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Osteogenic sarcoma

Report of one hundred and fifty-seven cases

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This study presents the clinical features, roentgenographic manifestations, pathological features, abnormal laboratory studies, treatment and prognosis of 157 cases of osteogenic sarcoma. The series is the fifth largest ever reported.

The authors favor prompt surgical ablation followed by controlled intensive chemotherapy as a prophylactic or combined chemotherapy and radiation therapy for metastatic osteogenic sarcoma.

All case reports but one present rare or unusual situations.

ALTHOUGH osteogenic sarcoma is the second most common malignant primary bone tumor, the incidence of its occurrence is relatively rare when compared with carcinomas generally. Of 40,282 cancer cases seen and treated at Henry Ford Hospital from 1916 to 1975, only 157 cases of osteogenic sarcoma were encountered. This means that osteogenic sarcoma represents 0.39% (or approximately four per thousand) of all the malignancies seen and treated at our medical center, yet only four other centers in the world have reported a larger series.

Our series of 157 osteogenic sarcomas consists of 133 primary osteogenic sarcomas, seven extraskeletal osteogenic sarcomas,[1-4] six parosteal (juxtacortical) osteogenic sarcomas,[5-7] eight Pagetoid osteogenic sarcomas[8-19] and three postradiation osteogenic sarcomas.[20-28] Excluding the 133 primary osteogenic sarcomas, clinical data on 24 cases of different osteogenic sarcomas are presented in the following tables.

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TABLE I

EXTRASKELETAL OSTEOGENIC SARCOMA

Case Number	Age & Sex	Site	Symptoms	Treatment	Results
1	5, M	Right gluteus maximus muscle	Mass in right buttock	Radical en bloc excision of right buttock	No recurrence or metastases 9 years after surgery
2	36, M	Left heel	Pain and swelling of the left heel	Below-knee amputation; wedge resection of left lung for solitary metastasis; chemotherapy	Died of widespread metastases 51 months after amputation
3	42, F	Left thigh and buttock	Pain and swelling of the left thigh and buttock	Four failed local excisions followed by left hemipelvectomy	No recurrence or metastases 13 months after hemipelvectomy
4	63, M	Left axilla	Large growing mass in left axilla	Left forequarter amputation	Free of malignant disease 11 years after forequarter amputation
5	66, F	Right thigh	Large tumor in right thigh	Left hip disarticulation	Died of bony and visceral metastases 26 months after hip disarticulation
6	70, M	Dura mater of the left	Severe headaches	Left frontal craniotomy	Died 2½ months after craniotomy
7	78, M	Right calf	Pain and swelling of the right calf	Radical excision of soft tissues of right calf	No recurrence or metastases 13 years after radical local excision

TABLE II

PAROSTEAL (JUXTACORTICAL) OSTEOGENIC SARCOMA

Case Number	Age & Sex	Site	Symptoms	Treatment	Results
1	20, M	Right distal humerus	Pain and swelling of the right elbow	High above-elbow amputation	No evidence of disease 9 years after amputation
2	31, M	Right distal femur	Pain and swelling of the right distal thigh	High thigh amputation	No evidence of recurrence or metastases 19 years after surgery
3	37, F	Right distal femur	Pain and swelling in the right knee region	High thigh amputation	Died 15 months later from widespread metastases
4	38, M	Left proximal tibia	Pain and swelling of the left popliteal fossa	Above-knee amputation	Died 21 months later from extensive pulmonary metastases
5	38, F	Left ulna	Pain and swelling of the left forearm	Above elbow amputation	Alive and well 5 years after amputation
6	45, M	Right proximal tibia	Pain and swelling of the right knee	Above knee amputation	Died 7 months later from metastases

TABLE III

SECONDARY OSTEOGENIC SARCOMAS

Predisposing Disease or Condition	Age & Sex	Site	Symptoms	Treatment	Results
Paget's Disease	46, F	Both femora and right ilium	Pain and swelling of both distal thighs and right ilium	Left hip disarticulation and chemotherapy	Died 15 months later from metastases
	56, F	Right femur	Pain and swelling of right thigh	Right high thigh amputation	Died 9½ months later from metastases
	58, F	Right tibia	Pain in vicinity of right knee	Above knee amputation	Died 6 months later from metastases
	59, F	Left tibia	Pain and swelling of left knee	Above knee amputation	Died 10 months later from metastases
	61, M	Left proximal femur	Pain in left hip	Hemipelvectomy	Died one year later from metastases
	61, F	Right femur	Pain in right knee	Hip disarticulation	Died 7 months later from metastases
	64, F	Left humerus	Pain and swelling of the left shoulder	Radiotherapy	Died 3 months later from metastases
	71, F	Frontal bone of skull	Pain and swelling of the forehead	Left frontal craniotomy	Died 2 weeks later from brain involvement

TABLE III (Continued)

SECONDARY OSTEOGENIC SARCOMAS

Predisposing Disease or Condition		Age & Sex	Site	Symptoms	Treatment	Results
External Radiation	Osteochondroma	29, M	Right pubic bone	Pain and swelling of right groin area	Symptomatic treatments because of widespread metastases	Died 4 months later from extensive pulmonary metastases
		32, F	Right ribs	Pain and swelling of right chest wall	Radical excision of right chest wall, including right ribs and part of right diaphragm, lung and abdominal wall	Died one month later from extensive metastases
	Giant Cell Tumor	67, M	Left distal femur	Pain and swelling of right distal thigh	High thigh amputation	Died 8 months later from extensive pulmonary metastases

Our 133 cases of primary osteogenic sarcoma included 68 males and 65 females, 124 of them white and 9 black. Table IV shows the age distribution.

TABLE IV

AGE DISTRIBUTION

Age	Number of Patients
0-9	4
10-19	39
20-29	21
30-39	16
40-49	10
50-59	15
60-69	19
70-79	7
80-89	2

The sites of primary osteogenic sarcoma were most frequently in the long bones, 49.8% of them (65 patients) being located in a femur. In 12 patients (9%), the primary site was a humerus and in 10 patients (7%) it was a tibia. Eight patients (6%) had primary osteogenic sarcoma in an ilium. The distribution in all the other patients was as follows: Five each in rib or mandible, four each in fibula, clavicle, maxilla or sacrum; three each in the thoracic spine or lumbar spine, and one each in pubic bone, radius, skull, orbit, hand or foot.

Among signs and symptoms, pain and swelling are the most common principal complaints. Pathological fractures occurred in four femora, three humeri, one tibia and one clavicle. Two vertebral and one cranial osteogenic sarcomas caused significant neurological deficits by invading the adjacent central nervous system. In addition, some patients had constitutional symptoms which included fever, malaise, weight loss, weakness and anorexia, especially when metastases had already occurred.

Examination often revealed localized swelling and tenderness. Synovial effusion, muscular atrophy, loss of joint motion, edema of the involved extremity, prominent superficial veins and other evidence of localized hyperemia were also present in several patients.

The majority of the osteogenic sarcomas originated in the metaphyseal portion of a long tubular bone. Their typical radiological features included intramedullary trabecular destruction, cortical erosion and perforation, adjacent soft tissue extension and invasion, fairly uniform radiopacity throughout the tumor, and prominent periosteal reaction. A few were purely osteolytic in nature. Their diagnoses were determined only after biopsies.

Pathologic features

1. Gross pathology

The sectioned surfaces of many of the osteogenic sarcomas were quite hard to the touch and had a whitish or yellowish-white appearance. The intramedullary involvement, cortical destruction and extraosseous extension could be seen readily. In contrast, the osteolytic and the less ossified osteogenic sarcomas tended to be softer and sometimes showed areas of necrosis, cystification, hemorrhage and telangiectasis. A few chondroblastic osteogenic sarcomas grossly resembled chondrosarcoma by their bluish-gray, glistening and lobulated appearance.

2. Microscopic pathology

The histological features in the series fell into one or more of the following four basic patterns:

a. Osteoblastic osteogenic sarcoma characterized by the presence of malignant-looking osteoblasts intimately associated with abundant tumor os-

TABLE V

LABORATORY VALUES OF PATIENTS AFFECTED BY OSTEOGENIC SARCOMA

Name of Laboratory Study	Total Number of Patients	Number of Patients with Abnormal Laboratory Study	Percent (%)
Alkaline Phosphatase	68	36	53.0
WBC	84	14	16.7
HGB	86	10	11.6
Calcium	62	2	3.2

teoid. (The majority of our cases belonged to this group.)

b. **Chondroblastic osteogenic sarcoma** — usually contains fields of malignant cartilaginous tissue in addition to malignant osteoblasts and associated tumor osteoid.

c. **Fibroblastic osteogenic sarcoma** — typically possesses many malignant-appearing fibroblasts associated with significant amount of intercellular collagen. However, on close scrutiny, scattered strands of eosinophilic, homogenous, afibrillar and amorphous substance, suggestive of osteoid, help to differentiate fibroblastic osteogenic sarcoma from the bona fide intraosseous fibrosarcoma.

d. **Poorly differentiated osteogenic sarcoma** — the least common osteogenic sarcoma, contains no malignant osseous or cartilaginous tissue and very little fibrous tissue. The cells are usually very pleomorphic and bizarre and show many mitotic figures.

Abnormal laboratory studies

Since alkaline phosphatase determinations did not become available to us as a routine laboratory study until about 1950, the values in Table V include only the pre-amputation serum alkaline phosphatase and calcium levels, white blood cell counts and hemoglobin values of the victims of osteogenic sarcoma in our series since 1950.

Treatment

Prior to the availability of chemotherapeutic agents, metastatic and unresectable osteogenic sarcomas were treated with radiotherapy, but now these same tumors are treated with either chemotherapy alone or in combination with radiotherapy.

In this series, 19 patients with osteogenic sarcoma of the extremities were given a tumor dose of radiation to the involved limbs, which were later amputated. None survived beyond two years after amputation.

Most of the other osteogenic sarcomas in our series were treated with amputation. Eighteen survived five years or longer.

Currently, our preferred treatment method consists of prompt amputation as soon as unequivocal tissue diagnosis has been estab-

lished. Soon after amputation, the patient is referred to an oncologist for combined prophylactic chemotherapy.

Two patients developed solitary pulmonary metastasis post-amputation and underwent lobectomies. One survived two years after lobectomy while the other lived only four months. We feel that in the absence of other metastases, excision of resectable pulmonary metastases should be coupled with carefully controlled and supervised long-term chemotherapeutic regimens.

Prognosis

Table VI shows the final outcome of our 133 cases of primary osteogenic sarcoma. Of the 113 fatalities, 98 (or 86.7%) occurred within the first two years after diagnosis. Of the 18 patients who survived five years or more, none succumbed to osteogenic sarcoma, a cure rate of this series to date which is equivalent to a five-year survival rate of 13.5%. Of the 18 survivors, 8 were males and 10 females (from a total of 68 male and 65 female patients). They represent a 11.8% five-year survival rate for the males and a 15.4% five-year survival rate for the females.

Eight patients were treated with amputation and adjuvant chemotherapy within the past three years. Their clinical data are summarized in Table VII. The data in Table VII reveal that two patients (or 25%) died within one year; four (or 50%) within two years; and seven (or 87.5%) within three years. Their survival statistics seem to indicate that modern chemotherapy in our limited experience prolongs survival time by about one year and does not appear to affect the long-term survival rate.

Furthermore, using the clinical data from these 157 patients, these observations can be made:

1. Extraosseous osteogenic sarcoma seems to have a better long-term prognosis than intraosseous osteogenic sarcoma. Of seven patients affected by extraosseous osteogenic sarcoma, four (or 57.1%) survived five years or longer.
2. Amputation was curative in three of six cases of parosteal osteogenic sarcoma with five-year follow-up.
3. None of our eleven cases of Pagetoid and postradiation osteogenic sarcoma survived beyond one year after diagnosis. The evidence suggests that secondary osteogenic sarcoma carries an extremely unfavorable prognosis.

TABLE VI

SURVIVAL STATISTICS FOR 133 CASES OF OSTEOGENIC SARCOMA

Deceased Patients		Surviving Patients	
Deaths within 2 Years	Deaths within 5 Years	Survivals under 5 Years	Survivals 5 Years or Longer
98	113	2	18

TABLE VII

CLINICAL DATA OF 8 PATIENTS TREATED WITH AMPUTATION AND ADJUVANT CHEMOTHERAPY

Case	Age & Sex	Site	Surgical Treatment	Chemotherapeutic Agents Used	Survival Time
1	13, M	Distal femur	High thigh amputation	Adriamycin	Died 8 months later
2	12, M	Distal femur	Above knee amputation	Adriamycin Vincristin	Died 11 months later
3	15, F	Distal femur	Hip disarticulation	Actinomycin-D Vincristin Cytoxan	Died 14 months later
4	25, M	Proximal humerus	Forequarter amputation	Actinomycin-D Oncovin Cytoxan	Died 21 months later
5	12, M	Distal femur	Hip disarticulation	Adriamycin Oncovin Cytoxan Ict	Died 27 months later
6	15, M	Proximal tibia	Above knee amputation	Adriamycin Actinomycin-D Cytoxan Oncovin	Died 32 months later
7	24, F	Distal femur	Hip disarticulation	Actinomycin-D Vincristin Cytoxan	Died 33 months later
8	15, F	Distal femur	Above knee amputation	Adriamycin	No recurrence or metastasis 25 months later

4. Unresectable osteogenic sarcoma (such as our 10 cases of vertebral and sacral osteogenic sarcoma) and metastatic osteogenic sarcoma were invariably found to be lethal. Radiotherapy uniformly failed to prevent the fatal outcome.

5. Not one among the nine patients with spontaneous pathological femoral, humeral, tibial and clavicular fractures survived. Perhaps the pathological fracture merely reflects the extremely destructive, invasive and metastasizable nature of the underlying osteogenic sarcoma.

6. The presence of abnormally high serum alkaline phosphatase level, leukocytosis, anemia and hypercalcemia, in the absence of any other detectable system diseases, indicates a poor prognosis. Not a single patient who had two or more of these abnormal laboratory values survived five years or more.

7. The presence of constitutional symptoms, such as fever, malaise, weight loss, weakness, anorexia, etc., drastically reduces the chances of long-term survival. Overt constitutional symptoms usually indicate that systemic metastases have already occurred.

Summary

Extraosseous osteogenic sarcoma, parosteal osteogenic sarcoma, and female patients tend to have a better prognosis. In contrast, secondary osteogenic sarcoma; metastatic and unresectable osteogenic sarcoma; pathological fractures; the presence of elevated serum alkaline phosphatase level, leukocytosis, anemia and hypercalcemia; and the presence of constitutional symptoms seem to indicate a poorer prognosis. The overall five-year survival rate for our 133 cases of primary osteogenic sarcoma is 13.5%.

We feel that prompt surgical ablation followed by carefully controlled intensive prophylactic chemotherapy is the treatment of choice. Excision of resectable pulmonary metastases should be combined with chemotherapy and radiotherapy for metastatic osteogenic sarcoma.

Some unusual and illustrative cases.

Case 1: A metacarpal osteogenic sarcoma successfully treated with ray amputation.

A 20-year-old white female, on March 24, 1947, complained of pain the past three months in the dorsolateral aspect of her right hand. There was no history of recent trauma. Examination revealed some swelling and tenderness of her 4th and 5th metacarpals. X-ray films (Figure 1-A) showed periosteal bone formation involving the adjacent sides of the 4th and 5th metacarpals. All laboratory studies were within normal limits. On April 7, biopsy of the 4th and 5th metacarpals gave a pathologic diagnosis of osteogenic sarcoma (Figure 1-B). Four days later the patient had amputation of the 4th and 5th digits of her right hand, including the corresponding metacarpals. Her postoperative course was uneventful. A 10-year follow-up at yearly intervals up to September 16, 1957, found her in excellent health with essentially normal use of her right hand.

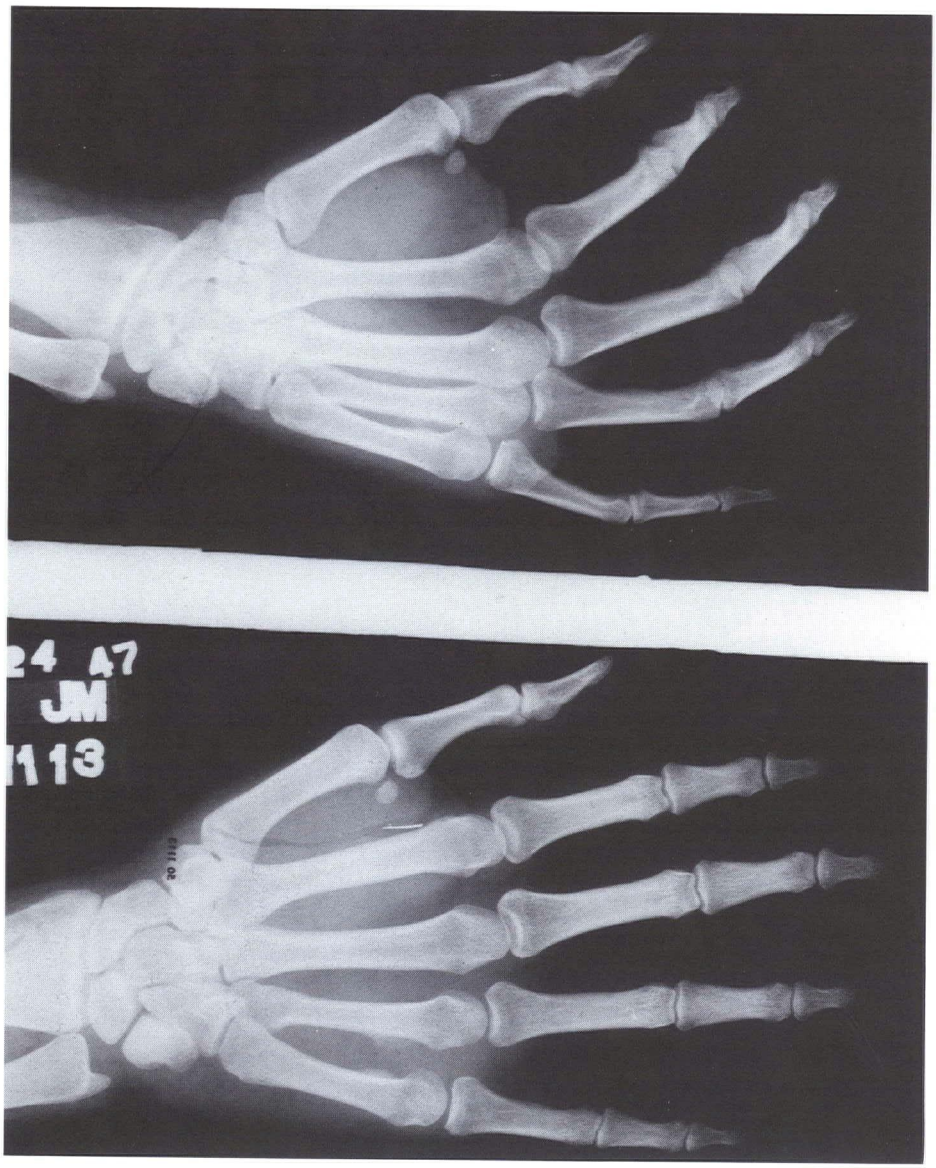

Figure 1-A
Roentgenograms of the hand show periosteal bone formation involving the distal portion of the adjacent sides of the fourth and fifth metacarpals.

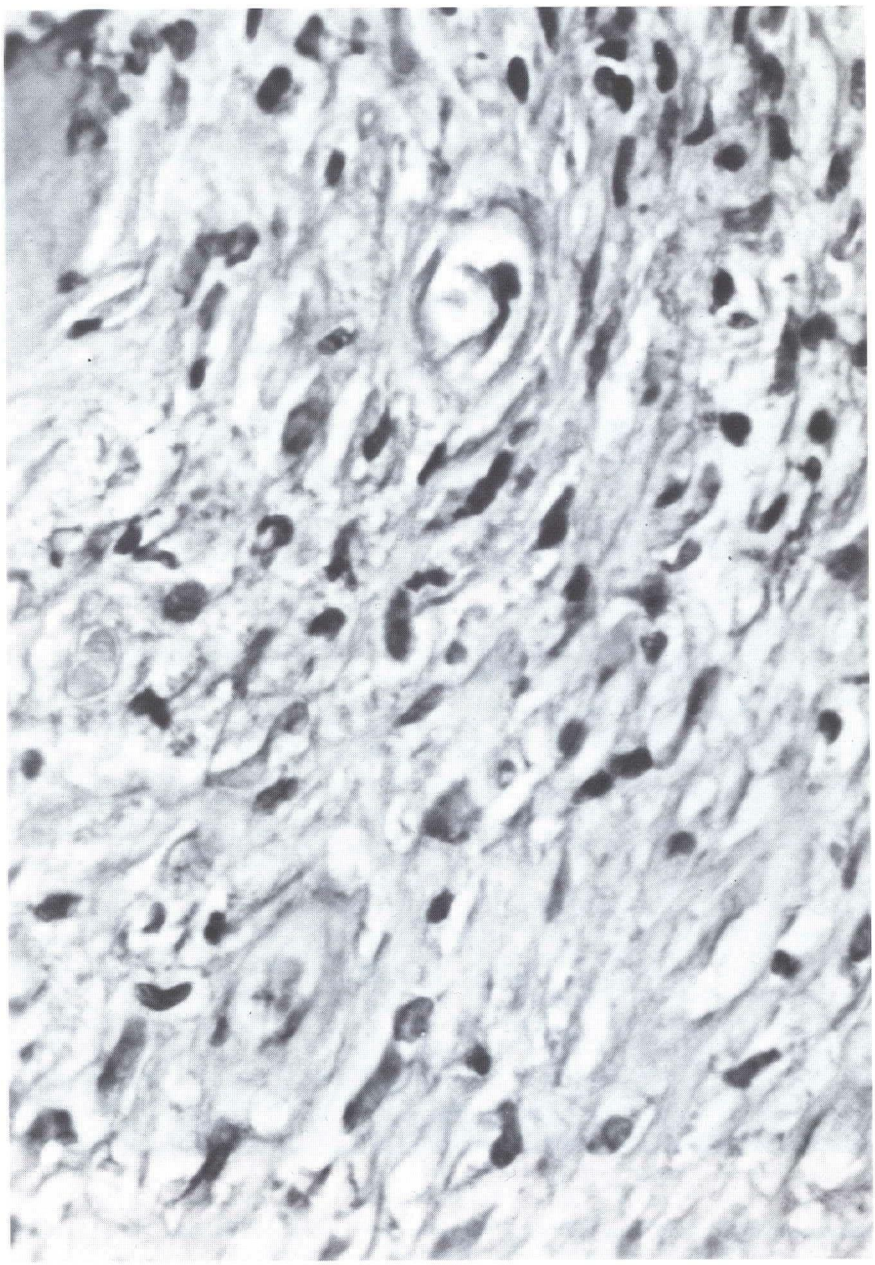

Figure 1-B
Photomicrograph (X 900) shows the pleomorphic and hyperchromatic cells intimately associated with the homogeneous, afibrillar tumor osteoid.

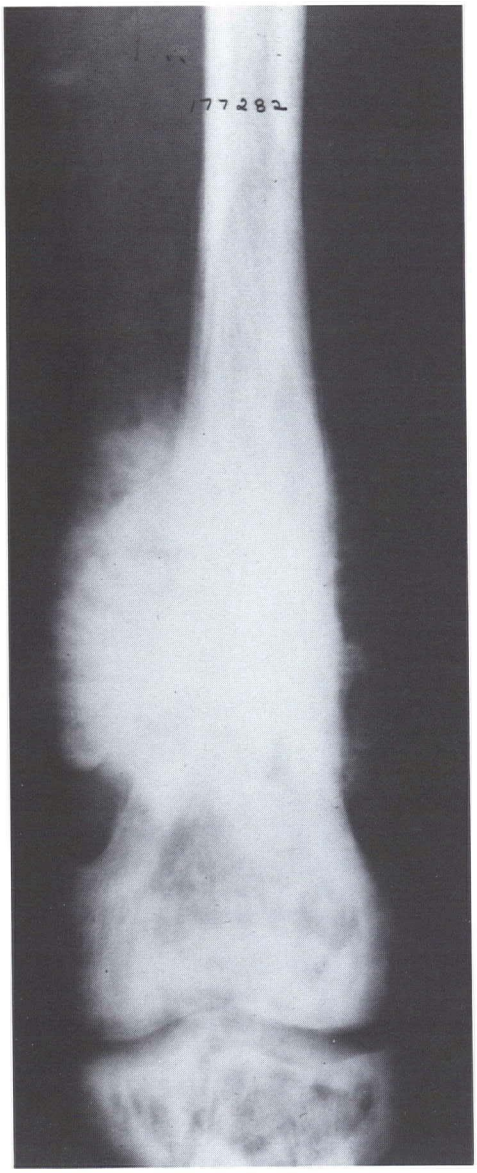

Figure 2-A

Roentgenogram of the femur shows the typical spotty demineralization, sclerosis and coarse trabeculation of Paget's disease. Also note the densely radiopaque tumor with the so-called "sunburst" appearance involving the lower third of the femur.

Case 2. A rare case of Pagetoid osteogenic sarcoma of the femur with metastasis to the ulna.

A 58-year-old white female, who had been known to have Paget's disease of her right femur and tibia for many years, was seen at Henry Ford Hospital on June 6, 1932, with the chief complaint of increasing pain and swelling of the right thigh the past several months. Examination revealed a very hard tumor, measuring 10 cm x 7.5 cm, which appeared to be firmly attached to the medial aspect of her right distal femur. X-ray photos (Figure 2-A) showed a large, densely radiopaque tumor involving the distal portion of the right femur. Three days later biopsy of the right femur revealed the presence of osteogenic sarcoma (Figure 2-B). On June 15, 1932, the patient underwent a high thigh amputation under general anesthesia. The following September, the patient was seen at our hospital again because of pain and swelling of her right forearm. Examination revealed a grossly swollen right forearm and a bony hard circumferential swelling of her proximal radius.

X-ray film (Figure 2-C) showed destruction of the radius with transverse radiopaque "sunburst" streamers extending into the adjacent soft tissues. Chest x-rays also showed the presence of several radiopaque nodules in both lungs. From then on, the patient's health rapidly deteriorated and she died three months later from extensive visceral and skeletal metastases.

Case 3. An extraosseous osteogenic sarcoma of the thigh which a hindquarter amputation failed to eradicate.

A 66-year-old white female was seen at our outpatient clinic on August 18, 1968, with the chief complaint the past 6 months of pain and swelling in her right thigh. Examination revealed a very large fusiform soft tissue mass which seemed to involve nearly the entire posteromedial aspect of her right thigh. X-ray photos (Figures 3-A and 3-B) showed a greatly enlarged soft tissue shadow of the posteromedial aspect of the right thigh. The right femur was not involved. Subsequent arteriograms (Figure 3-C) showed displacement of the femoral artery by the soft tissue tumor and the proliferation of the terminal branches of the right femoral artery in the tumor. Laboratory studies were all within the normal limits and radiological metastatic survey showed no evidence of metastases. On August 26, the patient's right thigh was biopsied. The pathologic diagnosis was extraosseous osteogenic sarcoma (Figures 3-D and 3-E). On September 3, the patient underwent a right hindquarter amputation. The surgical wound healed per primum.

Followed as an outpatient for two years after discharge from our hospital, the patient seemed to do fairly well until September 1970 when she started having constant pain in her right shoulder. X-ray photos taken on October 3, 1970, showed a

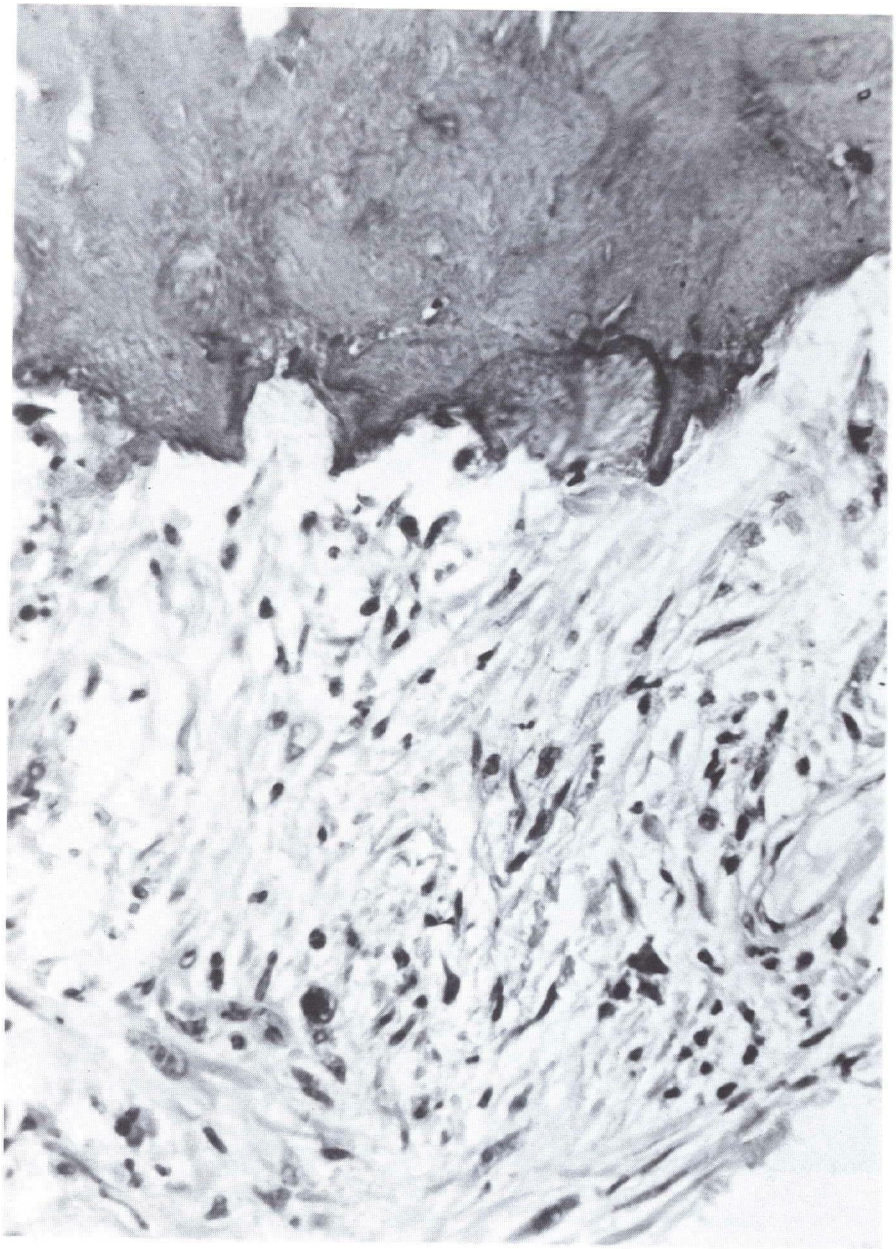

Figure 2-B

Photomicrograph (X 510) of a Pagetoid osteogenic sarcoma shows the dense cement lines and mosaic pattern of Pagetoid bone on one side and the malignant osteoblasts and the tumor osteoid on the other side.

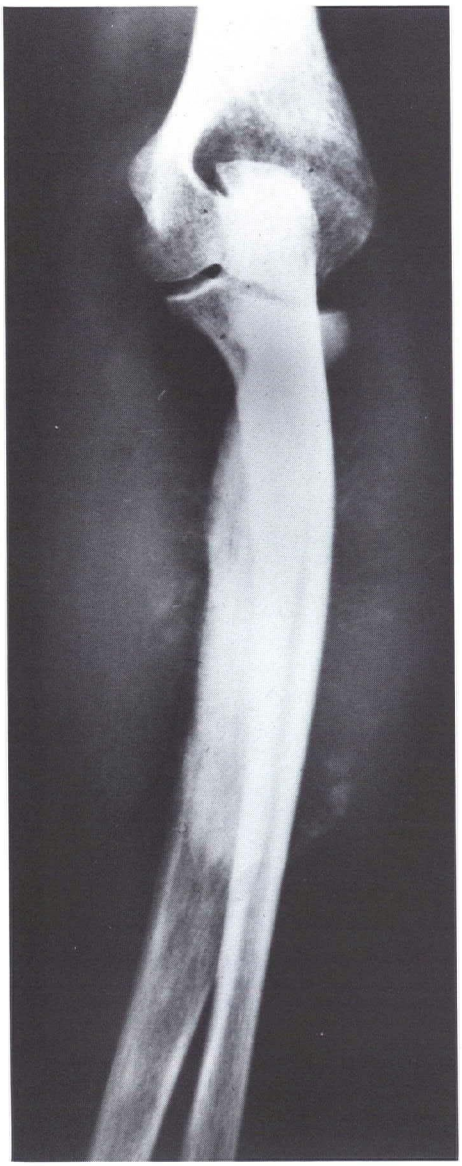

Figure 2-C

Roentgenogram of the forearm shows some cortical destruction of the proximal radius and the vertical "sunray" streamers extending from radius into the adjacent soft tissues.

destructive lesion of her left proximal humerus and several radiopaque nodules in her lung. The left humerus was given 3,000 rads of palliative radiotherapy which provided some pain relief. However, in spite of her radiotherapy and other supportive treatments, the patient's health rapidly deteriorated and she died on November 28, 1970, from widespread visceral and skeletal metastases.

Case 4. An osteolytic osteogenic sarcoma of the femur which caused pathological fracture and subsequent extensive destruction of the femur.

A 58-year-old white male with a four-month history of pain in his left leg was brought to our emergency room by ambulance on May 21, 1951, because of acute severe pain in his left leg and inability to bear any weight on the leg. X-ray photos taken at the emergency room showed a pathological fracture of his left distal femur. After admission to the hospital, subsequent chest x-rays revealed evidence of pulmonary metastases. To relieve pain and facilitate nursing care, the femoral fracture was stabilized with an intramedullary Hanson-Street rod (Figure 4-A). The tumor was also biopsied during the intramedullary rodding. The pathologic diagnosis was chondroblastic osteogenic sarcoma (Figure 4-B). The patient was treated with radiotherapy. Over the next five months, the femur was rapidly destroyed by the tumor until it almost completely disappeared (Figure 4-A). The patient finally died on November 25, 1951, from widespread pulmonary metastases.

Case 5. A secondary osteogenic sarcoma originating in a postradiation osteochondroma.

This 29-year-old white male had an osteochondroma excised from his right pubic bone when he was 18 years of age. Postoperatively, the right pubic bone was also treated with high dose of radiation. The patient was first seen at our hospital on May 10, 1949, with the chief complaint of several months of increasing pain and swelling in his right groin region. Examination revealed a football-size tumor which appeared to have originated in the pubic bone and then extended outward and downward to involve the adjacent soft tissues and the medial aspect of the right proximal femur. Irregular radiopacity was scattered throughout the tumor.

Subsequent biopsy revealed the presence of osteogenic sarcoma (Figure 5-B). No ablative surgery was attempted because the patient already had radiological evidence of pulmonary metastases. The patient's general condition ran a progressive downhill course and he expired in October, 1949. Autopsy revealed extensive visceral metastases.

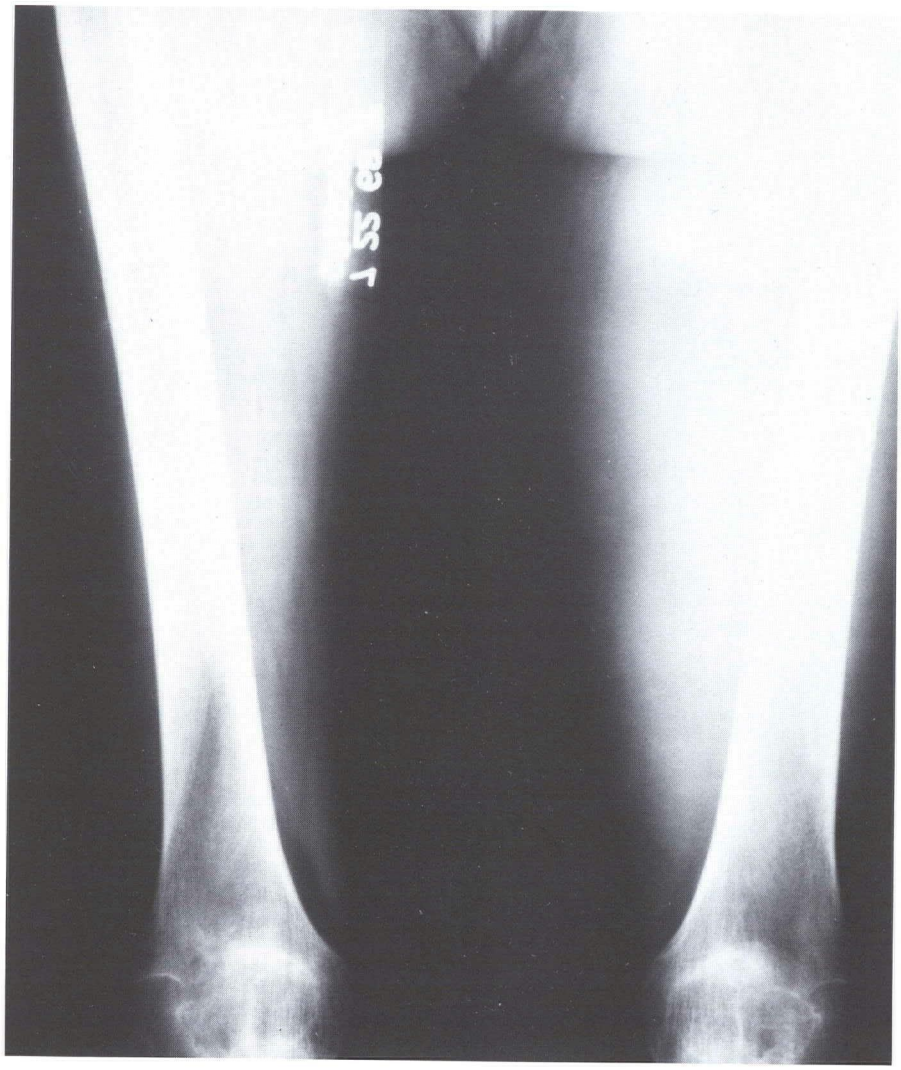

Figure 3-A
A-P view of both thighs shows a greatly enlarged soft tissue shadow which appears to involve the entire medial aspect of the right thigh.

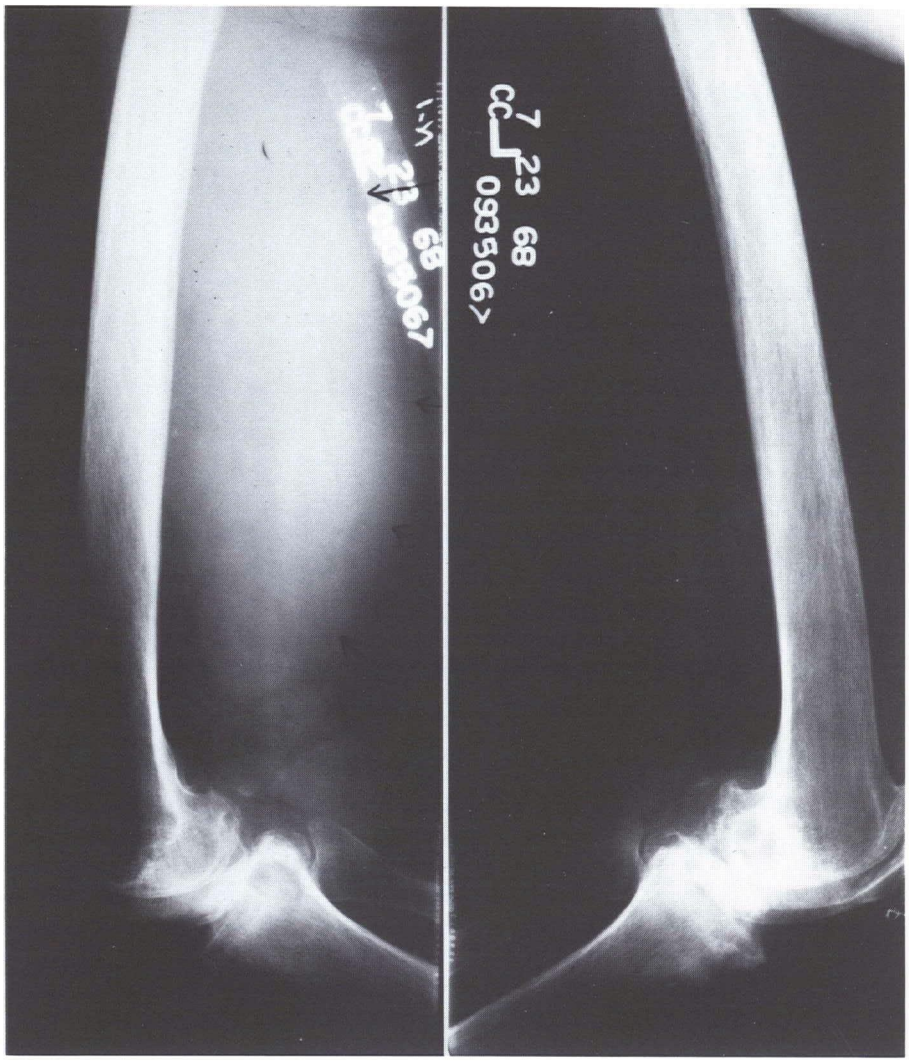

Figure 3-B
Lateral views of both thighs shows the huge fusiform enlargement of the soft tissue in the posterior aspect of the right thigh.

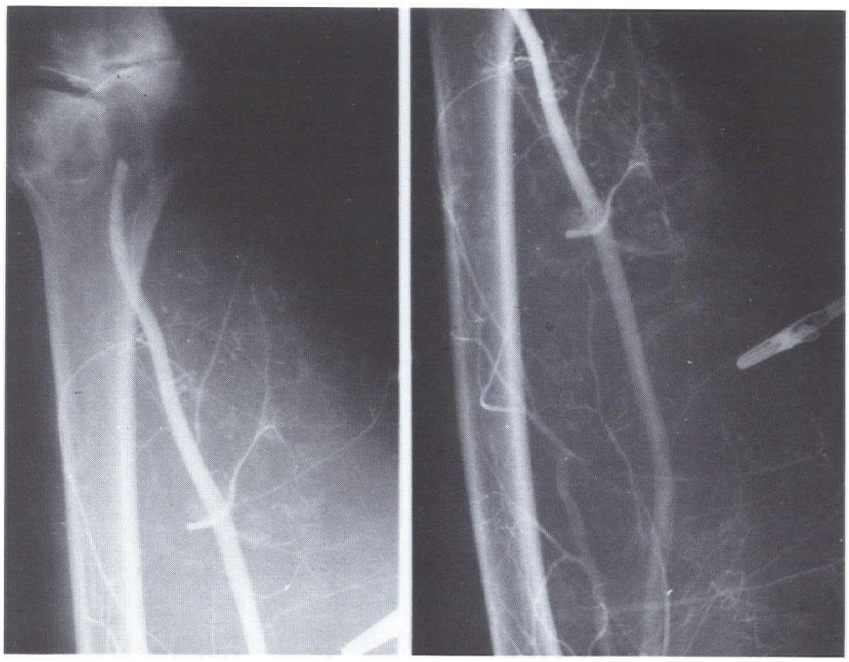

Figure 3-C
Arteriograms of the right thigh show displacement of the right femoral artery by the soft tissue tumor and the proliferation of the terminal branches of the femoral artery in the tumor.

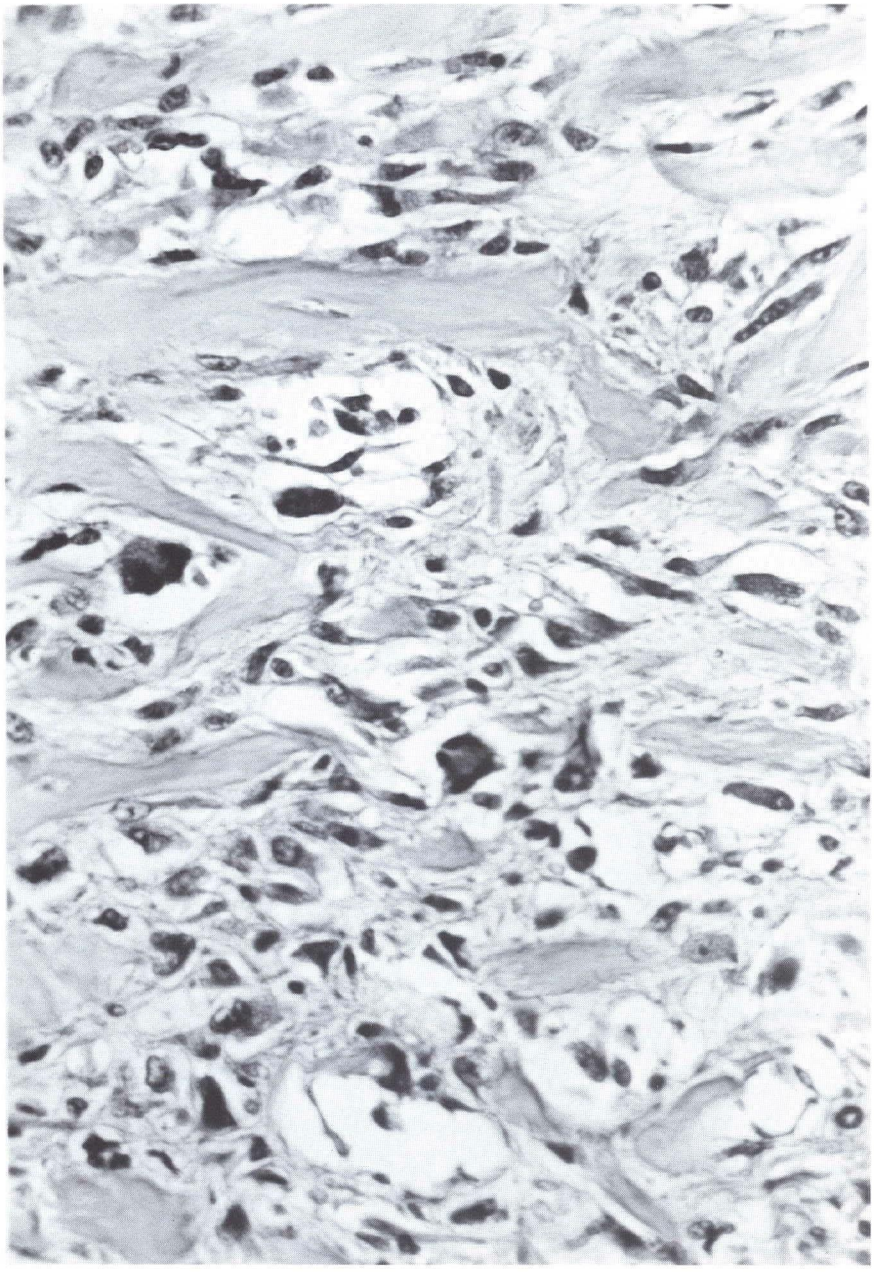

Figure 3-D
Photomicrograph (X 450) shows many malignant connective tissue cells actively producing tumor osteoid.

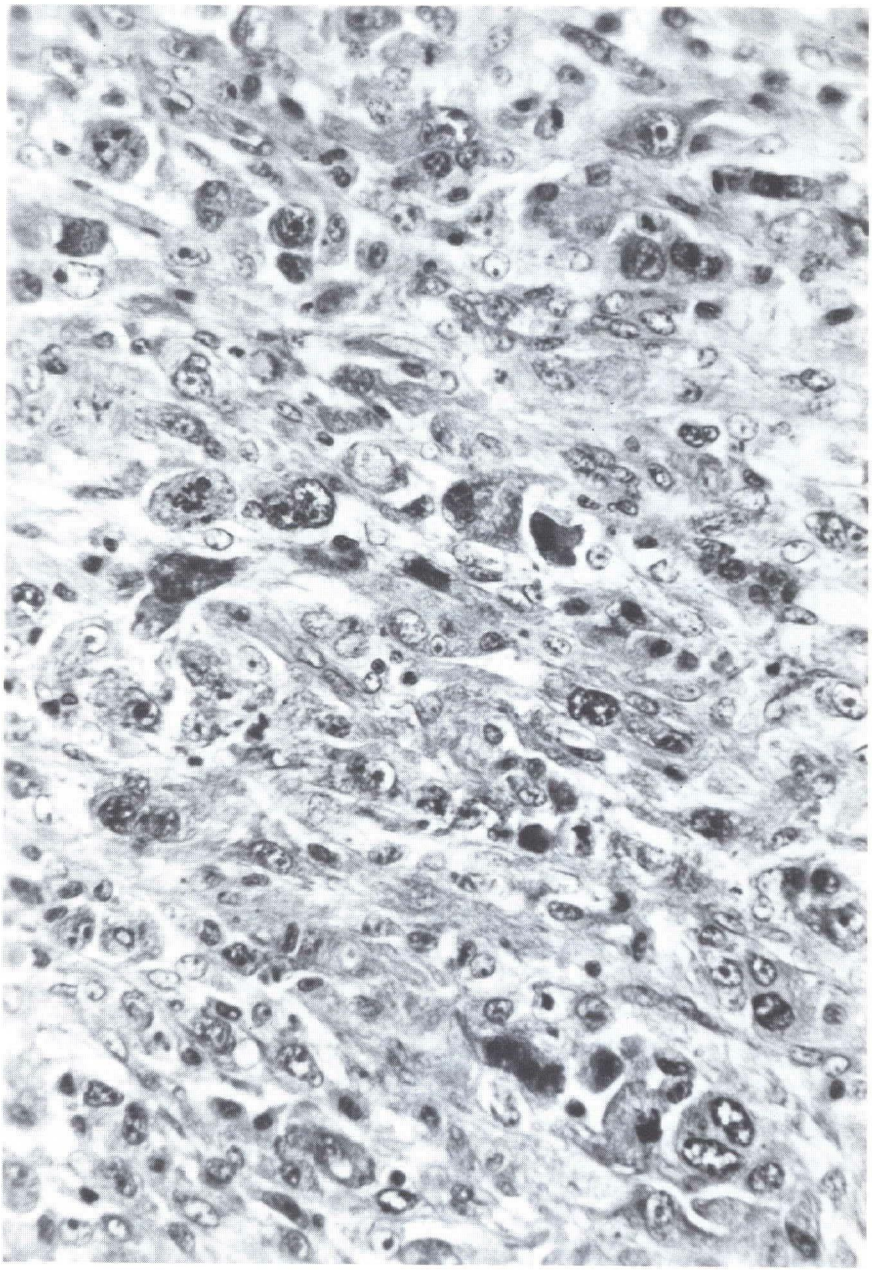

Figure 3-E
Photomicrograph (X 450) from a different part of the same tumor shows many extremely malignant-looking, pleomorphic and bizarre cells plus many mitotic figures.

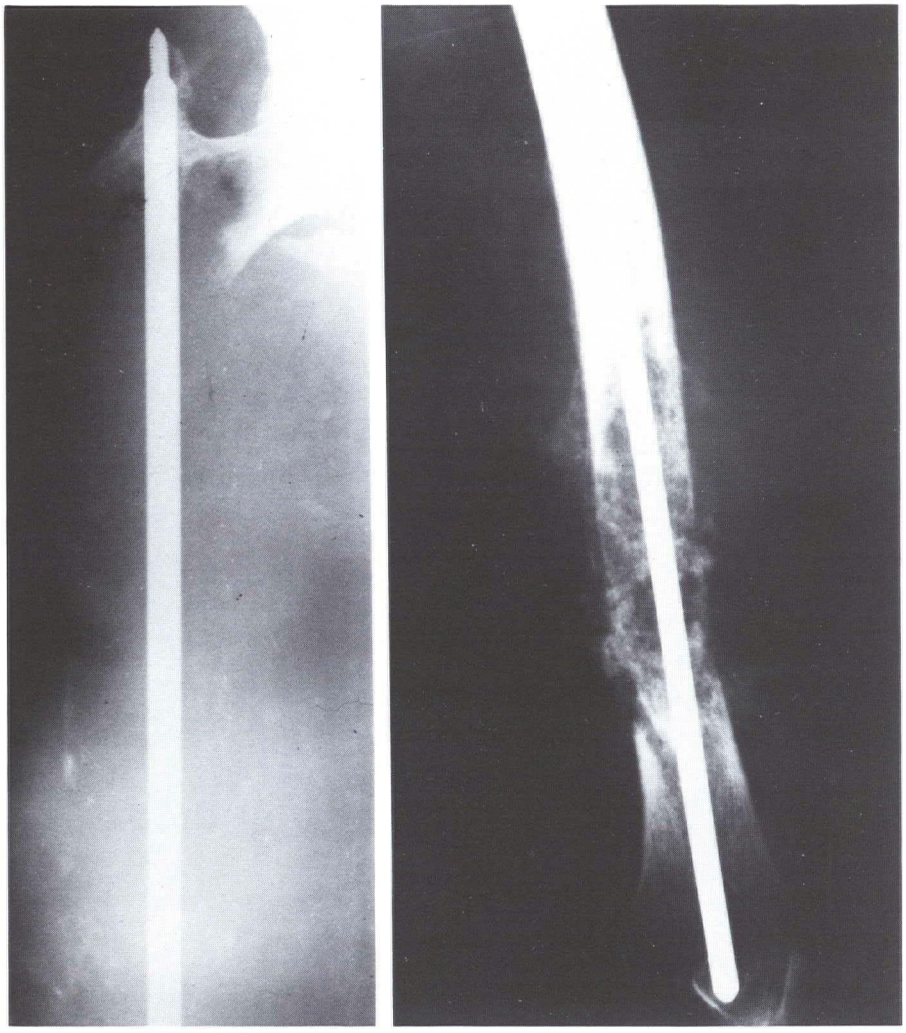

Figure 4-A

The roentgenogram on the right shows periosteal reaction and diffuse bone destruction of the distal femur. The pathological fracture has been stabilized by the intramedullary rod. The roentgenogram on the left shows nearly complete destruction of the whole femur.

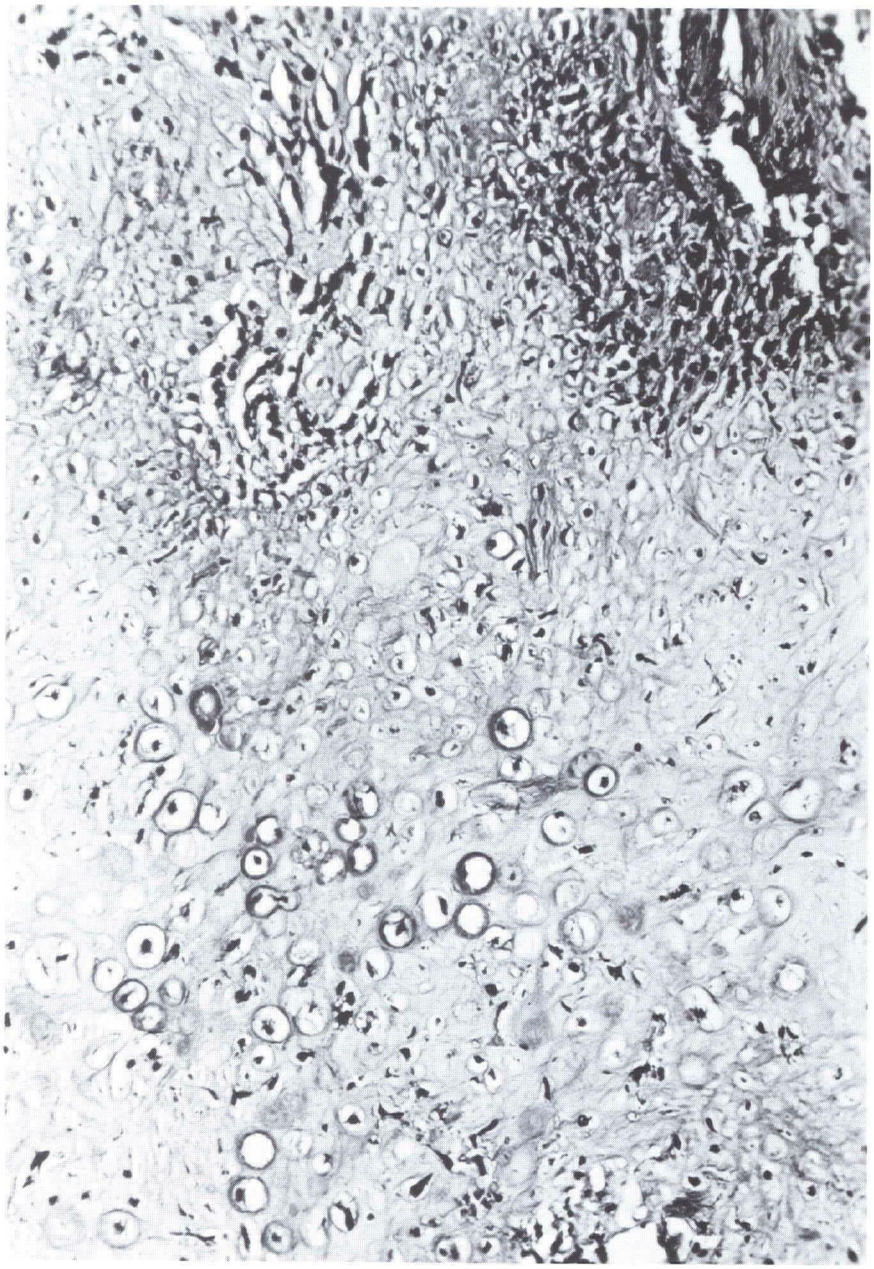

Figure 4-B
Photomicrograph (X 175) of a chondroblastic osteogenic sarcoma shows many extremely bizarre, pleomorphic and hyperchromatic cells. Quite a few of them are contained in lacunae.

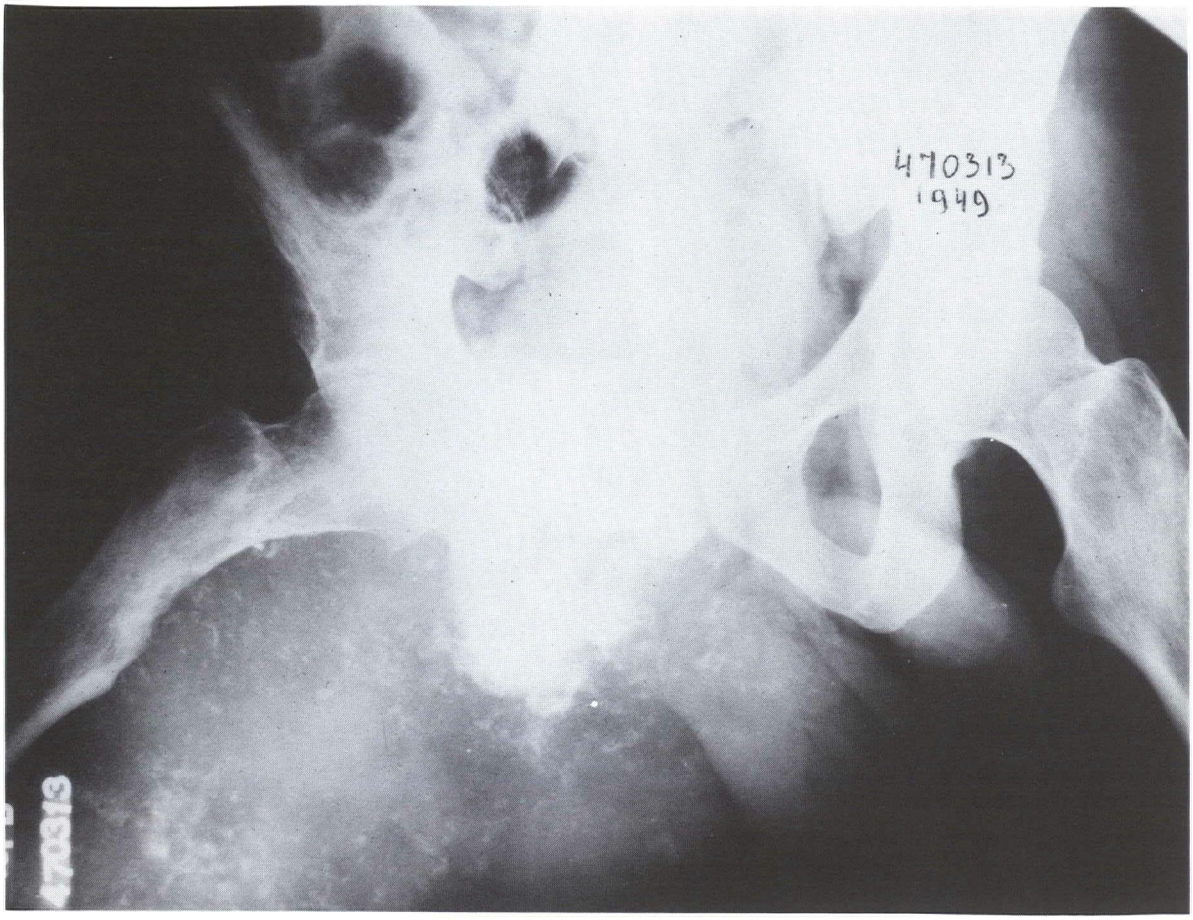

Figure 5-A

Roentgenogram of the pelvis shows a large radiopaque tumor with many foci of irregular calcification. Note the pubic bone involvement and the cortical erosion of the medial aspect of the right proximal femur.

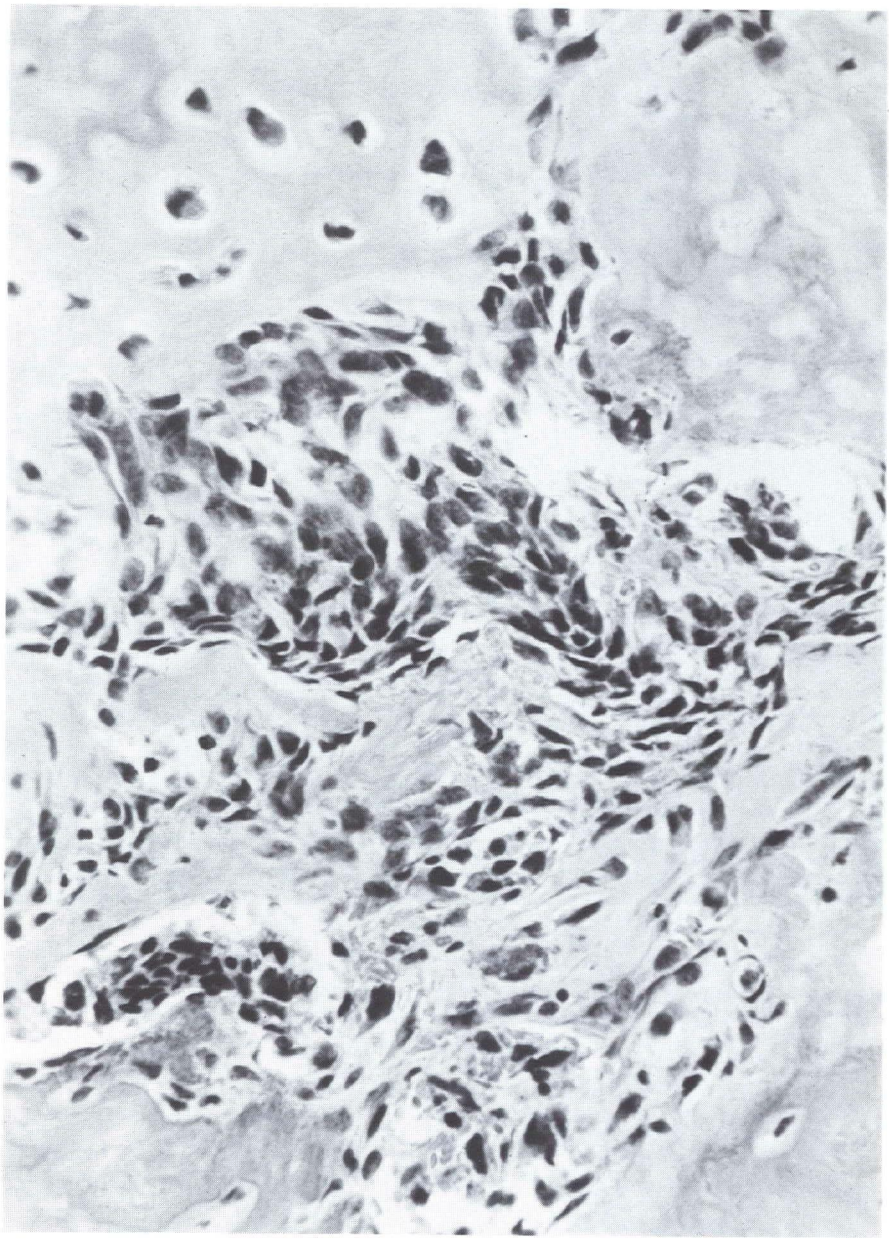

Figure 5-B
Photomicrograph (X 450) shows cords of malignant osteoblasts lining the newly formed tumor osteoid. Also observe the plump cells contained in lacunae.

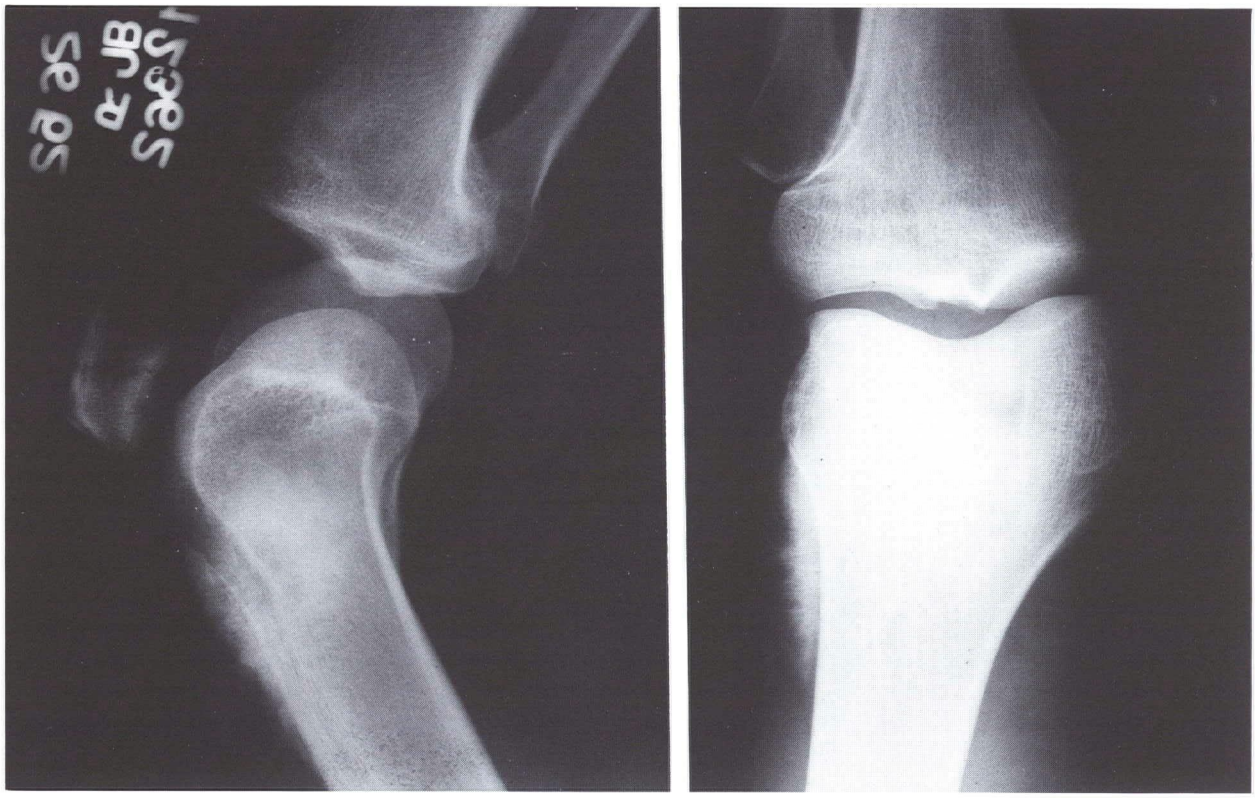

Figure 6-A
Roentgenograms of the knee show the sun-ray appearance of the periosteal bone formation. Note the complete absence of any intramedullary involvement.

Case 6. A parosteal (juxtacortical) osteogenic sarcoma of the femur cured by a mid-thigh amputation.

This 31-year-old black male was seen at our outpatient department on February 5, 1953, complaining of pain for the past 5 months in the medial aspect of his right knee. Examination revealed a hard mass about the size of an egg which appeared to be firmly attached to the medial supracondylar area of the right femur. X-ray photos (Figure 6-A) showed a brush-like radiopaque tumor occupying the medial supracondylar region of the right femur. The tumor appeared confined to the periosteal surface of the femoral cortex with no intramedullary involvement. All laboratory studies were within normal limits.

Biopsy of the femoral tumor three days later confirmed our clinical impression of parosteal osteogenic sarcoma (Figure 6-B). On March 2, 1953, the patient underwent a mid-thigh amputation of his right leg. His postoperative course was uneventful. Followed on an annual basis, the patient was last seen on October 31, 1972, approximately 19½ years after amputation. He wore a prosthesis and was in excellent health.

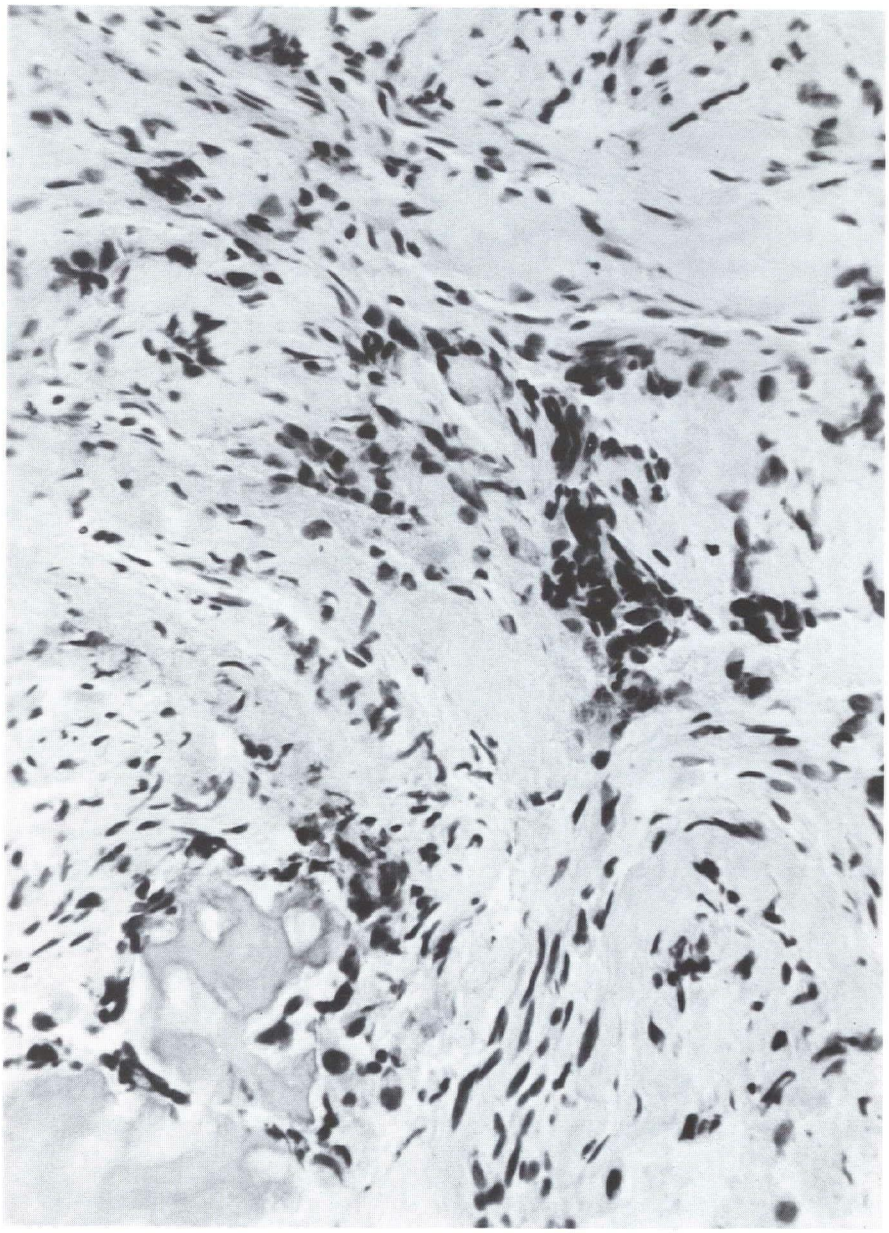

Figure 6-B
Photomicrograph (X 540) shows a fibrous stroma with moderate degree of nuclear pleomorphism. Note the immature bone formed by the stromal cells.

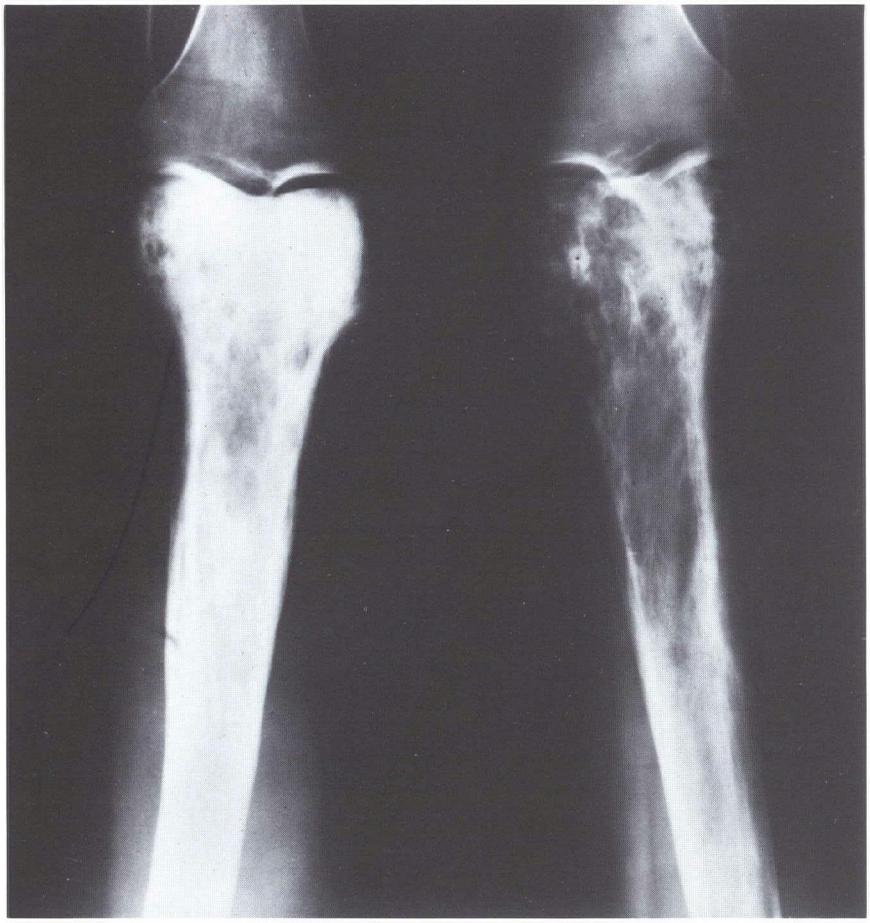

Figure 7-A

Roentgenogram of both femora shows the typical skeletal changes of Paget's disease. Also observe the large osteolytic lesion involving the medial supracondylar area of the left femur.

Case 7. Multiple osteogenic sarcomas complicating a case of polyostotic Paget's disease.

This 46-year-old white female had been known to have polyostotic Paget's disease of both her femora and right ilium for about seven years. She was seen at our outpatient clinic on October 25, 1970, complaining of pain and swelling of her left knee during the preceding three months. Examination revealed some diffuse tenderness in the medial supracondylar region of her left femur. X-ray photos (Figure 7-A) showed destruction of the medial supracondylar area in addition to the typical skeletal changes of Paget's disease. Laboratory studies showed serum alkaline phosphatase 17.7 (Bodansky units), calcium 10.5, phosphate 3.3, hemoglobin 13.2, and white blood cell count 5,500.

On October 30, 1970, a biopsy of the patient's left distal femur was performed. Pathologic diagnosis was osteogenic sarcoma complicating Paget's disease (Figure 7-B). On November 6, the patient underwent a left hip disarticulation. Postoperatively, the serum alkaline phosphatase level dropped to 7.2 (Bodansky units). Followed as an outpatient, she was brought to our emergency

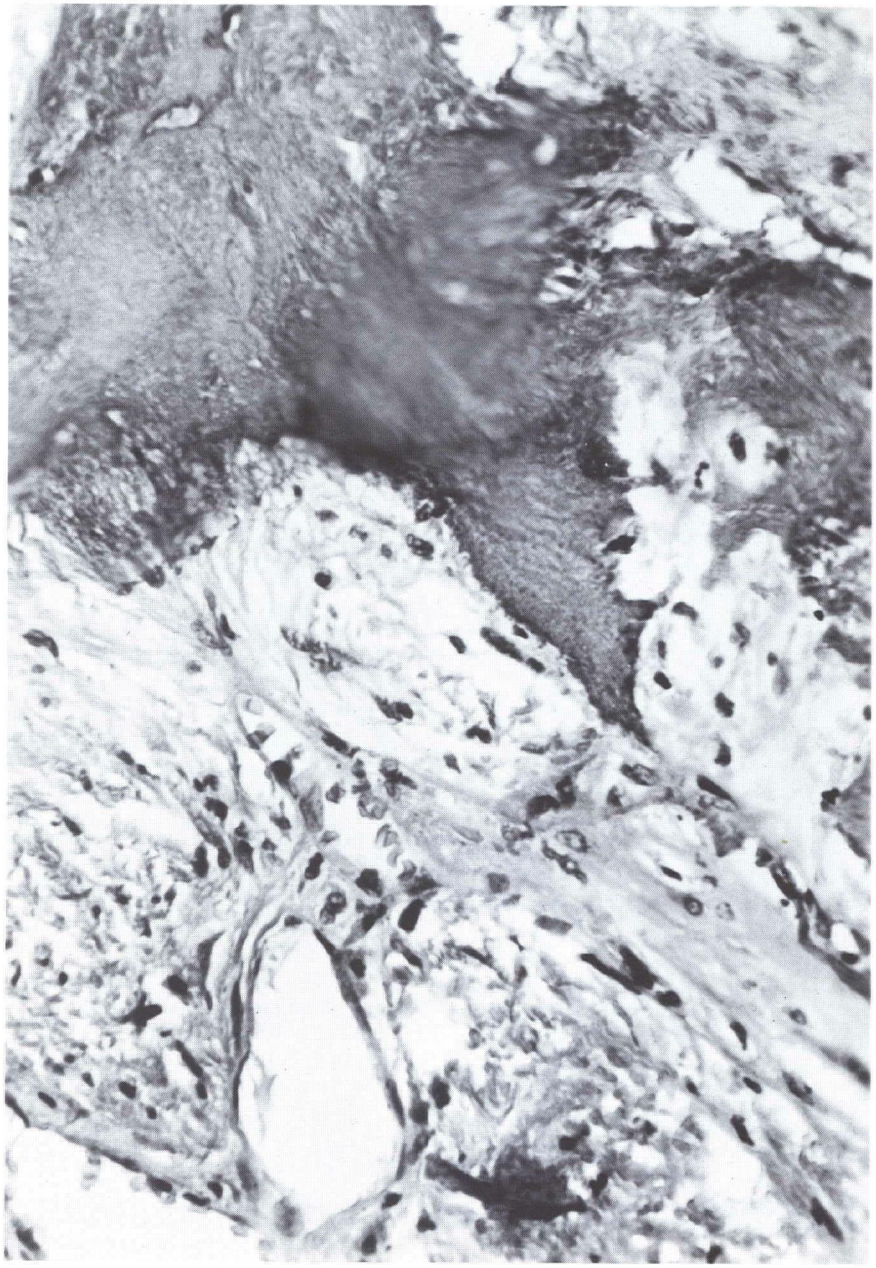

Figure 7-B
Photomicrograph (X 510) shows Pagetoid bone on one side and the hyperchromatic malignant cells and the homogeneous, afibrillar tumor osteoid on the other.

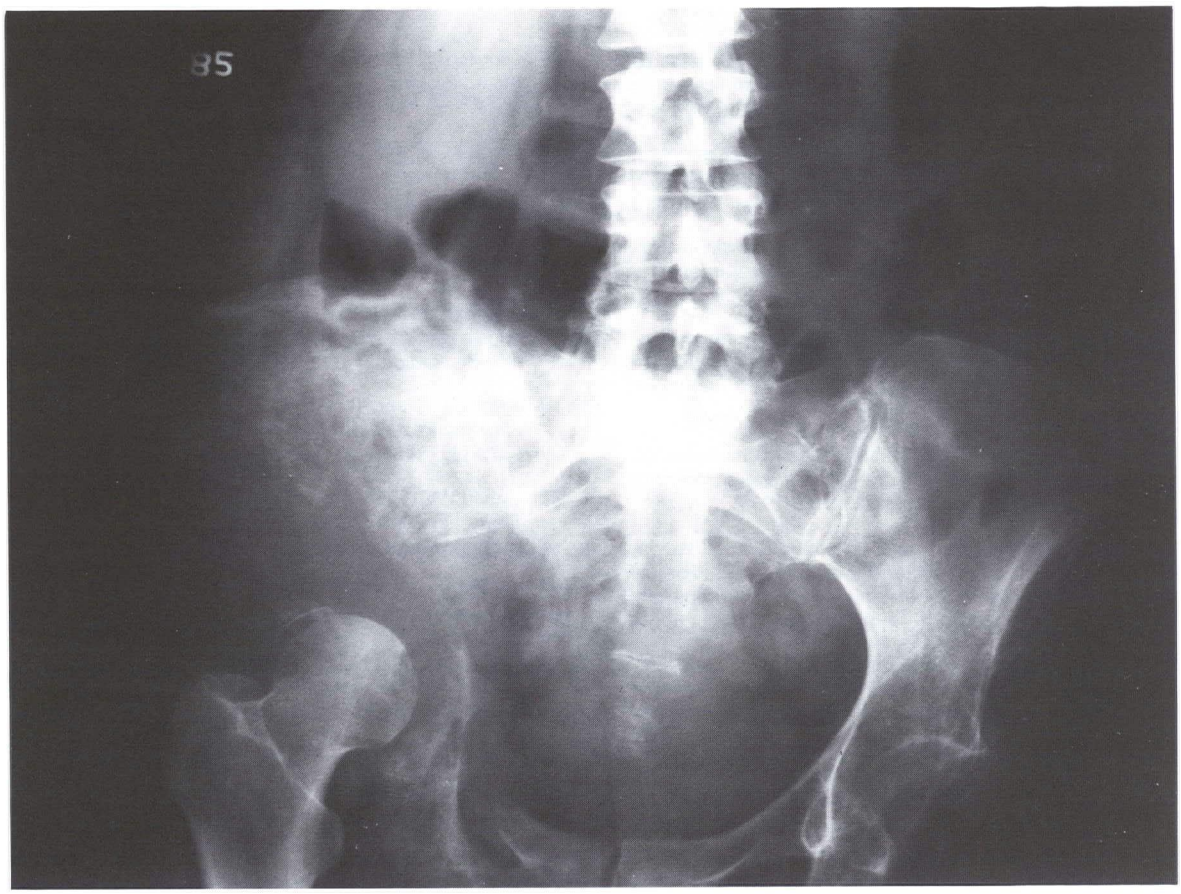

Figure 7-C

Roentgenogram of the hips shows extensive destruction of the right acetabulum and ilium on one side and the absence of the left femur on the other.

room by ambulance on June 22, 1971, because of severe pain in her right hip. The pain had been rapidly progressive for several weeks. Examination revealed a swollen and very irritable right hip. X-ray photos (Figure 7-C) showed extensive destruction of her right acetabulum and ilium. The patient also had radiological evidence of pulmonary metastases. The serum alkaline phosphatase was repeated and found to be elevated to 16.2 (Bodansky units). The right ilium was treated with 6,000 rads of palliative radiotherapy which brought about a drop of serum alkaline phosphatase to 7.2 (Bodansky units). However, in spite of radiotherapy, chemotherapy and other supportive measures, this patient's metastases continued to grow and spread and she finally expired on February 24, 1972, from widespread metastases. Autopsy also revealed the presence of Pagetoid osteogenic sarcoma in her right distal femur.

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