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Chondrosarcoma: A Report of 65 Cases

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A statistical analysis is presented of 65 cases of chondrosarcoma treated at Henry Ford Hospital from 1916 to 1976, with adequate follow-up data.

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Introduction

CHONDROSARCOMA is a relatively common, malignant, primary bone tumor, surpassed in frequency only by multiple myeloma and osteogenic sarcoma.

Chondrosarcoma usually originates in the large long bones (femur, tibia, humerus, etc.) or in the large flat bones (ilium, scapula, etc.).[1-8] Orthopaedic surgeons are frequently involved in treating this tumor, since surgical removal is still the treatment of choice.

This paper presents a statistical analysis drawn from 65 cases of chondrosarcoma treated at Henry Ford Hospital from 1916 to 1976, with adequate follow-up data. These include 54 cases of primary chondrosarcoma (a malignant cartilaginous tumor arising *de novo* in a particular site),[9-16] four of secondary chondrosarcoma (a malignant cartilaginous tumor arising in a preexisting bone or soft tissue lesion),[7-21] five of extraskeletal chondrosarcoma,[22-27] one of synovial chondrosarcoma,[28-32] and one of juxta-cortical (periosteal) chondrosarcoma.[13-14, 33-35] (See Table I)

Of the 65, the 54 cases of primary chondrosarcoma and the four cases of secondary chondrosarcoma are discussed in the following analysis. The remaining cases of extraskeletal, synovial, and periosteal chondrosarcoma were excluded because of their rare occurrence and unpredictable clinical behavior.

Clinical Features

Race, sex, and age distribution

There were 51 white and seven black patients, 31 of whom were men and the remaining 27 women. The youngest patient was 8 years old and the oldest 87 (Table I).

TABLE I

Type of Chondrosarcoma	Total Cases	Age	Sex	Location	Treatment	Results
Primary	54			See Figures 1 and 2		
Secondary	4	23	F	Left Distal Femur	Hip Disarticulation	Alive and Well 5 Years Later
		47	F	Proximal Phalanx of Right Index Finger	Ray Amputation	Alive and Well 10 Years Later
		46	M	Left Distal Femur	Hip Disarticulation	Died 8 Months Later from Metastases
		23	M	Left Proximal Femur	Hemipelvectomy	Died 6 Months Later from Metastases
Extraskeletal	5	87	M	Left Hand	Radical Excision Plus Irradiation	Died 17 Months Later from Metastases
		55	M	Right Vastus Medialis Muscle	Removal of Vastus Medialis Muscle	Alive and Well a Year Later
		28	F	Mediastinum	Radical Excision	Lost to Follow-up
		20	M	Left Vastus Lateralis Muscle	En-bloc Excision of Vastus Lateralis Muscle	Alive and Well a Year Later
		69	F	Right Thigh	Radical Excision Plus Irradiation	Died 8 Months Later from Metastases
Synovial	1	33	M	Right Knee	Above Knee Amputation	Alive and Well 7 Years Later
Periosteal	1	56	M	Right Thumb	Amputation of Thumb	Died 2 ¾ Years Later from an Automobile Accident

Tumor location

The femur, tibia, ilium, and humerus in descending order account for over 65% of the chondrosarcomas in our series (Table 2).

Signs and symptoms

Pain and swelling were the most common complaints. In addition, chondrosarcoma can cause many other symptoms by simply interfering with the normal functions of adjacent tissues. For example, pathological fractures occurred in two femora, one humerus, and one calcaneus in this series. One distal femoral and two proximal tibial chondrosarcomas were accompanied by significant muscular atrophy, synovial effusion, and loss of range of motion of the knees. By obstructing the superior vena cava, the mediastinal chondrosarcoma caused dyspnea, cyanosis, and marked swelling of the head, neck, and both upper extremities. One large, recurrent chondrosarcoma of the ilium grew into the abdominal cavity and caused complete bowel obstruction. Another lumbar chondrosarcoma produced pain, paresthesia, and muscular weakness of the leg by impinging on the lumbar nerve roots.

Roentgenographic Manifestations

Most of our chondrosarcomas were metaphyseal tumors, although large chondrosarcomas did also involve the diaphysis.

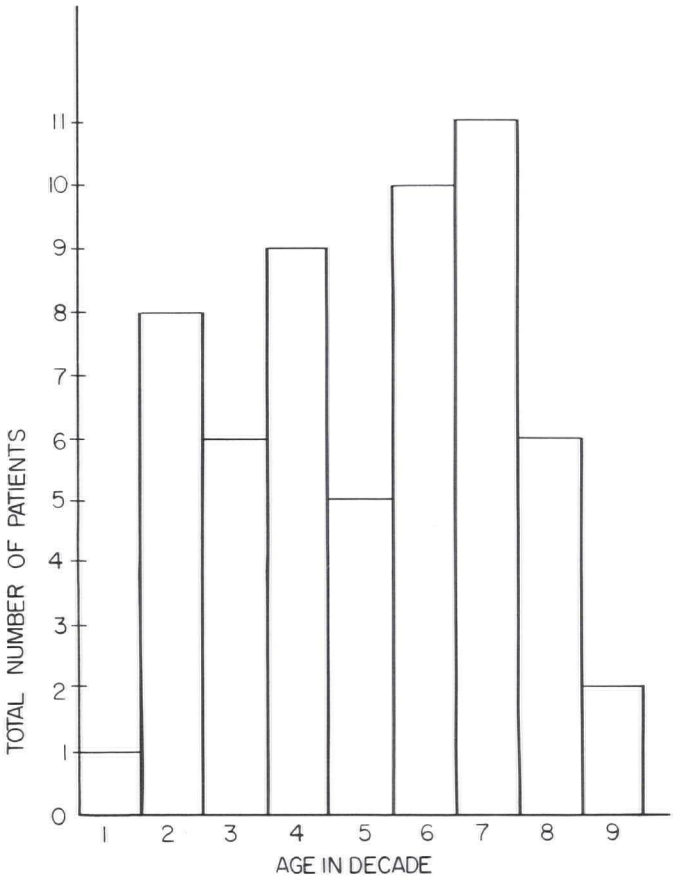

Fig. 1
Age distribution of 58 chondrosarcomas of bone

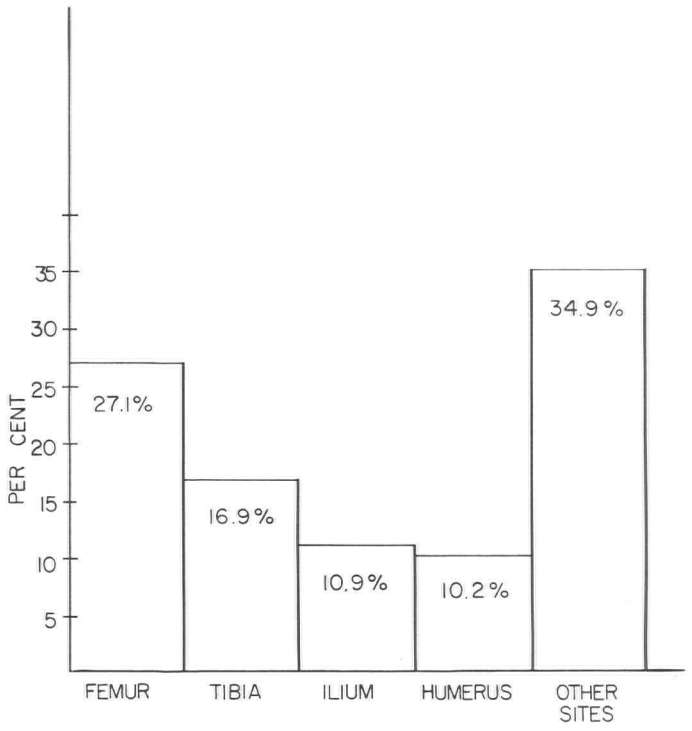

Fig. 2
Location of 58 chondrosarcomas of bone

When a chondrosarcoma is centrally located, trabecular and inner cortical destruction as well as outer cortical expansion, thickening, and sclerosis are the common radiological features (Figure 3). However, when a chondrosarcoma is peripherally located, the irregular calcification in its lobules frequently gives it a "cauliflower" appearance (Figure 4).

Pathology

1. Gross

The glistening, bluish-white appearance, the multi-lobulated configuration and the many foci of yellowish, chalky calcification identify it as a cartilaginous tumor. Foci of hemorrhage and cysts can also be seen (Figure 5).

2. Microscopic

It is our observation that chondrosarcoma can be differentiated from other benign cartilaginous lesions by its increased cellularity and mitotic figures, by the presence of pleomorphic, plump and hyperchromatic cells, by the frequent appearance of many double-nucleated lacunae, and by the existence of bizarre, giant cartilage cells (Figures 6 and 7).

Treatment

Before the advent of chemotherapeutic agents, inoperable primary and metastatic chondrosarcomas were treated with a tumor dose of irradiation of approximately 5,000 rads each. The same tumors are now treated with both irradiation and chemotherapy. Three patients in our series refused surgery and were treated with combined radiation and systemic chemotherapy. Unfortunately, they all succumbed to their disease within a short time. Of the three patients who underwent pulmonary resection for a well-localized pulmonary metastasis, only one of them is still alive more than two years after his lobectomy.

Over 90% of our patients had amputation or radical resection of one kind or other. The amputation was usually performed through the normal tissues proximal to the joint of the bone containing the chondrosarcoma (e.g., above-knee amputation for tibial or fibular lesion; hip disarticulation for distal femoral lesion; hindquarter amputation for proximal femoral lesion; above-elbow amputation for radial or ulnar lesion; shoulder disarticulation for distal humeral

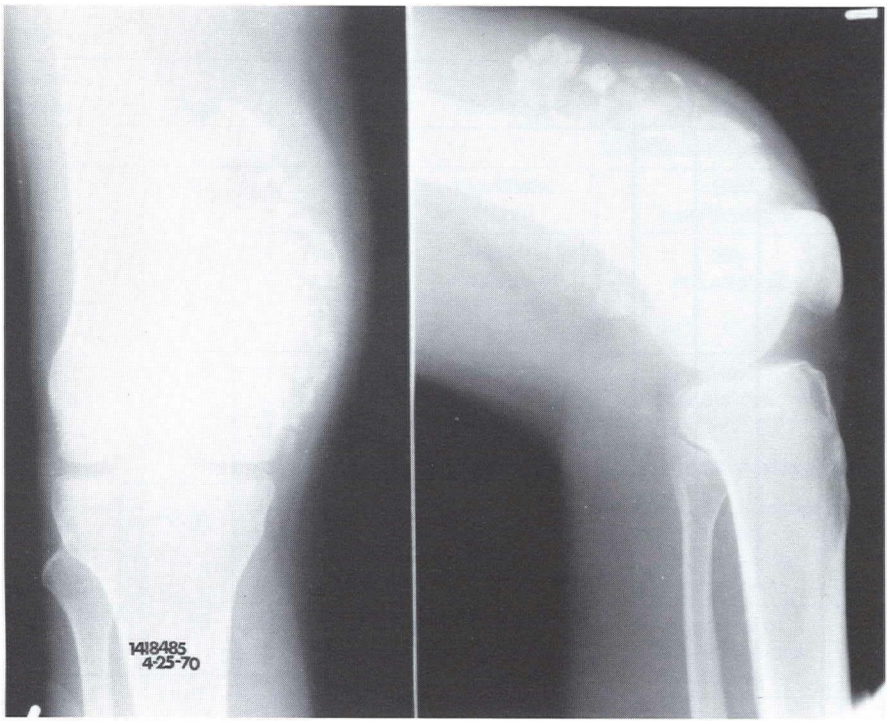

Fig. 4
Typical peripheral chondrosarcoma of the femur demonstrating the so-called "cauliflower" appearance of the tumors.

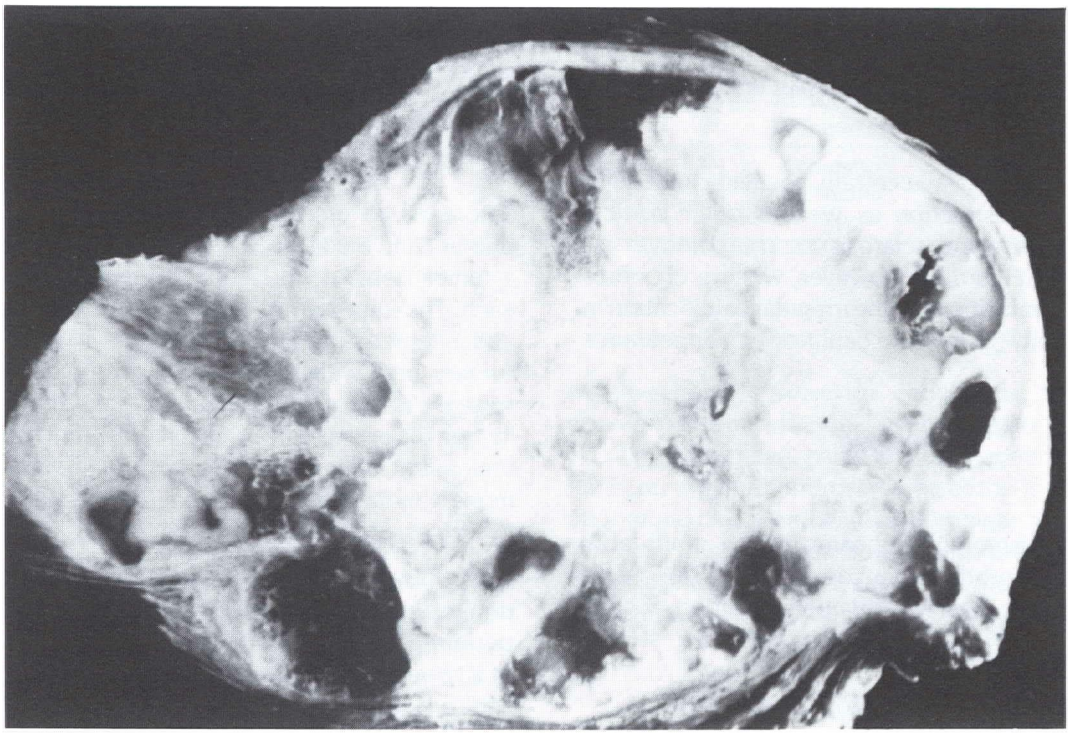

Fig. 5
The cross section of a large pelvic chondrosarcoma showing the glistening appearance, multilobulated configuration, and foci of chalky calcification, hemorrhage, and cysts.

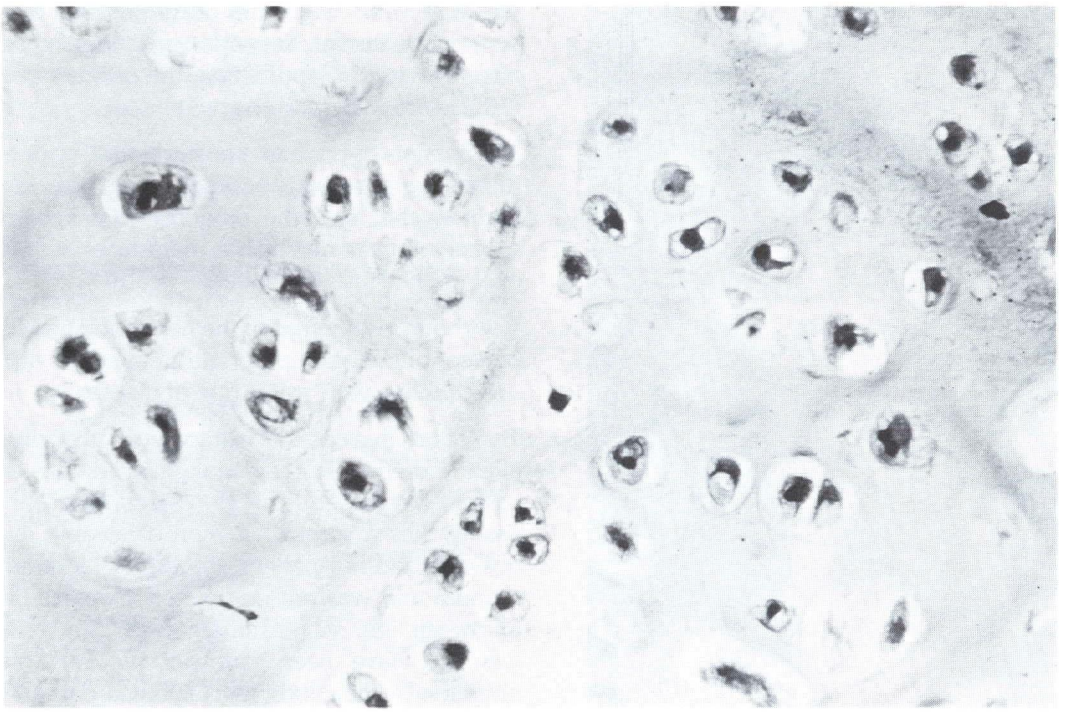

Fig. 6
Photomicrograph (X450, H & E) showing increased cellularity, plump and hyperchromatic nuclei, and several binucleated lacunae.

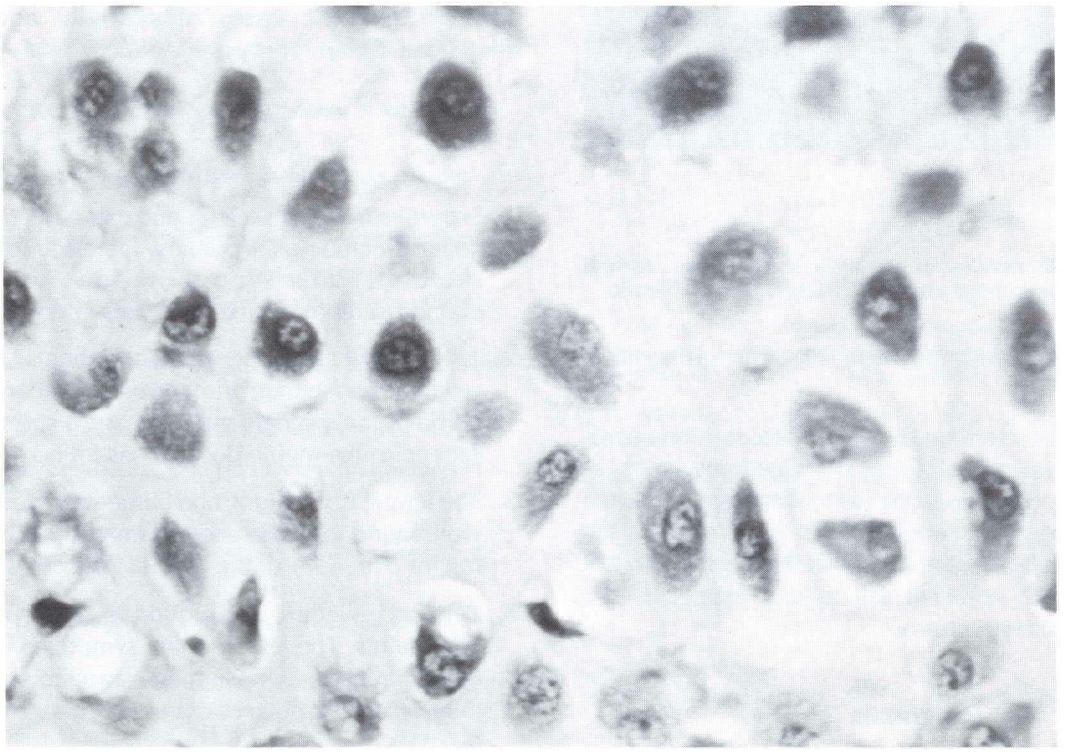

Fig. 7
Photomicrograph (X900, H & E) showing the large malignant cartilage cells with abundant cytoplasm which nearly fills the lacunae. Also note the large nuclei with prominent nucleoli.

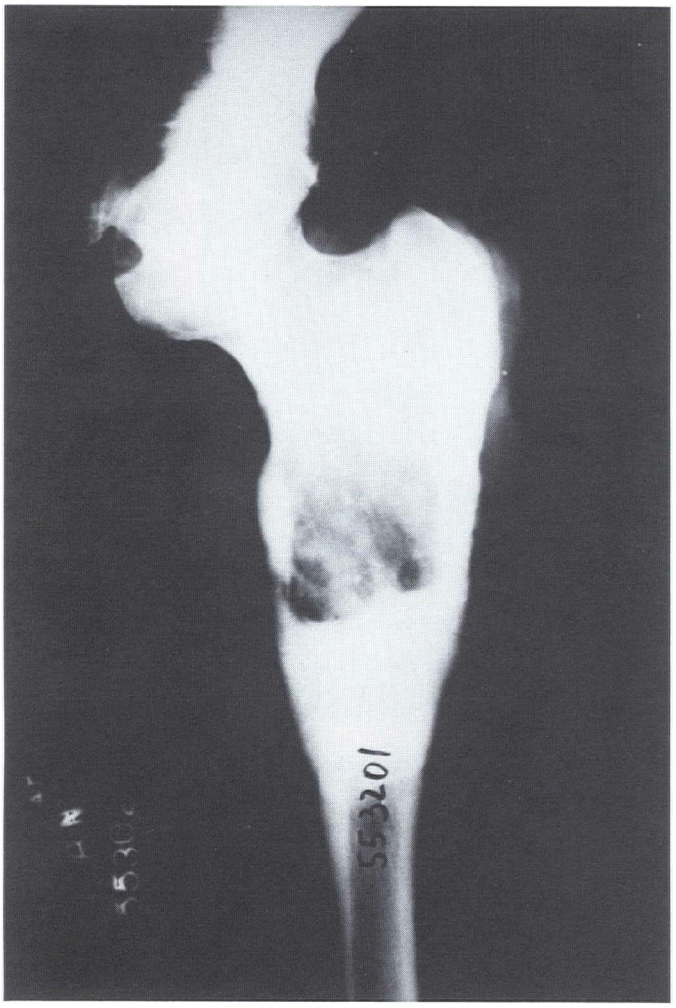

Fig. 3
Typical central chondrosarcoma of the femur showing inner cortical destruction and outer cortical expansion, thickening, and sclerosis.

lesion; and forequarter amputation for proximal humeral lesion).

However, at the present time, we prefer to combine chemotherapy with less radical surgical procedures, such as segmental resection followed by various methods of bone grafting or custom-made endoprostheses to replace the removed tumorous bones, provided that the tumors have not already invaded the adjacent soft tissues, especially the major neurovascular structures.

Prognosis

Follow-up periods of these 58 cases of chondrosarcoma ranged from one to 27 years. Thirty-seven patients (64%) died from their tumors within the first five years. Twenty-one patients (36%) were free of tumors five to 27 years after surgical treatment. Secondary chondrosarcoma seemed to have a worse prognosis than the primary chondrosarcoma (50% vs 36% five-year survival rate).

For patients who had constitutional symptoms, such as recent weight loss, weakness, malaise, fever, anorexia, night sweats, etc., the prognosis was very poor, because sooner or later detectable metastases would appear and eventually cause the patient's death.

The chondrosarcomas of the hands and feet had a favorable course following appropriate treatments; all patients have survived five to 17 years free of disease.

Two of our five cases of extraosseous chondrosarcoma have already died from their malignant disease, including one case of extraosseous chondrosarcoma of the hand, which usually has a somewhat more favorable long-term prognosis. In another patient, the tumor recurred in her mediastinum and was not completely excised. She was lost to follow-up and is presumed dead. Since two remaining survivors have been followed for only one year postoperatively, their long-term survival cannot be absolutely certain at the present time. These facts indicate that extraosseous chondrosarcoma appears to have a worse prognosis than skeletal chondrosarcoma.

We cannot draw any statistically significant conclusion about the single cases of synovial and periosteal chondrosarcoma.

Summary

1. Sixty-five cases of skeletal and extraskeletal chondrosarcomas, including 54 primary, four secondary, five extraskeletal, one synovial, and one periosteal chondrosarcoma, have been presented.
2. Fifty-one white and seven black patients (31 men and 27 women) had either primary or secondary chondrosarcoma. A fairly broad and even age distribution from the second to the eighth decade was found in this series.
3. The femur, tibia, ilium, and humerus in descending order were found to be the most common locations, accounting for over 65% of the chondrosarcomas in our series.
4. Pain and swelling were found to be the most common complaints. The less frequent symptoms include pathological fractures, effusion and impairments of adjacent joints, neurovascular compression, disuse muscular atrophy, etc.
5. Radical surgery was employed in most of the cases, but we now prefer segmental resection combined with

chemotherapy and bone grafting or endoprostheses. Irradiation and chemotherapy were reserved for inoperable chondrosarcomas or those with widespread metastases. Pulmonary resection for well-localized metastasis met with limited success.

6. Chondrosarcoma of the hands and feet had an excellent prognosis.

7. Secondary and extraosseous chondrosarcoma, as well as those exhibiting constitutional symptoms, tended to have a poorer prognosis.

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