff, of Denver, offered also an impression of parosteal osteosarcoma. Drs. J. W. Travis, of Topeka, and S. F. Crowley, of Colorado Springs, submitted myositis ossificans.

Operative findings: On October 7, 1966, an excision of the middle third of the left fibula was done; an **en-bloc** resection was attempted; however, there were finger-like extensions which were separately removed. The specimen measured 8.3 x 2.2 cm; the tumor itself measured 2.3 x 2 x 1.3 centimeters. On cross section the tumor did not seem to extend beyond the cortex and it had a pink-gray color.

Dr. Spjut: This is one of the most difficult cases in the Cancer Seminar. A differential diagnosis would include osteosarcoma, liposarcoma, myositis ossificans, traumatic periostitis or some other reactive lesion, or periosteal osteosarcoma. Throughout the sections, one finds numerous bizarre stromal cells along with abnormal mitoses. There is osteoid and bone formation. The small bits of cartilage and hemosiderin are present. The lesion appears to invade or at least involve skeletal muscle. There does not appear to be any particular attempt at maturation at what is presumably the periphery of this lesion. How does one put all of this together? The history of trauma which seems to be rather definite, might lead one to consider the possibility of some form of reactive lesion. However, one might ask the question, did the trauma merely call the attention of the patient to a pre-existing lesion? This commonly happens. We are

fortunate that we do not have this case for an immediate diagnosis. Even after having studied this slide almost daily for a month, it is difficult to arrive at a comfortable diagnosis. What can we rule out? Liposarcoma does not seem to be a reasonable diagnosis. Radiographically at least it does not appear to be an invasive lesion, but one that either arises from bone or periosteum. The few liposarcomas that have been reported as primary in bone, have been intraosseous lesions causing destruction of bone. We do know that cartilage and bone occasionally may be seen in liposarcoma, however, I think that this diagnosis would not stand up under careful scrutiny. To me, the main differential diagnosis lies between an osteosarcoma, perhaps periosteal in origin or a reactive lesion secondary to the trauma. The presence of hemosiderin does not help one way or another. The histological feature that we would like to see in order to rule in the possibility of a traumatically induced lesion, would be maturation at the periphery of this mass. Except for one area, the periphery does not appear to be maturing. In this one area, however, there is mature bone being formed with a slightly less cellular stroma between the bony trabeculae. Does this represent maturation of this lesion in the peripheral portion? Also at the periphery, if one studies the skeletal muscle fibers, one finds the many multinucleated giant cells apparently representing regenerative skeletal muscle fibers. Many of the bizarre stromal cells also appear to be arising from the skeletal muscle fibers. In myositis ossificans, arising secondary to trauma, one often sees bizarre osteoid and stroma which, on biopsy, may be impossible to distinguish from a sarcoma. Thus, even though we don't have a very good zoning effect described by Ackerman, I feel that the best diagnosis in this case is a reactive lesion secondary to the trauma. This is based on

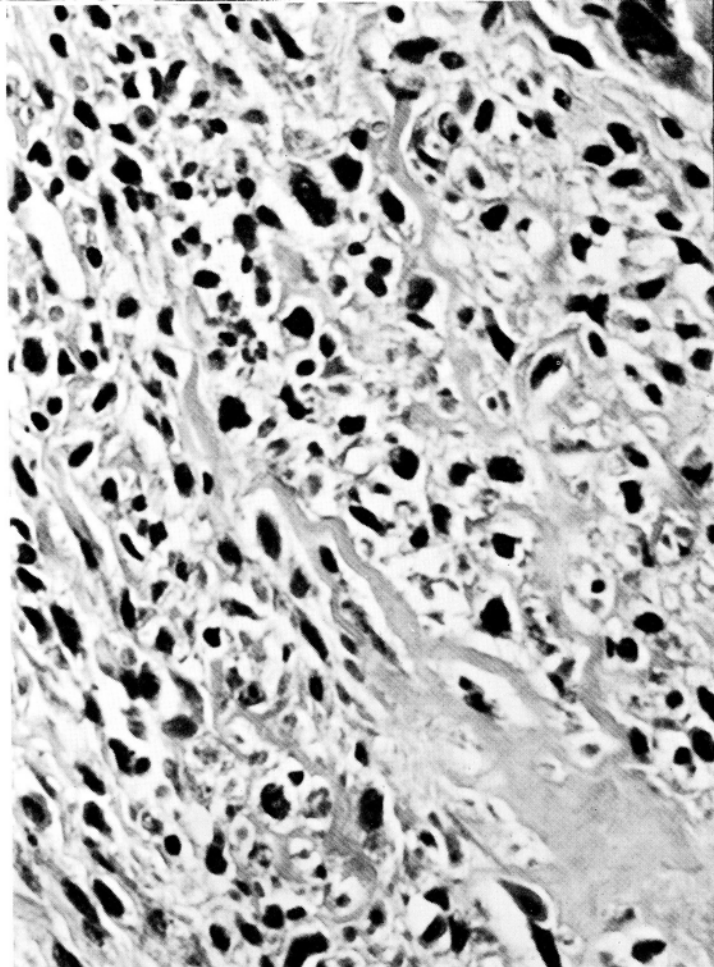


Fig. 3—The interior of the lesion presents cellular characteristics of malignancy. Hematoxylin-eosin 370X.

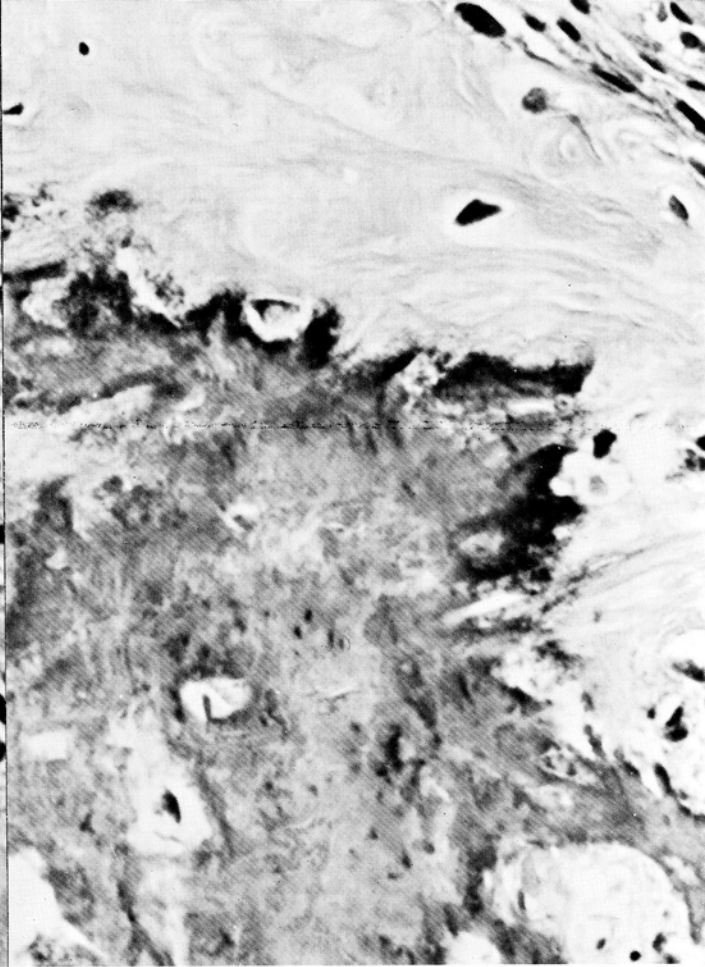


Fig. 4—Again at the periphery attempts at mature bone formation are noted. Hematoxylin-eosin 370X.

the rather skimpy histological evidence of mature bone formation towards the periphery of this lesion. Usually the periphery is more mature two months after trauma. I believe that we are helped a little by the radiographic findings in that I have failed to see any periosteal reaction immediately adjacent to this lesion, nor do I see evidence of cortical destruction. Osteosarcoma remains a distinct possibility in this case.

Dr. Spjut's diagnosis: Probable MYOSITIS OSSIFICANS

Histopathologic Diagnoses Submitted by Mail

Osteosarcoma	65
Myositis ossificans	38
Parosteal sarcoma	29
Fibrosarcoma	3
Others	4

Dr. Spjut: I have given my reasons for ending with a diagnosis of myositis ossificans although I still have a small reservation for osteosarcoma. Parosteal osteosarcoma is worthy of consideration but histologically we have a very undifferentiated lesion whereas with juxtacortical osteosarcoma one expects to see rather mature tissue and we have very little of that except for a few specks. Fibrosarcoma I think is out, because we do have osteoid formation as an integral part of this lesion.

Dr. Regato: Dr. L. Lowbeer, of Tulsa, and Dr. J. B. Frerichs, of El Paso, also submitted a diagnosis of myositis ossificans. Dr. G. Simon, of New Haven, preferred to designate it as a pseudo-malignant osseous tumor of the soft tissues. Dr. F. Schajowicz, of Buenos Aires, and Dr. M. H. McGavran, of Saint Louis, preferred osteosarcoma. Dr. M. R. Abell, of Ann Arbor, wavered between both of these diagnoses.

Subsequent history: In June, 1967, the patient was reported to be asymptomatic.

Dr. Hatcher: I first thought that this might be a marching fracture, but there is no discernible fracture line going through the bone and there is no fusiform reactive new bone around the entire circumference of the area. My next thought was subperiosteal osteoid osteoma: an osteoid osteoma does not necessarily have to have a very dense nidus and sometimes they may produce a reaction in the soft tissue similar to this. Frankly, I didn't think of an osteosarcoma from the roentgenogram. Whatever it is, I think it is very likely that the patient is cured and left with no disability.

M. Kepes, M.D., Kansas City, Mo.: I thought this was a difficult tumor. In the periphery there is some maturation and there is involvement of the muscles, an apparent reaction to injury; it does look like myositis ossificans in many respects; we have been pre-warned that myositis ossificans can look extremely immature but, somewhere, we have to draw the line between immaturity and malignancy. If there are many mitotic figures, that may mean young tissue, but once we get to malignant type of mitosis, atypical mitosis, this seems to be beyond the scope of just immaturity. I wonder if this is a case of myositis ossificans that turned into sarcoma. As Dr. Spjut pointed out, it should have been much more mature as far as peripheral ossification is concerned.

Dr. Spjut: Dr. Ackerman, in a paper that is so widely quoted, describes the presence of bizarre mitotic figures in these lesions; you could say that these don't mean very much since the patients have all done very well. If these were extrasosseous osteosarcomas, one would expect the majority of these patients to have died. As far as sarcomas

arising in myositis ossificans, this is a most difficult thing to prove; unless you had a biopsy prior to the malignant change, you don't really know what the pre-existing lesion was. These diagnoses are often made on the form that the lesion takes: we see a lesion in soft tissue near bone that radiographically looks like myositis ossificans and it turns out to be malignant and we draw the conclusion that perhaps this was a pre-existing myositis ossificans; this is very difficult to prove. Some of the patients that have been diagnosed as having a malignant lesion arising in myositis ossifications have done very well; I think that occasionally one runs into a generous pathologist and he may just over-diagnose this lesion.

A. O. Severance, M.D., San Antonio, Texas: I don't want to disappoint you. So far there are no metastases. Three weeks ago the patient was seen by his physician and he is still well with no evidence of residual tumor or recurrence. I would like to point out that the tumor arose apparently on the margin of bone and extended into the cortex only a very tiny bit microscopically. The medullary cavity was not involved. After some difficulty, we decided against myositis ossificans on the basis of the unusual cellularity and on what we interpreted as actual invasion of surrounding muscles by a malignant tumor, so we favored parosteal osteosarcoma.

A. M. Ginzler, Warren, Ohio: I wavered from myositis ossificans to osteogenic sarcoma. There is a fascinating book of lectures of Dr. James Ewing to the Junior class in Pathology at Cornell University in 1932, where he describes a case in which he had made the diagnosis of osteogenic

sarcoma in a high school football player and they were about ready to amputate the leg at Memorial Hospital of New York when a report came back from Dr. Frank Mallory of Boston of myositis ossificans.

M. R. Abell, M.D., Ann Arbor, Michigan: Our experience has been a little bit different. We've had a similar case which we thought was a classic myositis ossificans; one of the pathologists who has written on this subject visited us, saw the patient and said, "Son, don't let them take your leg off"; a year later he had pulmonary metastases. The suggestion that a malignant change may have arisen in myositis ossificans is not out of place, I think. We have great difficulty in deciding between these two lesions; we have to sit on the fence and leave it up to the clinician and up to the radiologist.

C. P. Schwinn, M.D., Los Angeles, California: I reviewed recently about 15 cases of myositis; we also have about 28 cases of parosteal sarcomas. I think the remark about the immaturity of the lesion is pertinent. I think the myositis ossificans sometimes appears very immature and very cellular and for this reason you may think it is a malignant tumor; but if you look at it carefully with the viewpoint that it is immature rather than malignant, you can resolve this difficulty. I thought these sections showed some malignant cells, particularly the fibroblastic ones at the periphery. I would consider this an osteosarcoma.

References

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9. Giant-cell Tumor of the Sphenoid

Contributed by **L. C. Griff, M.D.**, Harrisburg, Pennsylvania and **T. Zuck, M.D.** and **J. T. Decker, M.D.**, Denver, Colorado

THE PATIENT was a 13-year old girl in December, 1965, when she complained of frontal pains and diplopia of six weeks' duration. Examination revealed palsies of the 3rd and 6th left cranial nerves; visual fields, electroencephalograms and brain scans were all within normal limits.

Dr. Edeiken: The single lateral right roentgenogram of the skull reveals erosion of bone anterior to the clivus destroying the dorsum sella at its base. The upper portion of the dorsum is intact. One anterior clinoid is elevated and both show erosion. There is some increased bone density in the region of the sphenoid sinus. The arteriogram reveals anterior displacement of the carotid artery in the region of the sella.

Likely lesions in this situation are chordoma, aneurysm, carcinoma of the sphenoid sinus and glioma. Because the lesion extends from the posterior portion of the dorsum, and seems to have started in the posterior fossa near the clivus, the most likely possibility is chordoma. Occasionally, chondrosarcomas are found in this area, but they usually contain calcifications. Other lesions such as osteosarcoma and nasopharyngeal tumor extending into the skull may occur. Aneurysms do not usually occur at this site, but they could cause the same type bone destruction.

Dr. Edeiken's impression: CHORDOMA.

Roentgenologic Impressions Submitted by Mail

Chordoma	33
Craniopharyngioma	22
Eosinophilic granuloma	12
Others	29

Dr. Edeiken: A craniopharyngioma can occur anywhere in the sella, above the sella and it can occur in this age group; it is a likely diagnosis I suppose but you should see some calcification within it. There is no reason why this can't be histiocytosis X; I think that is a very good thought.

Dr. Regato: Dr. B. L. Pear, of Denver, also suggested chordoma. Dr. M. Bischoff, of Denver, preferred craniopharyngioma, and Dr. F. Wilson, of Colorado Springs, eosinophilic granuloma.

Operative findings: On February 3, 1966, a fronto-temporal craniotomy was done. The specimen removed consisted of a small fragment of bone and soft tissue. On March 8, 1966, she was re-operated upon; several lobules of the tumor were removed from the area of the cavernous sinus with brisk bleeding; eventually, the surgeons gave up the tumor as "inoperable".

Dr. Spjut: From a rather small bit of tissue, we can make a diagnosis of a giant cell lesion. Is this a giant cell tumor, a giant cell reaction, or some type of central nervous system tumor with numerous giant cells? In consulting with our neuropathologist and neuroradiologist, the possibility of a central nervous system tumor seems unlikely; the radiographs seem to bear this out. We have no clinical evidence of hyperparathyroidism. It is possible that this is a giant cell reaction to some other lesion, say of the bone, but from the biopsy one would have to say that the multinucleated giant cells plus the stroma are compatible with the giant cell tumor, even though it had been stated previously that a patient of this age would rarely be affected by giant cell tumor. We are all aware that giant cell tumors are found

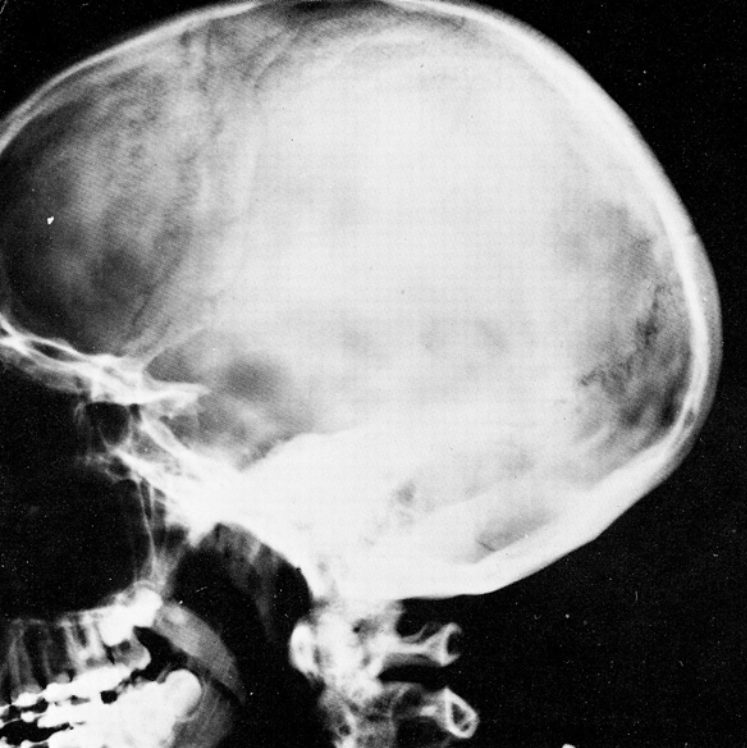


Fig. 1—Erosion of the dorsum sellae at its base.

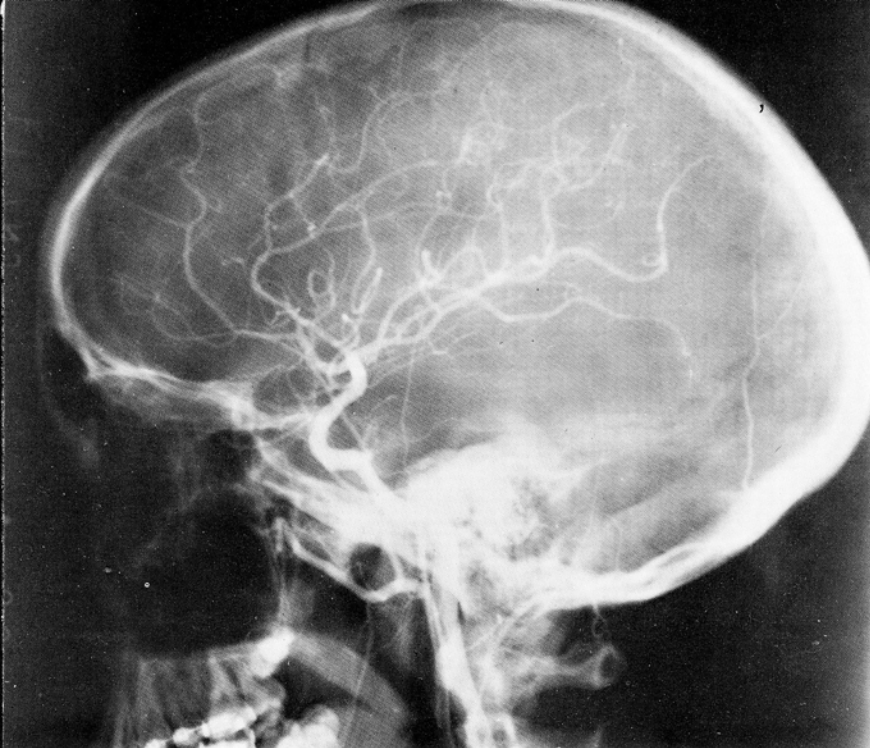


Fig. 2—Anterior displacement of the carotid artery in the region of the sella.

more commonly in the long bones or many other bones than the skull. The few giant cell tumors that have been reported in the jaw and skull generally have been some other giant cell lesion, for example the giant cell reparative granuloma. The latter lesion is seen most frequently in the mandible or maxilla. Histologically, the multinucleated cells are more sparse than one observes in a giant cell tumor, but this may be difficult to judge in a given lesion. The exact classification of this case is difficult. I know of no histological stains that would help us to differentiate the reactive lesion from the genuine tumor. It has been described that the multinucleated cells of the giant cell tumor contain an abundance of acid phosphatase, but even this is not specific as the giant cells of giant cell reparative granuloma are also acid phosphatase positive. From the literature a few acceptable giant cell tumors have been reported that involve the bones of the skull. Among the cases that I have been able to find have been individual case reports of giant cell tumor of sphenoid bone (girl age 12) and of the temporal bone (woman age 50).

Dr. Spjut's diagnosis: GIANT-CELL TUMOR.

Histopathologic Diagnoses Submitted by Mail

Giant-cell tumor, unclassified	79
Giant-cell tumor, benign	26
Giant-cell tumor, malignant	7
Eosinophilic granuloma	6
Hyperparathyroidism	5
Others	18

Dr. Spjut: We've taken the stand that one cannot necessarily predict from a histological section whether a giant cell tumor is benign or malignant. It is true that the majority of the giant cell tumors fall into a category the behavior of which suggests that they are a benign lesion. Then there are the borderline lesions in which the stroma appears more active, mitotic figures are seen and the nuclei of the giant cells also undergo alterations that are of some concern. The out and out malignant giant cell tumors are comparable to an osteosarcoma; so, instead of saying a giant cell tumor is benign or malignant when we are talking about the genuine article, we merely say this is a giant cell tumor and occa-

sionally go along with Lichtenstein's classification, saying grade I or grade II, or grade III, with III being the obviously malignant one. This may very well be a giant cell tumor but one would have to say that we don't have features that say this is an out and out malignant tumor. We don't have any histological features here, other than the multinucleated giant cells, that would go along with an eosinophilic granuloma. We don't have the histologic background that we would like to see. Hyperparathyroidism is a lesion that histologically could be indistinguishable from a giant cell tumor. But I think we have to consider the clinical findings plus the radiographic findings and this would help us to rule out the possibility of hyperparathyroidism.

Dr. Regato: Dr. D. Dawson, of Colorado Springs, and Dr. Claire Langston, of Denver, also submitted a diagnosis of giant-cell tumor. Dr. M. R. Abell, of Ann Arbor, preferred giant-cell reaction. Dr. C. P. Schwinn, of Los Angeles, offered Brown tumor of hyperthyroidism, a diagnosis which was also suggested by Dr. L. Lowbeer, of Tulsa.

Subsequent history: This specimen was submitted to the AFIP (Accession No. 1195768). A diagnosis of giant-cell tumor was rendered (J. C. Gallagher, M.C.). The distinguished pathologists of that renowned and trusted center went further to advise, without regard to the location of the tumor (!) that "complete surgical removal is the only safe therapeutic procedure; radiation therapy should be avoided; irradiation of this lesion may convert it into a sarcoma".

J. L. Eller, M.D., Aurora, Colo.: In spite of the AFIP recommendations, this child was treated upon advice of our consultant therapeutic radiologist: she was irradiated through two lateral fields, 7 x 6 cm in diameter and received a mid-line cranial dose of 5000 R in 45 days. The irradiation was followed by epilation and marked subjective improvement. The present radiograph of the skull shows bone regrowth and reformation of the destructed area.

Dr. Hatcher: It seems to me that the warning from the AFIP is probably sound for the average giant cell tumor at the distal end of the femur but here is a giant cell tumor that, if it continues to grow, is going to cause lots of trouble and it seems that it is not feasible to remove that tumor

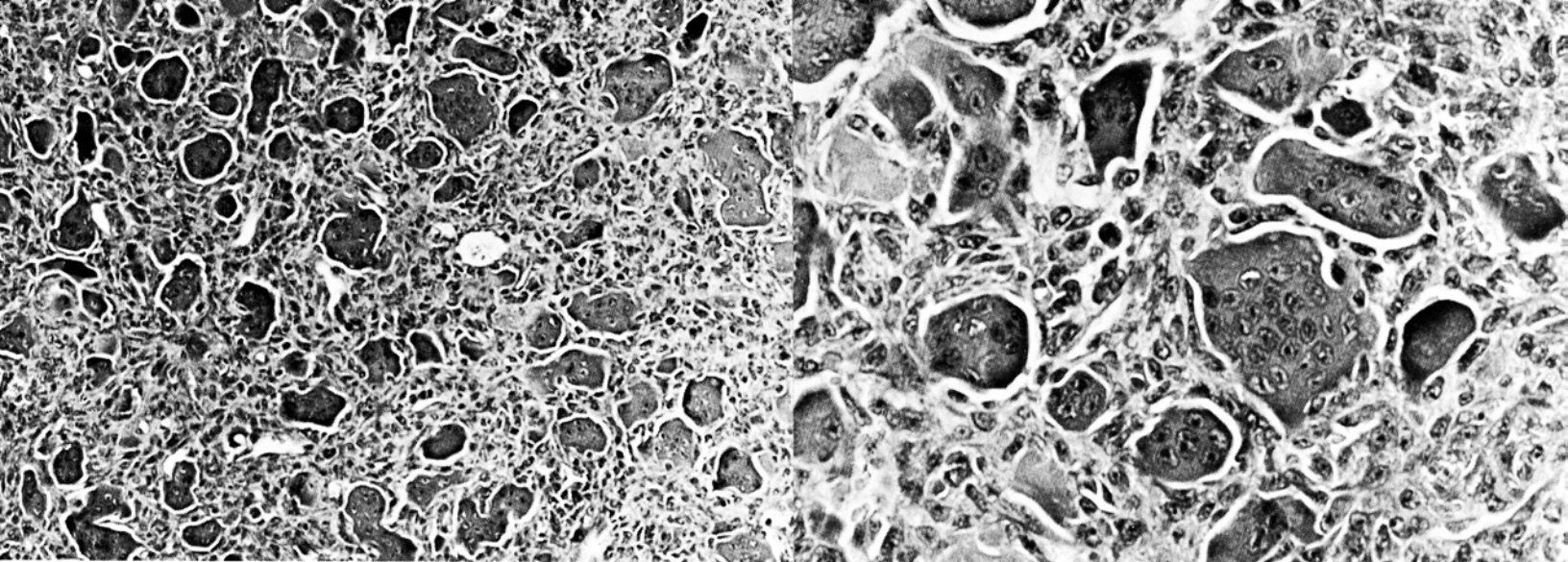


Fig. 3—The numerous multinucleated giant cells are apparent. Hematoxylin-eosin 100X.

Fig. 4—With higher magnification the giant cells and the stroma seem to be well differentiated. Hematoxylin-eosin 300X.

entirely. Therefore, I think that radiation is the one form of treatment that may be of benefit. The evidence that some giant cell tumors may be converted to malignant tumor by irradiation is still open to question.

Dr. Regato: Anyone who would say that a benign giant cell tumor becomes malignant because of irradiation will also have to explain how there are cases of giant cell tumors that were only curetted and later were said also to have become malignant. It seems evident to me that such tumors were malignant in the first place and that they were misdiagnosed as benign; neither the curetting nor the radiotherapy had anything to do with the subsequent manifestations of malignancy. In a patient like this such concept denies a curative form of treatment to a child whose lesion is entirely incurable by surgery. The dogmatic manner in which the report was given led first to abstention. I was consulted at Fitzsimmons Army Hospital and I advised that radiotherapy be given; I did not know until this moment that my advice had been followed.

M. R. Abell, Ann Arbor, Michigan: We were reluctant to use the term giant-cell tumor unless the lesion fits clinically in respect to the age and unless it is in the usual location. Seeing only a small amount of tissue we preferred to designate it as a benign giant cell reaction.

L. Lowbeer, M.D., Tulsa, Oklahoma: Is there any information available on blood calcium and blood phosphorus?

Dr. Regato: I was prepared for that question; we called Fitzsimmons but the chemistries are not available.

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10. Juxta-cortical Chondroma of the Finger

Contributed by **R. L. Homer, M.D., R. P. Spurk, M.D. and L. E. Olsen, M.D.,**
 Denver, Colorado

THE PATIENT was a 70-year old woman in November, 1966, when she gave a history of progressive enlargement of the left index finger for fifteen years; in recent years most tenderness and limitation of motion of the finger had resulted. On examination there was a diffuse enlargement of the proximal phalanx of the left index finger with 30° flexion contracture of the middle joint.

Dr. Edeiken: The roentgenogram reveals a soft tissue mass anterior to the proximal phalanx of the left index finger. It has been present for fifteen years. There are flocculent calcifications within this mass and involvement of the adjacent bone by erosion.

Tumors to be considered in this region are giant-cell tumors of tendon sheath, periosteal chondroma, chondroblastoma, chondrosarcoma, and synovial sarcoma. The presence of the flocculent calcification indicates cartilaginous tumor and since the soft tissue mass extends peripherally to this and there is erosion of bone, one must assume that this is a cartilaginous tumor that has undergone malignant change. Giant-cell tumor, tendon sheath, and fibrosarcomas

will never show a flocculent calcification in the roentgenograms. Synovial sarcomas frequently reveal calcifications which may be rather dense. One would expect synovial sarcoma to contain calcifications throughout the soft tissue tumor.

Dr. Edeiken's impression: CHONDROSARCOMA secondary to periosteal chondroma.

Roentgenologic Impressions Submitted by Mail

Chondroma	25
Chondrosarcoma	18
Giant-cell tumor	18
Fibrosarcoma	15
Others	20

Dr. Edeiken: The giant-cell tumor will not show calcification out into the stroma; it is very unusual for it to do that, although once in awhile you have a little bleeding area and you will get some calcification. The soft tissue mass of fibrosarcoma should not show calcification. This doesn't mean that calcification or ossification or even periosteal proliferation doesn't occur, but these tumors seem to have a



Fig. 1—Diffuse enlargement of the proximal phalanx of the left index finger.

preference for destroying either bone or periosteum as soon as it's met. A good general rule, however, giant-cell tumors and fibrosarcomas should not contain calcium.

Dr. Regato: Dr. L. O. Martinez, of Miami, also offered an impression of chondrosarcoma. Dr. R. Henschele, of Saginaw, preferred chondroma.

Operative findings: On November 17, 1966, a firm mass was found arising from the volar surface and bulging on both sides of the flexor tendons of the left index finger. A frozen biopsy was done, followed by a ray amputation of the finger through the proximal portion of the second metacarpal bone.

Dr. Spjut: This tumor is of cartilaginous origin and presents a differential diagnosis between a benign cartilaginous tumor or a chondrosarcoma. The long history in this case is not particularly helpful as a chondrosarcoma may well have been present for years prior to a confirmed diagnosis. A few chondrosarcomas arising in the small bones of the hands and the feet have been reported, so this is a valid consideration in this lesion. Radiographically, this lesion appears to have arisen outside the bone and is not central in origin. Taking the radiological findings together with the histological findings, one could classify this as a juxtacortical chondroma. How do we know this is not a well-differentiated chondrosarcoma? Most of the cells in this lesion are poorly preserved or perhaps dead. In areas of viable cartilage the cells appear essentially normal, but occasionally, one encounters binucleated cartilage cells. It has often been stated that a histological criterion for the diagnosis of well-differentiated chondrosarcoma is the finding of binucleated cartilage cells. However, in instances of cartilaginous tumor arising outside bone, for example synovial chondromatosis or juxtacortical chondroma, there are often atypical nuclear features that one might associate with a well-differentiated chondrosarcoma. In other words, a few atypical cells are allowable in the diagnosis of lesions in this location. This case fits very well with the lesion that Jaffe designated as the juxtacortical chondroma. In fact, one of the illustrations in his paper describing this lesion is similar



Fig. 2—Soft tissue mass anterior to the proximal phalanx of the index finger with erosion of adjacent bone.

to the case at hand. Despite the few binucleate cells, this is a benign lesion. It is important in the diagnosis of cartilaginous tumors to know the size and location of the lesion, as all criteria are not applicable to all lesions.

Dr. Spjut's diagnosis: JUXTA-CORTICAL CHONDROMA.

Histopathologic Diagnoses Submitted by Mail

Chondrosarcoma	53
Enchondroma	35
Osteochondroma	30
Juxta-cortical chondroma	12
Others	9

Dr. Spjut: I have already given you my reasons for saying this is not chondrosarcoma. The films would indicate that this is not an enchondroma. We have a lesion that is basically a cartilaginous tumor and it does not have the gross morphological features of an osteochondroma: we don't have a very good combination of bone and cartilage.

Dr. Regato: Dr. F. Schajowicz, of Buenos Aires, Dr. A. M. Ginzler, of Warren, Ohio, and Drs. Magda and John Kepes, also made a diagnosis of juxta-cortical chondroma. Dr. C. P. Schwinn, of Los Angeles, preferred the designation of enchondroma (periosteal chondroma). Dr. M. R. Abell, of Ann Arbor, suggested synovial osteochondromatosis. Dr. M. H. McGavran, of Saint Louis, and Dr. R. Hall, of Fort Sam Houston, preferred chondrosarcoma.

Subsequent history: The patient was last seen in September, 1967. There was no evidence of recurrence.

Dr. Hatcher: Most tumors of the phalanges of the hand or foot are cartilaginous, and most of them are benign, although once in awhile there is a frank chondrosarcoma primary in those small bones. One exception to this is the Maffucci syndrome — a combination of enchondromatosis plus hemangiomas. These cartilaginous tumors quite frequently become malignant but unfortunately for the chondrosarcoma part of it, most patients die of carcinoma elsewhere in the body, before the chondrosarcoma catches up with them. If the same histologic appearance of these benign cartilaginous tumors is seen centrally in a major bone, one

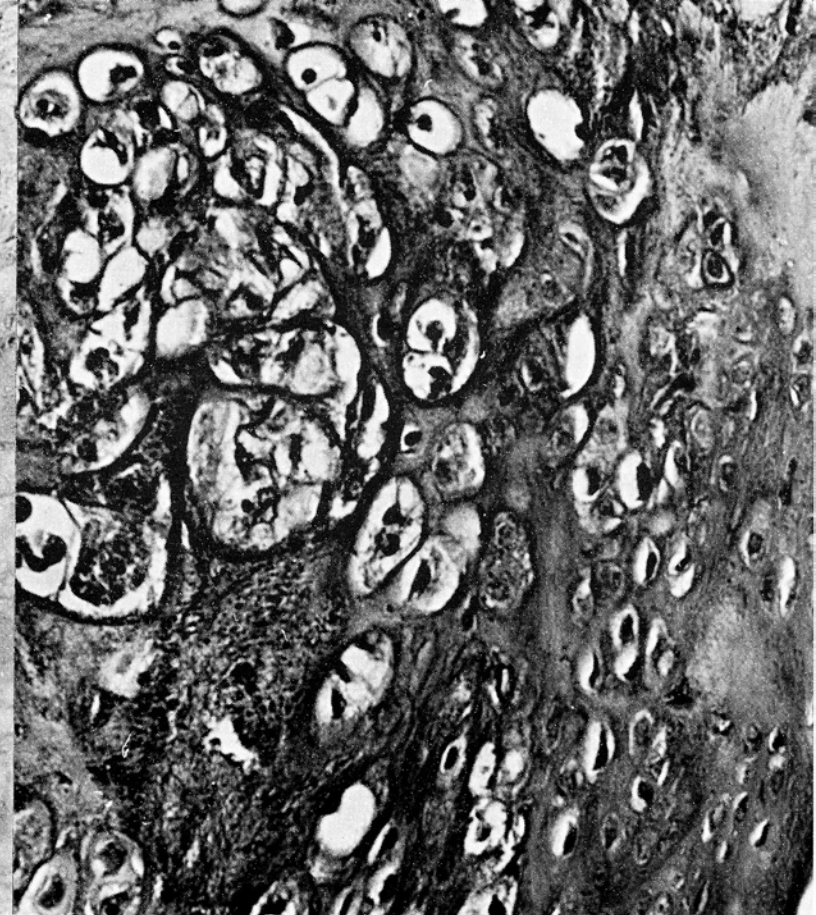
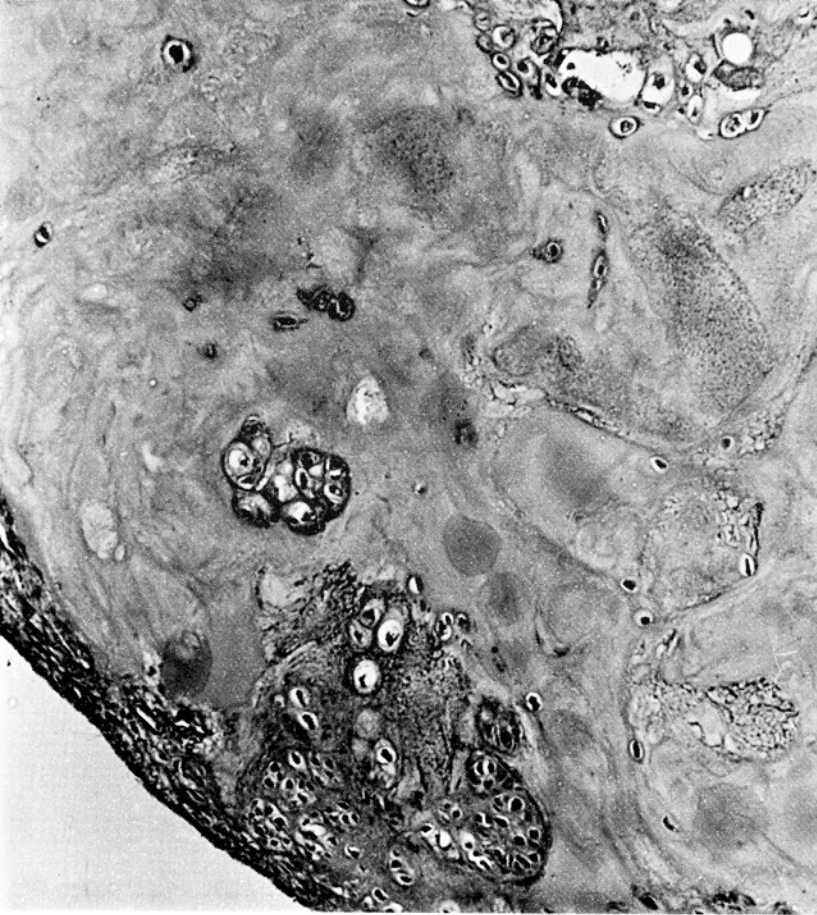


Fig. 3—Hyaline cartilage that appears nearly normal. Hematoxylin-eosin 100X.

Fig. 4—Under higher magnification occasional binucleate cells are noted. Considering the entire case these are not to be misconstrued as evidence of malignancy. Hematoxylin-eosin 300X.

would think more of a sarcoma and treat it much more radically. This woman is 70 years of age and her finger apparently almost useless: the ray-like amputation gives her a better looking hand.

M. R. Abell, Ann Arbor, Michigan: I would like to ask Dr. Spjut how he would differentiate this, histologically, from synovial chondromatosis. Is there a difference or do you have to rely solely on what the surgeon finds on the basis of minimal amount of cortical thickening of bone? Synovial chondromatosis can give you some pretty large lesions particularly in the hip joint and cause some deformity of the adjacent bone.

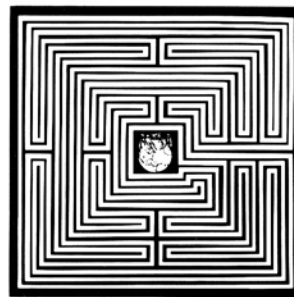
Dr. Spjut: In making that distinction one would rely a great deal on what the surgical findings were; if the surgeon said that this was confined to a tendon sheath or was confined to a joint then I would go along with synovial chondromatosis. Histologically, if you take fields out of this

lesion, it would be hard to say it isn't. I don't know whether a synovial chondromatosis of the finger would cause this much reaction along the cortex of the phalanx.

Dr. Edeiken: I have never seen synovial chondromatosis or synovial osteochondroma outside the joints. First of all, there has to be synovia — and I appreciate there is synovial tissue around the tendons—but I have never seen it; perhaps those are the extra-osseous chondromas that we're told about. But to make that diagnosis at the present stage, it must be within a joint and it is usually within a large joint.

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11. Solitary Chondroblastic Sarcoma of Metacarpal Region

Contributed by H. G. Kroll, M.D., J. W. Travis, M.D. and W. W. Scammon, M.D.
Topeka, Kansas

THE PATIENT was a 45-year old man in September, 1965, when he sustained trauma on the metacarpal region of the left hand; there was a history of pre-existing swelling of six months' duration in this area. Examination revealed a tender and diffuse tumefaction of the dorsum of the left hand and thumb.

Dr. Edeiken: The roentgenograms reveal a soft tissue mass with erosion at the base of the second metacarpal. There is no calcification within this lesion. The erosion of the base of the second metacarpal indicates that this is a malignant lesion.

The differential diagnosis should include fibrosarcoma, synovial sarcoma, and metastatic disease. There is nothing distinctive about this lesion to lead one to the proper diagnosis. Synovial sarcomas will frequently show calcifications but the majority do not. Fibrosarcomas never calcify. In a 45-year old man metastatic lesions must be considered, but without a history of a primary tumor, one could hardly make this diagnosis.

Dr. Edeiken's impression: FIBROSARCOMA.

Roentgenologic Impressions Submitted by Mail

Metastatic carcinoma	29
Enchondroma	12
Giant-cell tumor	10
Fibrosarcoma	9
Chondrosarcoma	7
Others	27

Dr. Edeiken: Metastatic carcinoma to the soft tissues of the hand is a fairly rare thing; you wouldn't expect three separate bones to be involved in the same locale without bones in other areas being involved. This is not an enchondroma: it could be a periosteal chondroma, but there is no

calcification and I have never seen one grow this large outside of the bone. This could be a giant-cell tumor of the tendon sheath but not a giant-cell tumor of the bone. This could be a chondrosarcoma the same as it could be a fibrosarcoma; there is no calcification in it and there is no way to make the roentgen diagnosis.

Dr. Regato: Dr. L. O. Martinez, of Miami, also submitted a diagnosis of fibrosarcoma. Dr. J. C. Lemon, of Denver, suggested metastatic carcinoma. Dr. R. Henschel, of Saginaw, preferred chondroma; Dr. J. Cox, of Colorado Springs, chondrosarcoma.

Operative findings: On September 9, 1965, a surgical exploration was done; a sequestrum was removed from what appeared to be a granulomatous lesion. A diagnosis of chondrosarcoma was rendered and, in December, 1965, an amputation of the left index finger with the second metacarpal bone was done.

Dr. Spjut: This is an interesting case from the standpoint of the histological differential diagnosis. Numerous possibilities present themselves, at least to the person who has never seen a tumor like this before, and that includes me. The differential diagnostic possibilities that I considered were sweat gland carcinoma, liposarcoma, glomus tumor, rhabdomyosarcoma, and cartilaginous tumors of bone. Which can we eliminate? The glomus tumor seems to be one of the first to be eliminated in that we do not have a vascular lesion with the cells surrounding the vessels, although a glomus tumor may have the mucoid and myxoid stroma seen in this lesion. One wonders about skeletal muscle tumors because of the dense acidophilia of the cells, but pleomorphism is lacking and cross-striations are not demonstrable. Could this be a myxoid liposarcoma that is secondarily involving the bone? Roentgenographically, this seems to be a

Fig. 1—Lateral and front view showing soft tissue mass with erosion of the second metacarpal without calcification.

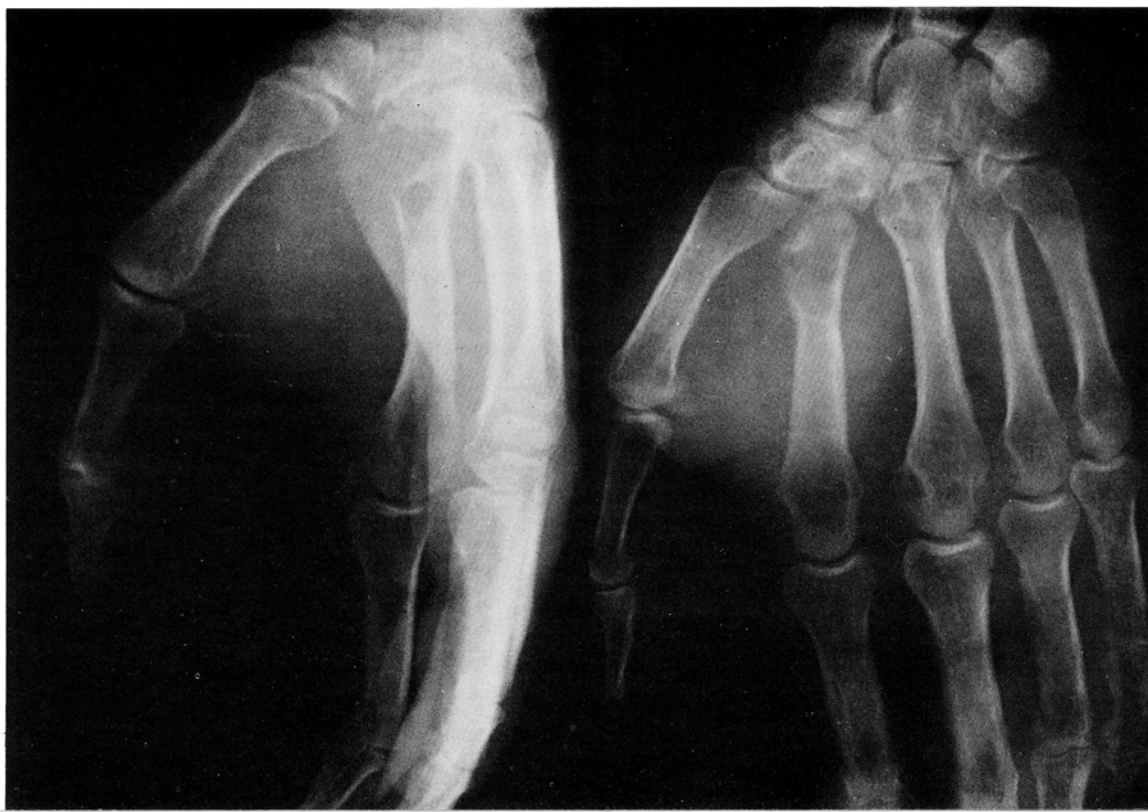
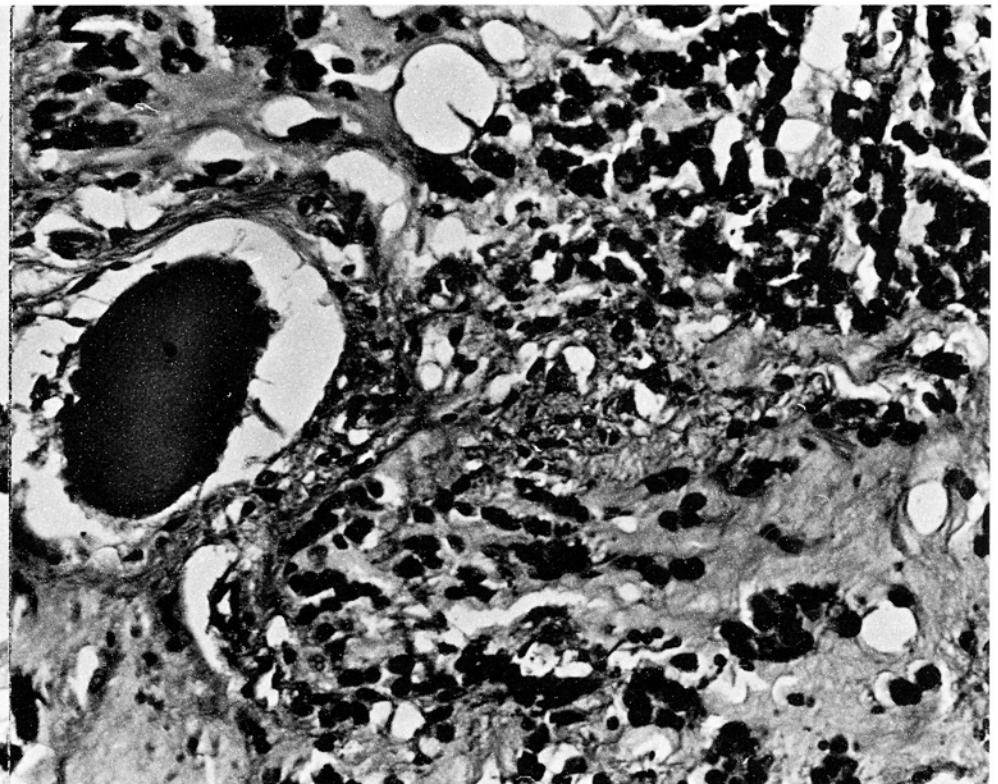
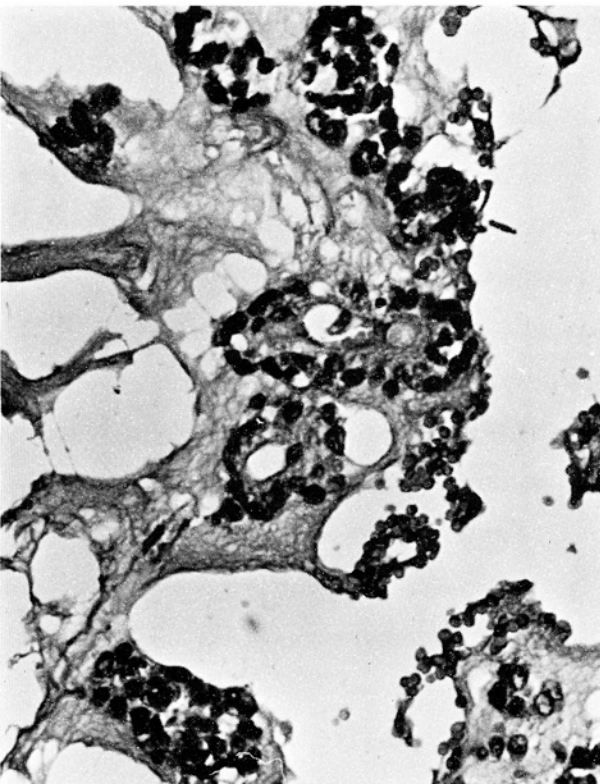




Fig. 2—Surgical specimen showing tumor removed with index finger and metacarpal bone.

lesion that is coming out of the bone and invading soft tissues. In addition, the cytologic pattern of this tumor does not appear to be characteristic of liposarcoma; the cells are only occasionally vacuolated and signet ring cells and identi-

Fig. 3—The mucoid background with aggregates of small round cells is noted. Hematoxylin-eosin 300X.



fiable fat cells are not seen. A tumor of sweat gland origin certainly seems possible. Many of the sweat gland carcinomas, however, are described as being clear cells, whereas this lesion has cells which are densely acidophilic. We have seen sweat gland tumors composed of cells as acidophilic as those in this case. Also, a sweat gland tumor may have a mucoid and myxoid background and a cartilaginous component.

Could this be a primary tumor of bone? If so, one of the major considerations would be a tumor of cartilaginous origin. In the section that I had, I was unable to demonstrate a cartilage, but the histological pattern of this tumor would fit with a tumor that has been described by Lichtenstein and designated as a chondroblastic sarcoma. This tumor may have a myxoid background, and the cells are well-delineated with a dense acidophilic cytoplasm and dark ovoid or round nuclei as noted in this case. The cells often line up in strands. The prognosis of the lesions is uncertain as only a few cases have been reported but the suggestion by Lichtenstein has been that it is good. Thus the diagnosis in this case is solitary chondroblastic sarcoma.

Dr. Spjut's diagnosis: SOLITARY CHONDROBLASTIC SARCOMA.

Histopathologic Diagnoses Submitted by Mail

Chondrosarcoma	45
Metastatic carcinoma	35
Glomus tumor	14
Chondro-myxoid fibroma	14
Others	31

Dr. Spjut: I believe we could say that this tumor would fit into the broad category of chondrosarcomas, although a most unusual variant. One might have to consider metastatic carcinoma of the breast, maybe even of the lung, but we have this unusual mucoid pattern to contend with. Glomus tumor, I've already discussed. Chondromyxoid fibroma is a reasonable consideration; all I can say is that this one does not present the characteristic features that would allow us to make that diagnosis. The characteristic features are those of lobulation of the tumor not only grossly but histologically with a migration of the nuclei of the lobules toward the periphery; there are some atypical chondromyxoid fibromas that have been described, of course.

Fig. 4—The cells have dense cytoplasm and are compatible with immature cartilage cells. A cross-sectioned skeletal muscle fiber is isolated by the tumor. Hematoxylin-eosin 300X.

Dr. Regato: Dr. H. L. McGaffey, of Idaho Falls, and Dr. R. M. Sherwin, of Colorado Springs, offered a diagnosis of mixed tumor. Dr. M. H. McGavran, of Saint Louis, and Dr. C. Langston, of Denver, preferred metastatic carcinoma, of the lung or stomach, probably and respectively. Dr. C. P. Schwinn, of Los Angeles, suggested chordoid tumor and Dr. F. Schajowicz, of Buenos Aires, an adenoid basal-cell epithelioma of the sweat glands. This patient's slide had been seen by Drs. M. B. Dockerty and D. C. Dahlin, of the Mayo Clinic, who rendered a diagnosis of myxoid chondrosarcoma.

Subsequent history: In September, 1967, there was no evidence of recurrence or metastases.

Dr. Hatcher: This tumor appears to be a relatively benign thing provided it is removed; the very site itself favors a cartilaginous origin.

L. Lowbeer, M.D., Tulsa, Oklahoma: Dr. Spjut, in the paper to which you were referring to, by Lichenstein, two or three of the cases which he calls chondroblastic sarcomas were multicentric. I don't know whether a skeletal survey exists in this particular case.

Dr. Spjut: That is right and, in addition, the lesion is called mesenchymal chondrosarcoma; it is also described as multicentric, but we don't have any evidence of other lesions in this patient.

J. W. Travis, M.D., Topeka, Kansas: Skeletal surveys were negative in this patient.

C. P. Schwinn, M.D., Los Angeles, California: This term chordoid was coined by Dr. Stewart; he jokingly postulated that they arose from the sesamoid bone but he was not serious about that. He called them chordoid because they resembled chordomas; they also look like the so-called spindle cell chondrosarcoma that you see occasionally in the long bones; they have been reported as soft part chondrosarcomas, on occasion. Of the five we have, three have metastases or are dead.

W. J. Frable, M.D., Richmond, Virginia: We had a case similar to this when I was a resident; it was in the foot, and we didn't know what it was at the time. We sent it to Dr. Stout who had seen two such cases, both of which had metastasized. He felt that they were related to embryonal cartilage and used the term embryonal chondrosarcoma; ours also metastasized very widely.

W. L. Miller, M.D., El Paso, Texas: Dr. Spjut, I think it might have been interesting to have stained this for acid or sulfate mucosaccharide substance. I wonder if you did an alcine blue stain at low pH or did any particular mucin stains on this lesion?

Dr. Spjut: We did mucin stains which were negative. I didn't alter the pH on the alcine blue. I could try that when I get back.

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12. Chondroblastoma of the Talus

Contributed by **J. W. Doucette, M.D.** and **A. C. Sudan, M.D.**, Denver, Colorado

THE PATIENT was a 12-year old girl in June, 1967, when she was examined because of a swelling of the right ankle following trauma to her toe while swinging. A small, apparently cystic lesion of the right talus had been noticed one year previously.

Dr. Edeiken: This is almost an impossible case to evaluate on a roentgenographic basis. The patient did present this lesion one year previously, but we do not have the roentgenograms; it must not have been considered a significant lesion because no biopsy was obtained. Since it followed trauma, one might consider aneurysmal bone cyst and osteoblastoma. One year later, apparently it was operated and we have a post-operative film. There is an irregularity of the inferior margin of the calcaneus with some evidence of increased density. This area of increased density suggests that there is an actively growing tumor rather than a benign cyst. The possibility of osteoblastoma seems good. Unicameral bone cyst, eosinophilic granuloma, chondroblastoma and infection are other possibilities.

Dr. Edeiken's impression: OSTEOLASTOMA.

Roentgenologic Impressions Submitted by Mail

Bone cyst	20
Chondroblastoma	15
Osteoblastoma	14
Chondro-myxoid fibroma	9
Non-osteogenic fibroma	7
Others	28

Dr. Edeiken: Bone cyst is a good thought — chondroblastoma and osteoblastoma, I'll just put together and they're good thoughts too. I didn't mention chondromyxofibroma; I see none of their characteristics: usually they have a very

thick bone reaction around them and they show big septae. Non-osteogenic fibromas are rare in this area.

Dr. Regato: Dr. J. F. Wilson and Dr. C. H. Taggart, of Colorado Springs, diagnosed an aneurysmal bone cyst. Dr. E. Salzman, of Denver, offered osteoblastoma and Dr. J. C. Lemon, also of Denver, chondroblastoma.

Operative findings: On July 6, 1967, the lesion of the medial aspect of the right talus was curetted; bone chips from the iliac crest were impacted in the defect. The material removed was spongy but gritty and brown-red in color.

Dr. Spjut: The differential diagnosis in this lesion revolves around giant cell tumor, a malignant cartilaginous tumor or a chondroblastoma. Again, we are dealing with a patient who is 12 years old; this makes the diagnosis of a giant-cell tumor a little more difficult to confirm. Histologically, the lesion does not have the features that are entirely characteristic of giant-cell tumor although there are numerous multinucleated cells. If one pays close attention to the intervening cells, we see that they are rounded and fairly well demarcated, rather than the more spindly stromal cell seen in the giant-cell tumor. Scattered foci of calcification are present. These may be seen in occasional giant-cell tumors. What about chondrosarcomas? It is a distinctly uncommon lesion in this age group. From the histological standpoint, even though this is a cellular lesion, there is a uniformity to the cell type involved. We do not have the bizarre nuclei that one might expect with a chondrosarcoma of this degree of cellularity. This lesion is quite characteristic of a benign chondroblastoma: the very well outlined rounded cells that form the background stroma, the multinucleated giant cells that are not atypical and the foci of calcification,



Fig. 1—Irregularity of the inferior margin of the calcaneus with some increased density.

fit well with this diagnosis. The most common locations of this lesion have been in the epiphysis of long bones, although as more cases are found, more unusual sites are reported; other cases having involvement of the talus have been recorded. From our electron microscope studies, we have been able to demonstrate that the cells in the lesion are similar to the normal chondroblast: this lesion appears to have been aptly named. For all practical purposes, this is a benign lesion and should be treated as such. There have been rare cases reported of acceptable malignant chondroblastoma. Local recurrence after curettage is not unusual.

Dr. Spjut's diagnosis: CHONDROBLASTOMA.

Histopathologic Diagnoses Submitted by Mail

Giant-cell tumor	43
Malignant giant-cell tumor	9
Chondroblastoma	37
Aneurysmal bone cyst	12
Hyperparathyroidism	9
Others	30

Dr. Spjut: If we don't have a giant-cell tumor, we obviously can't have a malignant giant-cell tumor. We have no features that I know of to support the diagnosis of aneurysmal bone cyst. I don't see how we could support hyperparathyroidism; in the pediatric age group it is an extremely rare lesion and I don't know how we should be making that diagnosis without some laboratory confirmation.

Dr. Regato: Dr. T. H. McConnell, of Dallas, and Dr. R. Schultz, of Sioux Falls, also made a diagnosis of chondroblastoma. Dr. M. R. Abell, of Ann Arbor, suggested a giant-cell reaction but wondered about histiocytosis X. Dr. L. Lowbeer, of Tulsa, interpreted the lesion to be a cross of chondromyxoid fibroma and chondroblastoma, also known as enchondromatous giant-cell tumor.

Subsequent history: A slide of this case was submitted to Dr. Henry L. Jaffe by the contributors; he wrote: "I have considered and ruled out a giant-cell tumor and a benign vascular tumor. It is my impression that the lesion is actually a benign chondroblastoma, though somewhat atypical in its histologic composition."

A slide was also submitted to the AFIP (Accession No.

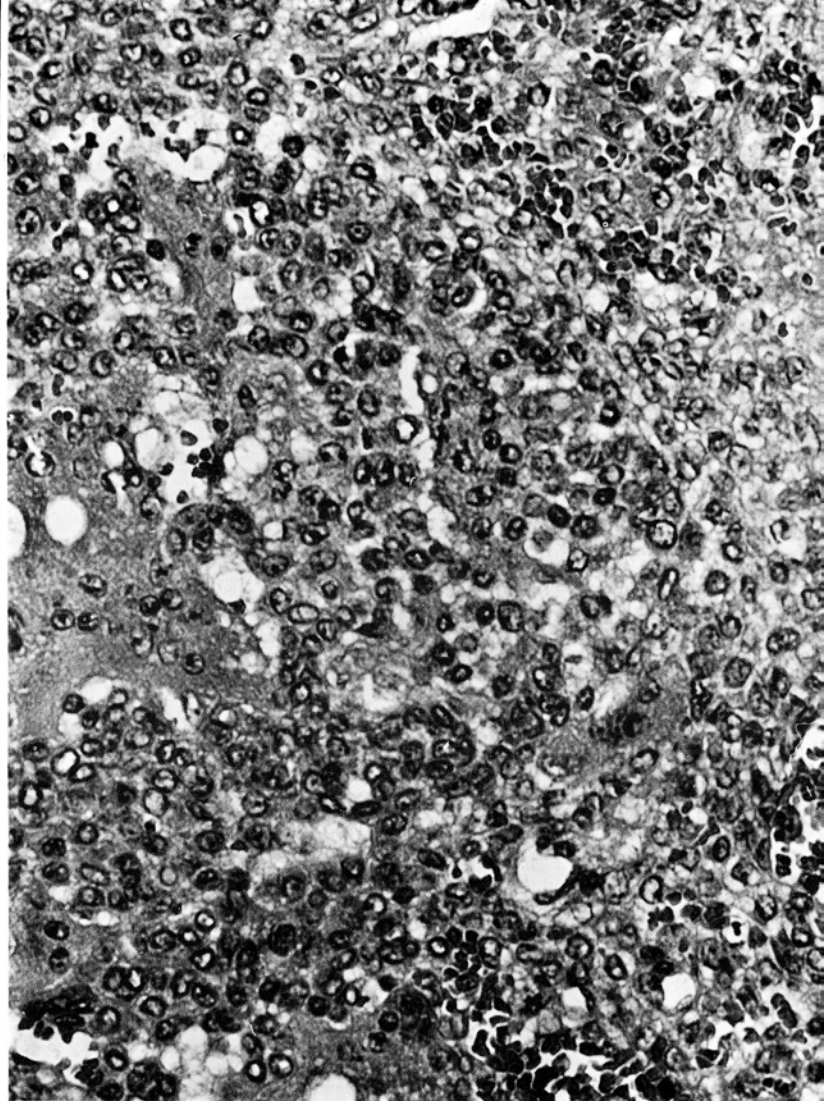


Fig. 2—Uniformity of the cells in a chondroblastoma. Multinucleated giant cells are present. These are found commonly in this lesion. Hematoxylin-eosin 300X.

1249744) and a report was rendered by Dr. Lent Johnson: "The choice would lie largely between a chondroblastoma and a giant-cell tumor. By convention, chondroblastoma is the modern term for the old epiphyseal chondromatous giant-cell tumor of Codman and to date virtually all such lesions have been regarded as being epiphyseal in location. The talus has no epiphysis. Therefore, unless the evidence were overwhelming, one would hesitate to use this term. Furthermore, there is no real good evidence of chondroblastic activity. In some occasions hyalinized collagen or osteoid may be found in giant-cell tumors without their being malignant. Judging from the size of the lesion on X-ray and the amount of material received in the laboratory, the radiolucent lesion seems to have been filled with the material and, hence, the staff would prefer to use the term giant-cell tumor. It appears to be benign, and if all of the material has been removed followed by adequate chemical cauterization, the staff would anticipate that there would be little likelihood of further trouble with the tumor. Giant-cell tumors in the carpal bones tend to be somewhat atypical, as in this case."

In September, 1967, the patient was reported doing well although not yet allowed to bear weight on her foot. The bone grafts seem to have taken well.

Dr. Hatcher: Benign chondroblastoma is one of the most satisfactory bone lesions to treat surgically. It seems that a little curettage, obviously not affecting all of the

tumor cells, is sufficient to cause a healing reaction. Ordinarily in young children of this age it isn't necessary to pack the area with bone grafts because they heal so rapidly. I don't know of any benign epiphyseal chondroblastomas that have ever been said to become malignant. Once in awhile one of these lesions is situated in one of the epiphyses of a knee joint and may produce a reaction within the joint which may lead to contracture and stiffness of that joint; in general, they run a pretty harmless course. I suspect many of them may heal spontaneously.

Dr. Spjut: Dr. Carlos Perez-Mesa sent us a case like this a few years ago. I remember it was a chondroblastoma from the upper end of the femur that had been curetted several times and that the patient developed nodules in the lungs; thus, Dr. Perez-Mesa thought that it was a malignant chondroblastoma. I wonder if he has some follow-up information on this patient.

C. Perez-Mesa, M.D., Columbia, Missouri: The last I heard was about 4 years ago: the patient refused treatment;

despite the presence of two large metastases in the lung, he got married and his wife was pregnant about six months later.

Dr. Regato: Dr. Sudan, were there any calcium and phosphorus determinations?

A. C. Sudan, M.D., Denver, Colorado: We did those tests but they were within normal limits.

L. Lowbeer, M.D., Tulsa, Oklahoma: I know that Dr. Johnson calls these cases enchondromatous giant-cell tumors and I believe he has been quoted with the word "enchondromatous" left out. It is inconceivable that he would call that a true giant-cell tumor.

Dr. Regato: Dr. Sudan reaffirms that Dr. Johnson reported benign giant-cell tumor.

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13. Osteosarcoma of the Distal End of the Humerus

Contributed by **R. Marcial-Rojas, M.D.**, San Juan, Puerto Rico

THE PATIENT was a 16-year old boy in December, 1957, when he was hit by a baseball on the right arm and complained of severe pain.

Dr. Edeiken: There is an osteolytic lesion in the distal end of the humerus with a pathologic fracture. Even though the extremities of the lesion are well defined, there is one area at the distal end, of gradual transition from abnormal to normal bone which indicates rapid growth. There is no periosteal reaction or soft tissue mass.

Fig. 1—Osteolytic lesion of the distal end of the humerus with pathologic fracture, without periosteal reaction.



The early closure of the epiphyses suggests a hormonal abnormality, perhaps hyperthyroidism due to thyroid carcinoma or functional adrenal cortical tumor. With this evidence it is most likely a functioning thyroid metastasis. There is also a suggestion that the soft tissues show loss of the normal fascial planes and fat lines consistent with increased metabolism. However, normal children at this age may show this change.

In this age group one must always consider osteosarcoma as a likely choice if a bone lesion reveals malignant characteristics. However, one would expect periosteal reaction and some evidence of bone tumor formation.

Dr. Edeiken's impression: METASTATIC CARCINOMA (Thyroid?).

Roentgenologic Impressions Submitted by Mail

Osteosarcoma	28
Ewing's sarcoma	19
Fibrous dysplasia	10
Metastatic carcinoma	7
Others	32

Dr. Edeiken: In an Ewing's sarcoma we would not expect a localized lesion and would expect to see some periosteal reaction.

Dr. Regato: Dr. L. O. Martinez, of Miami, and Dr. J. C. Lemon, of Denver, also submitted an impression of metastatic carcinoma. Dr. M. Bischoff, of Denver, and Dr. J. Cox, of Colorado Springs, preferred Ewing's sarcoma. Dr. A. Litch, of Roswell, and Dr. J. F. Wilson, of Colorado Springs, offered osteosarcoma.

Operative findings: In January, 1958, the patient was operated upon; a frozen section diagnosis of giant-cell tumor was made and the lesion treated by curettement and impaction of bone chips. Three months later there were signs of recurrence in the roentgenogram and an amputation was done.

Dr. Spjut: In this case we have a fracture through what was probably a pre-existing lesion of bone. The problem that might arise is: could the lesion possibly have been secondary to the fracture? Histologically, we have lesional tissue that suggests more than a reaction to injury. The possibilities would include osteosarcoma and of the benign lesions, osteoblastoma. Throughout the section, one sees osteoid and calcified osteoid associated with fairly bizarre

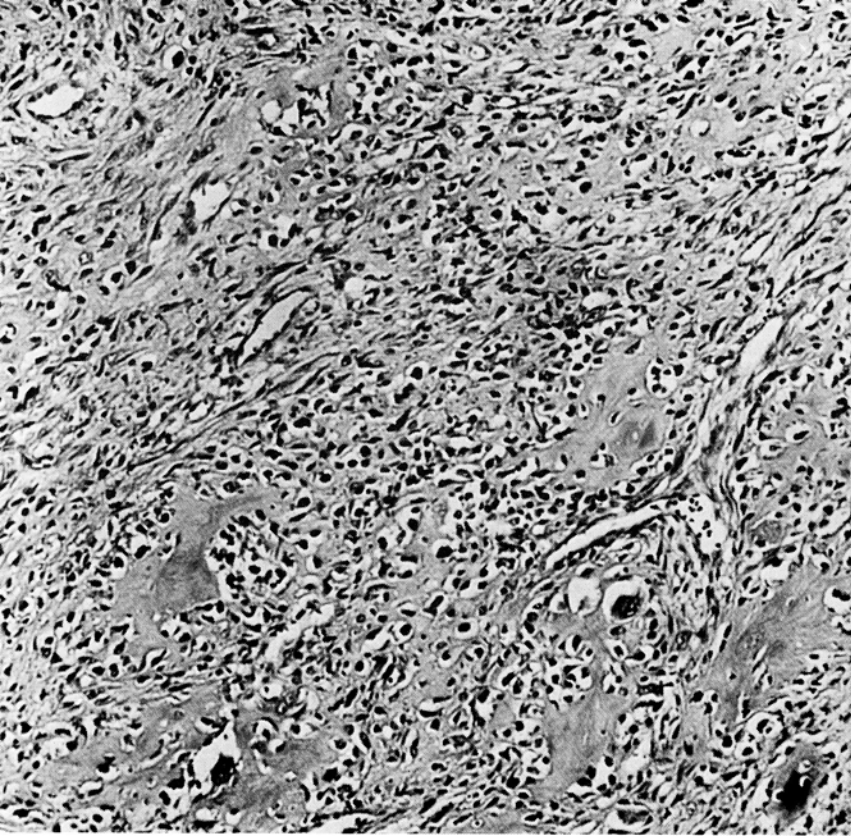


Fig. 2—Osteoid surrounded by pleomorphic osteoblasts and a malignant stroma indicate that this is an osteosarcoma. Hematoxylin-eosin 120X.

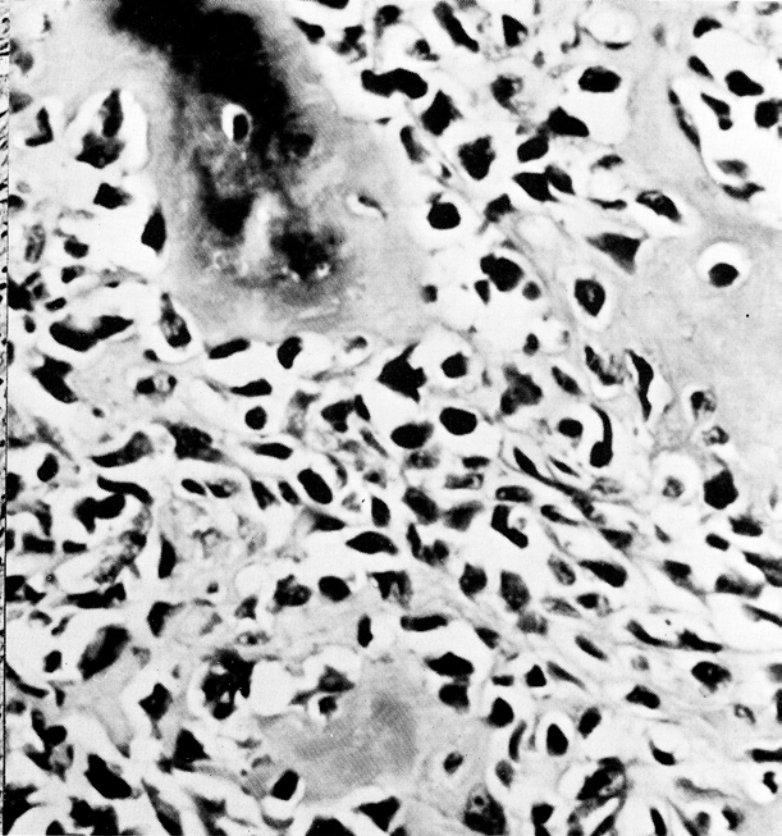


Fig. 3—Higher magnification of malignant osteoid and stroma. One spicule of osteoid is partly calcified. Hematoxylin-eosin 370X.

cells, presumably osteoblasts. The intervening stroma is vascular but it is generally not as vascular as one might expect to see with osteoblastoma. In addition, as one examines the cells associated with the osteoid, there are numerous bizarre features that lead one to consider that these are not benign osteoblasts. With osteoblastoma, to be sure, there are numerous osteoblasts around the osteoid and bony trabeculae, but under scrutiny these appear to be fairly uniform and do not suggest malignant neoplastic change. In this lesion, it appears that we have a malignant osteoid associated with a malignant stroma, giving us the histologic features we would need in order to make a diagnosis of osteosarcoma.

Oddly enough, pathological fractures through osteosarcomas are not common, having been reported in approximately 15% of cases. A question often asked about osteosarcoma is the frequency with which lymph nodes are involved. In a recent study from Memorial Hospital an incidence of approximately 3% was found. We have seen only occasional instances in which regional lymph nodes have been involved by osteosarcoma. The question of grading of osteosarcomas in relationship to prognosis has been discussed many times. Price indicates that there is a rather close correlation between the grade and the prognosis. In our own experience we find no correlation whatsoever in the grade and the expected survival. This corresponds to the observations that have been made at the Memorial Hospital in New York. In reviewing a large series of osteosarcomas with Drs. Ackerman, Edeiken, and Farrell, we found that there were no specific radiologic or histological findings that correlated with a predictable outcome. The histologic pattern, osteoblastic, fibroblastic, and chondroblastic, have been related to different prognoses by Dahlin.

Dr. Spjut's diagnosis: OSTEOSARCOMA.

Histopathologic Diagnoses Submitted by Mail	
Osteosarcoma	63
Osteoblastoma	32
Giant-cell tumor	15
Others	25

Dr. Spjut: This lesion does not have the characteristic histological features of osteoblastoma; I realize that this is a lesion that does cause considerable difficulty especially if you happen to get small curettings from a small osteoblastoma. I don't see how anyone would possibly support the diagnosis of giant-cell tumor; the lesion is in the wrong part of the bone and, although we do have a few multinucleated giant cells in the lesion, this does not make it a giant-cell tumor; you would have to say this is a malignant giant-cell tumor, in other words, an osteosarcoma.

Dr. Regato: Dr. A. O. Severance, of San Antonio, and Dr. M. H. McGavran, of Saint Louis, also submitted a diagnosis of osteosarcoma. Dr. R. A. Brooks, of Lincoln, and Dr. C. P. Schwinn, of Los Angeles, qualified it as a low-grade osteosarcoma. Dr. R. Boyer, of Colorado Springs, and Dr. F. Schajowicz, of Buenos Aires, preferred benign osteoblastoma.

Subsequent history: The patient did well until 1962 when he presented a pneumothorax and a small pulmonary lesion was found. A segmental resection was done but a pulmonary recurrence took place; the patient expired in 1963. At autopsy there were disseminated pulmonary metastases.

R. Marcial-Rojas, M.D., San Juan, Puerto Rico: This case presented certain interesting features; on the original curettings the pathologist called it a giant-cell tumor. The orthopedic surgeon knew it couldn't be a giant-cell tumor because of the age and location of the tumor. Dr. Lichtenstein was consulted and wrote that he did not know what the tumor was, that it was an offbeat osteochondroid tumor that defied a name, but that he was certain that it was benign. However, it was acting malignant locally and the boy received the amputation. At autopsy the left lung was solid as a rock with metastatic osteogenic sarcoma. The metastases were localized to the left lung. The right lung had three isolated metastases.

Dr. Hatcher: Without the pathologic fracture, the tumor in that region might have been resected with some success; once there is a fracture there is hemorrhage out into the

fascial planes and tumor cells are carried outside the field: the chances of survival are lessened. This appears to be histologically a relatively low-grade sarcoma but as far as the patient is concerned, it is a very high grade sarcoma.

Dr. Edeiken: A pathologic fracture as a presenting symptom is most often seen in benign disease and in this age group is usually seen with unicameral bone cysts; there is a perfectly good reason for that and that is that the benign lesions tend not to cause symptoms and, therefore, there is nothing done until it has eroded the cortex and weakened the bone. Osteosarcomas and other malignant tumors cause pathologic fractures in about 20% of the patients.

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14. Malignant "Lymphoma" of the Head of the Femur

Contributed by **J. A. del Regato, M.D.** and **M. Berthrong, M.D.**, Colorado Springs, Colorado

THE PATIENT was a 62-year old man in December, 1962, when he gave a one-year history of pain in the left hip which had become recently severe; there was a history of compression fracture of the right femoral head which had been treated by prosthesis. On examination, there was shortening of several centimeters in the left lower extremity.

Dr. Edeiken: The roentgenograms reveal a large osteolytic lesion in the subtrochanteric area of the left femoral neck and shaft with pathologic fracture. The destruction is incomplete leaving many trabeculations behind. Soft tissue calcification is present at the inferior medial margin. There is callus formation. A roentgenogram taken three months after the initial study shows no change.

The most common primary malignant lesions in the neck of the femur are chondrosarcoma and fibrosarcoma. They usually do not attain this size and frequently develop a surrounding sclerotic margin. Chondrosarcomas of this size will usually show calcifications. Fibrosarcomas will never calcify the matrix and therefore this tumor cannot be excluded. The soft tissue calcifications noted at the inferior margin of the fracture are probably related to hematoma and not to calcification within soft tissue tumor. If it were tumor calcification we would have to consider a primary osteosarcoma or chondrosarcoma.

In this age group, one must also consider metastatic disease. Metastases often are completely osteolytic and do not often leave behind bone trabeculations or spicules of bone as seen in this instance. Multiple myeloma and other round-cell tumors such as lymphoma and reticulum-cell sarcoma cannot be excluded. From the standpoint of differential diagnosis, the most likely lesion in this age group would be either metastatic disease, multiple myeloma and other round-cell tumors.

Dr. Edeiken's impression: ROUND-CELL TUMOR (Lymphoma? Plasmacytoma?)

Roentgenologic Impressions Submitted by Mail

Osteosarcoma	25
Reticulum-cell sarcoma	22
Chondrosarcoma	10
Metastatic carcinoma	9
Myeloma	7
Others	26

Dr. Edeiken: Osteosarcoma is not very common at this age but there is an increased incidence of osteosarcoma in the older age groups not attributable to Paget's disease. I can't find anything on the roentgenogram that would lead me to that diagnosis although it's a possibility. Chondrosarcoma is possible but we do not have calcification. Metastatic carcinoma is another excellent possibility.

Dr. Regato: Dr. R. Henschel, of Saginaw, submitted osteosarcoma. Dr. J. T. McClintock, of Denver, offered chondrosarcoma. Dr. A. Ritch, of Roswell, suggested solitary myeloma. Dr. R. P. Spurk, of Denver, and Dr. L. O. Martinez, of Miami, preferred reticulum-cell sarcoma.

Operative findings: In December, 1962, a biopsy was done and a diagnosis of reticulum-cell sarcoma of bone was rendered.

Dr. Spjut: Histologically, this lesion is difficult to classify. One has to consider the possibilities of malignant lymphoma, histiocytosis and, because of the identifiable plasma cells, a myeloma. To me, the dominant cells are not plasma cells, even though there are a number present. The dominant cells seem to be a lymphocyte and a large cell that has an indented nucleus. In some areas cells that appear to be reticulum cells dominate the pattern. Other than the background cells that in some areas appear to be histiocytes, I don't believe that this histologic pattern is strongly suggestive of a histiocytosis. Multinucleated giant cells and eosinophils are absent or very few in number. A reticulum stain of the section reveals that in the areas in which reticulum cells are abundant that reticulum fibers are prominent. I would favor designating this lesion as a malignant lymphoma of bone and from the clinical standpoint, apparently primary. I'm not sure that I can subclassify the tumor; perhaps it is a lymphoma. One may often see mature lymphocytes mixed with a lesion of bone designated as a primary reticulum-cell sarcoma of bone. We would agree with Ivins' and Dahlin's designation of this type of lesion as a malignant lymphoma rather than specifically a reticulum-cell sarcoma. There is some importance in delineating this lesion in bone, as it has been shown that the expected 5-year survival rate is approximately 35%. As to location, the long bones of the upper and lower extremities are the most common sites of these lesions. The flat bones have been the seat of a few.

Dr. Spjut's diagnosis: MALIGNANT LYMPHOMA (?)



Fig. 1—Osteolytic lesion of femoral neck with pathologic fracture (1962).

Histopathologic Diagnoses Submitted by Mail

Myeloma	34
Reticulum-cell sarcoma	33
Malignant lymphoma	23
Hodgkin's disease	17
Eosinophilic granuloma	10
Histiocytosis	8
Others	21

Dr. Spjut: Myeloma one has to consider because we do have recognizable plasma cells. Then the question that I really can't answer is: could the background cell here really represent an undifferentiated plasma cell? I suppose that is a possibility. Perhaps the clinical work-up of this patient would give us some information as to the serum electrophoresis that might help us in this differential diagnosis. Hodgkin's disease is a rather uncommon malignant lymphoma of the bone; in this case, in order to make that diagnosis, one would have to demonstrate satisfactory Dorothy Reed-Sternberg cells and I could find none. In the absence of that one histological feature, we could not support Hodgkin's disease. There are a few eosinophils in here to be sure and I don't think we have much else to support a diagnosis of eosinophilic granuloma; we don't have a background cell of the histiocyte; we have previous few, if any, multinucleated cells. The dominant cell here seems to be along the lymphoid series. It would be the same for the histiocytosis.

Dr. Regato: Dr. M. H. McGavran, of Saint Louis, Dr. M. R. Abell, of Ann Arbor, and Dr. F. Schajowicz, of Buenos Aires, all were agreed on a diagnosis of malignant lymphoma. Dr. C. P. Schwinn, of Los Angeles, preferred the designation "malignant tumor of the reticulo-endothelial system" meaning Hodgkin's, leukemia, et cetera, but "not histiocytosis X". Dr. T. H. Snider, of Fort Sam Houston, made a diagnosis of histiocytosis X. Dr. L. Lowbeer, of

Fig. 2—Immature lymphocytic cells that composed a major portion of the lesion. Hematoxylin-eosin 300X.

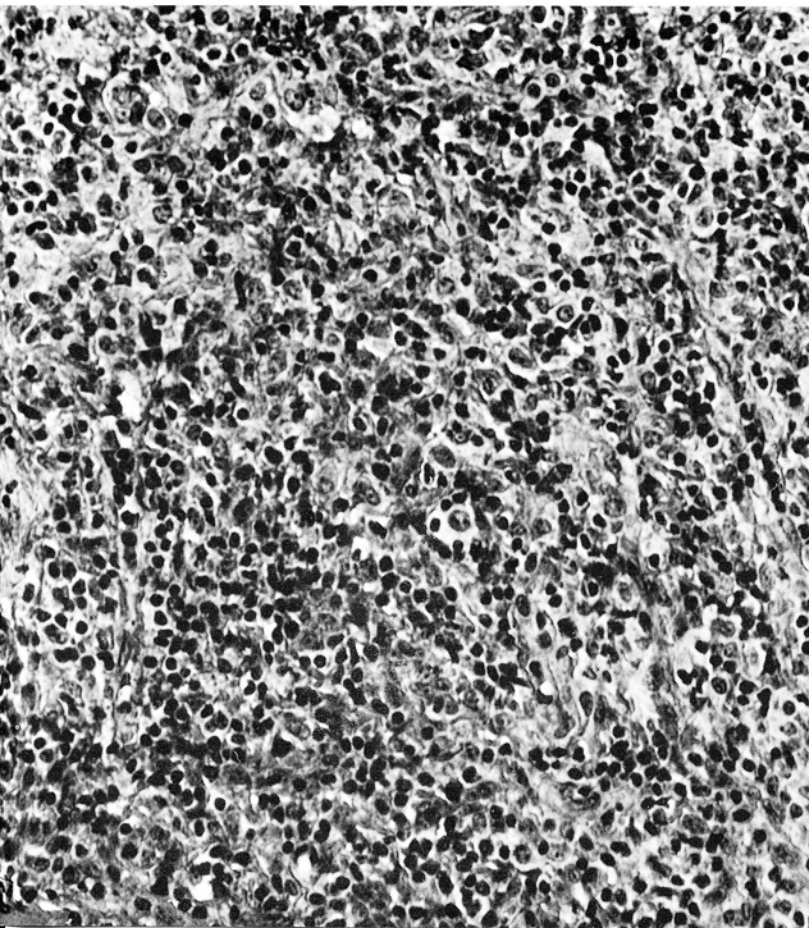
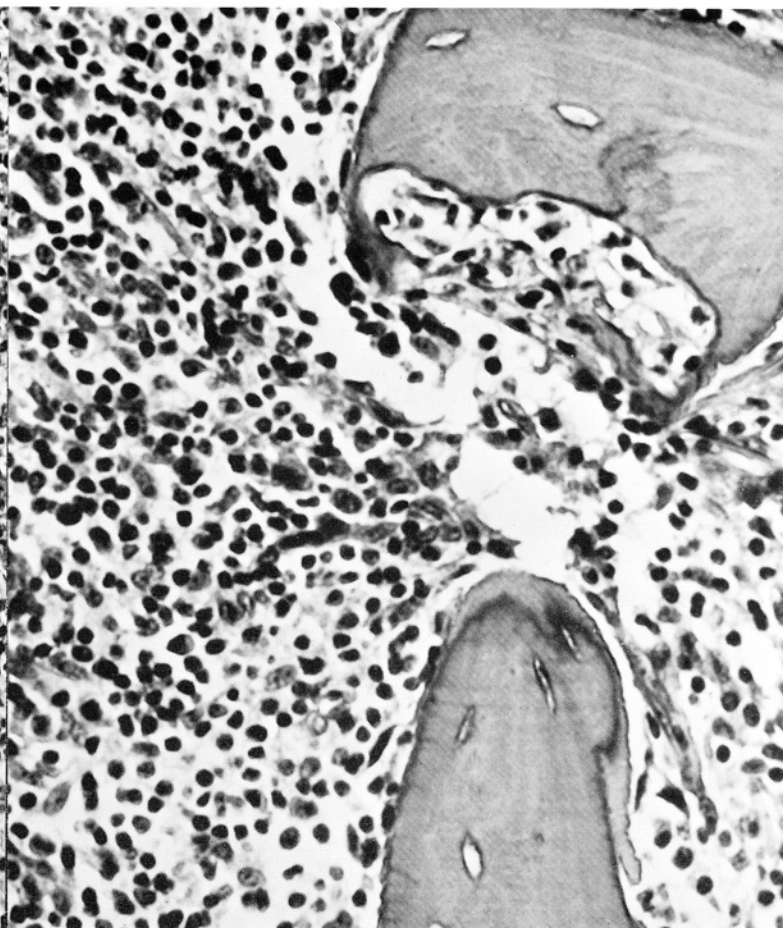


Fig. 3—Residual bone in a field of immature lymphoid cells. Hematoxylin-eosin 300X.



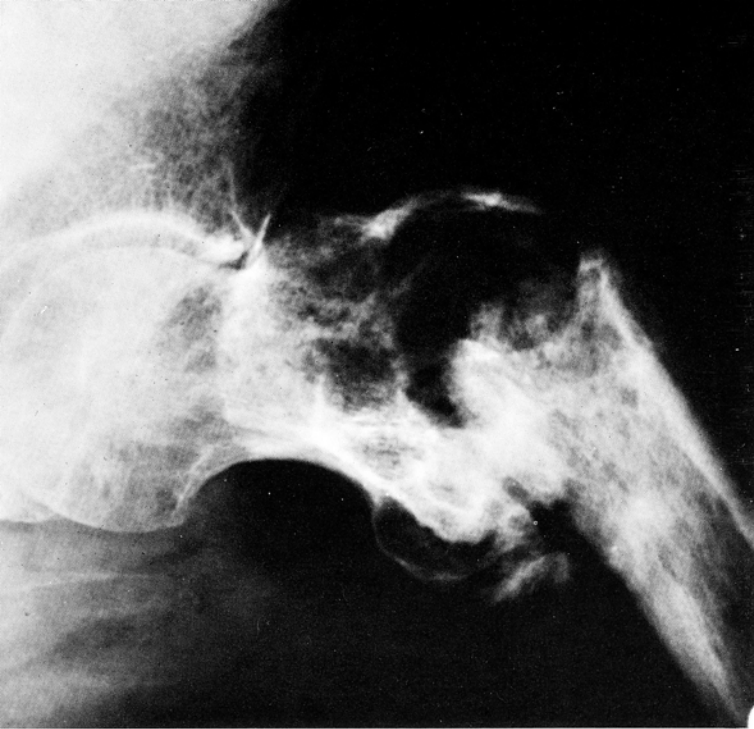


Fig. 4—Roentgenogram taken in September, 1966, showing fracture but no recurrence of tumor after radiotherapy.

Tulsa, gave it the designation of pleomorphic marrow tumor and wondered if it could be Hodgkin's sarcoma. Dr. H. L. McGaffey, of Idaho Falls, called it a reticulum-cell sarcoma. Dr. B. Sharma, of Portland, Oregon, and Dr. R. Marcial-Rojas, of San Juan, saw it as a solitary myeloma. Dr. T. H. McConnell, of Dallas, preferred Ewing's sarcoma. Dr. W. R. Platt, of Saint Louis, saw a granulomatous process, and Dr. A. O. Severance, of San Antonio, called it an eosinophilic granuloma.

Subsequent history: From December 19, 1962, to February 15, 1963, the patient received Cobalt-60 teletherapy; the tumor was irradiated through an anterior and a lateral field receiving a calculated dose of 6,600 rads in 58 days.

There was early recalcification of the lesion; the patient left his crutches, walked with the help of a cane and, later, without one. He did well until February, 1966, when he presented with a left lower abdomino-pelvic mass, 18 x 22 cm in diameter, fixed to the bone; there was no bony destruction, the IVP showed displacement of the left ureter.

It was assumed that the mass represented nodular lymphatic metastases and he was submitted to irradiations; from February 14, 1966, to April 16, 1966, he received a calculated dose of 6,570 rads at the pelvic mid-plane, in 61 days. There was only a slight regression of the mass. A needle biopsy was done which showed malignant cells compatible with a diagnosis of reticulum-cell sarcoma. The patient submitted to a surgical exploration which revealed a large retroperitoneal mass which was removed: it consisted, for the most part, of blood and necrotic material. Microscopic examination was reported as showing apparently malignant cells in the periphery of the mass. Post-operatively, the patient developed a new large hemorrhagic mass for which he was successfully operated upon. In September, 1966, the patient presented sudden pain in the left hip and the roentgenograms showed a fracture but no recurrence of the tumor; the patient refused pinning. In March, 1967, the patient was again ambulating comfortably.

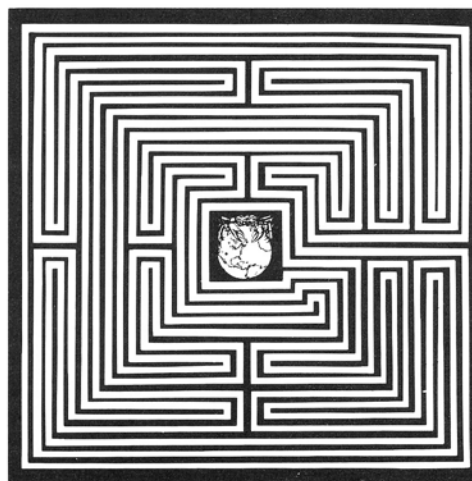
Dr. Spjut: Dr. del Regato sent me an extra slide on this case from the abdominal mass. However, this slide does not help in the diagnosis of this lesion as it shows extensive necrosis of tumor that is unidentifiable plus extensive recently extravasated blood.

Dr. Regato: In December, 1962, his total serum proteins were 5.3 gm %, the albumin being the main factor reduced; relative percentages of globulins were normal. The bone marrow biopsies have all been reported as normal. The patient has had no further bony manifestations to date, five years after initial radiotherapy.

Dr. Hatcher: In the older person, it's very difficult to be sure that the lesion in the bone is a primary reticulum-cell sarcoma. Very often there may be a primary focus elsewhere in the lymphatics which has already metastasized to the skeleton. Reticulum-cell sarcoma has very largely been taken out of the surgeons' hands except for the purpose of biopsy. In general, the preferred treatment for either reticulum-cell or lymphoma is radiotherapy rather than surgery; there may be occasional instances in which one would prefer to excise the lesion surgically. One might consider a palliative removal of his lower extremity but most people are reluctant to lose an extremity unless they have to and the chances of getting union of the fracture after the tumor is killed by irradiation is pretty good.

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15. *Metastatic (renal?) Carcinoma of the Iliac Bone*

Contributed by **W. K. Bullock, M.D.**, Los Angeles, California and **D. R. Dickson, M.D.**
and **P. Riemenschneider, M.D.**, Santa Barbara, California

THE PATIENT was a 54-year old man in April, 1962, when he complained of pain in the right side of the pelvis.

Dr. Edeiken: There is a large osteolytic lesion in the lateral aspect of the right iliac wing. A soft tissue mass is associated. Several pathologic fractures are also present. It is interesting to note that the margin is sclerotic. Bone reaction in multiple myeloma is so rare that if it occurs in a lesion, one should exclude this diagnosis. Most other malignant tumors of bone may have a sclerotic margin and therefore the differential diagnosis occurring in this age group should include metastatic lesions as well as primary tumors. Metastatic lesions from the kidney often produce a large osteolytic lesion with associated soft tissue mass. Chondrosarcoma and osteosarcoma may occur at this site but usually there is some matrix calcification to lead one to the diagnosis. Fibrosarcomas may also occur at this site and calcification of the matrix does not occur. They are also slow growing and may have a sclerotic margin.

The roentgenographic diagnosis cannot be made in this case with certainty, but it appears to be due to a slow growing lesion which may either be metastatic or of primary nature. The two most likely considerations are metastatic carcinoma of the kidney and fibrosarcoma.

Fig. 1—Osteolytic lesion of iliac wing with sclerotic margin.



Dr. Edeiken's impression: 1) METASTATIC CARCINOMA, (Kidney?) 2) FIBROSARCOMA.

Roentgenologic Impressions Submitted by Mail

Metastatic carcinoma	19
Plasmacytoma	16
Chondrosarcoma	15
Reticulum-cell sarcoma	7
Fibrosarcoma	6
Others	25

Dr. Edeiken: I don't like that margin; it is unusual in metastatic carcinoma; it is unheard of in plasmacytoma. This is a favorite place for chondrosarcoma. I would like to see it a little closer to the sacroiliac joint, but not necessarily. There is no calcification of the matrix in this case so that this diagnosis cannot be excluded or included. This could well be a reticulum-cell sarcoma.

Dr. Regato: Drs. M. Bischoff, of Denver, and J. F. Wilson, of Colorado Springs, also offered fibrosarcoma. Dr. E. Salzman, of Denver, suggested plasmacytoma. Dr. B. L. Pear, of Denver, preferred metastatic carcinoma, possibly renal.

Operative findings: On April 9, 1962, a biopsy was done followed by extensive bleeding.

Dr. Spjut: Histologically, this case has been a puzzle. Several diagnoses have been considered and among them: metastatic renal cell carcinoma, xanthogranuloma, rhabdomyosarcoma, leiomyosarcoma, fibrosarcoma, and liposarcoma. If this were a rhabdomyosarcoma or a leiomyosarcoma, one would expect this to be an invasive lesion. Apparently, we have no historical or physical evidence to suggest that this patient had a soft tissue mass. Liposarcomas, however, have been reported as being primary in bone and with the spindle cell component, the clear cells, and the cells that appear to contain fat, one may have to consider this as a possibility. Xanthogranulomas, benign and malignant, occur in soft tissues. Whether this is such a lesion is difficult to prove. The numerous vacuolated cells, the spindle cell component, plus the numerous inflammatory cells, lymphocytes and plasma cells, suggest this lesion. However, in reviewing the papers reported on malignant xanthogranulomas of soft tissue, this lesion does not have the storiform pattern of the spindle cells which apparently is characteristic of this disease. Perhaps the most enticing lesion to be considered is that of the metastatic renal cell carcinoma. It is well known that the poorly differentiated renal cell carcinomas may resemble a sarcoma as this lesion does. In fact, a pseudosarcomatous pattern, pure and mixed with clear or granular cells, has been listed in classifications of renal cell carcinomas. Also, we do have a destructive lesion of bone which would fit with renal cell carcinoma. Metastases to bone are present in one-third of the patients with renal cell carcinoma, and represent the most common site in one series. It would be interesting to know whether fat stains were done on this lesion and if they were positive. This may not be especially helpful, as renal cell carcinoma, liposarcoma, and xanthogranuloma would probably be positive. Fibrosarcoma might be ruled out. We did oil-red-O stains on the extra sections sent to us but residual lipid could not be identified. My tentative diagnosis is metastatic renal cell carcinoma with liposarcoma, fibrosarcoma, and xanthogranuloma as possi-

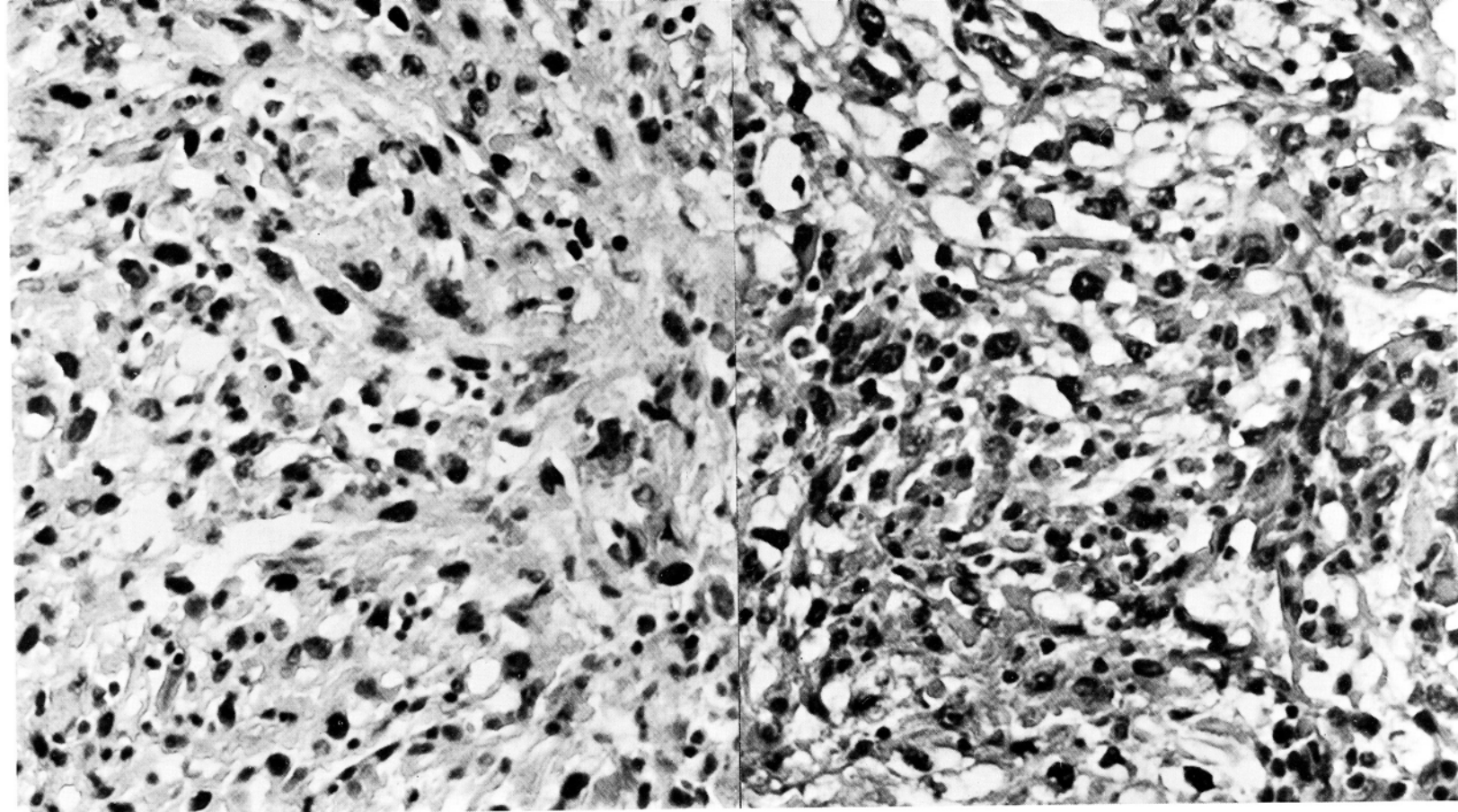


Fig. 2—Pleomorphic spindle-cell stroma. Hematoxylin-eosin 370X.

Fig. 3—In other areas the cells appear to have a clear cytoplasm suggesting renal cell carcinoma. Inflammatory cells are present. Hematoxylin-eosin 370X.

bilities that cannot be ruled out. It is noted that this case does come from a patient who was first seen in 1962, suggesting the possibility that if this is metastatic carcinoma, that the patient may still be alive. This may be against a metastatic lesion.

Dr. Spjut's diagnosis: Probable METASTATIC CARCINOMA (Kidney?)

Histopathologic Diagnoses Submitted by Mail

Liposarcoma	41
Metastatic carcinoma	39
(Kidney 28)	
Fibrosarcoma	12
Rhabdomyosarcoma	12
Other sarcomas	30
Others	12

Dr. Spjut: I can't say that this is not liposarcoma and I leave that as a possibility. Most fibrosarcomas are not this pleomorphic. Usually when a fibrosarcoma becomes poorly differentiated it becomes more cellular than pleomorphic, but occasional ones may be pleomorphic. I have mentioned the possibility of rhabdomyosarcoma and then I suppose there are many other sarcomas that people might consider.

Dr. Regato: Dr. D. Dawson, of Colorado Springs, also

made a diagnosis of probable metastatic renal carcinoma. Dr. W. R. Platt, of Saint Louis, offered liposarcoma; Dr. A. O. Severance of San Antonio, rhabdomyosarcoma. Dr. C. P. Schwinn, of Los Angeles, fibrosarcoma; Dr. M. H. McGavran, of Saint Louis, xanthofibrosarcoma. Dr. R. Schultz, of Sioux Falls, offered Hodgkin's; Dr. R. Boyer, of Colorado Springs, malignant Schwannoma. Dr. G. Simon, of New Haven, concluded to an osteolytic osteosarcoma and wondered if it was secondary to Paget's disease.

Subsequent history: It was decided to irradiate the patient pre-operatively then a hemipelvectomy was attempted and abandoned because of bleeding. Additional radiotherapy was given but the patient continued to have increasing pain. He expired on August 25, 1963; no autopsy was done.

Dr. Hatcher: There is evidence in favor of a carcinoma metastatic from the kidney: these tumors are quite characteristically bloody and sometimes one has to obtain the biopsy from the drapes after he has cut into the tumor.

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OUR GUEST SPEAKERS



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