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MESENCHYMAL CHONDROSARCOMA — A CASE REPORT WITH BRIEF REVIEW OF THE LITERATURE

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Sumrnary

A 21 year old Chinese female was found to have a thoracic tumour on routine chest radiography. At operation, the tumour was observed to arise from the left sixth rib and was adherent to the underlying pleura. It was excised with the rib. Histological sections showed a lesion composed of foci of cartilage alternating with sheets of malignant spindle cells. In areas, a marked haemangiopericytomatous pattern was present. These unique features are diagnostic of a rnesenchymal chondrosarcoma. The clinical features of 85 of the approximately 125 reported cases of rnesenchymal chondrosarcoma are reviewed.

INTRODUCTION

Mesenchymal chondrosarcoma is a rare but clinicopathologically distinct neoplasm of bone and extraskeletal soft tissues. It has not previously been described to occur in the Malaysian population.

CASE REPORT

The patient is a 21 year old Chinese girl who was found on routine chest radiography (Fig. 1) to have a tumour involving the left sixth rib. This was diagnosed radiologically as an osteochondroma. At operation, a lobutated grey mass measuring 5 x 4 x 3cm was found arising from the inner surface of the rib. The tumour was hard in consistency and contained foci of calcification. The rib was excised leaving behind residual tumour adherent to the pleura.

Histology

The tumour was fixed in neutral buffered formalin and routinely processed. 5 pm thick paraffin sections were obtained and stained with haemalurn and eosin. Silver impregnation for reticulin fibres was also performed. Sections showed a lesion dominated by sheets of spindle cells with uniform hyperchromatic nuclei (Fig. 2). Mitotic figures were rare. Numerous capillary spaces were present thus giving a markedly haemangiopericytomatous appearance (Figs. 3, 4 and 5). isolated areas of well formed cartilage with foci of calcification were present (Fig. 5). The transition from 'spindle-cell to chondroid areas was abrupt. Osteoid was not seen. These unique histological features are diagnostic of mesenchymal chondrosarcorna.

DISCUSSION

Mesenchymal chondrosarcoma was first described as a distinct skeletal neoplasm by Lichtenstein and Bernstein[1] in 1959. Three years later, Dahlin and Henderson[2] noted its occurence in the cranial dura mater. Since then, a total of at least 125 cases have been reported from throughout the world. In addition, some of the lesions described by Hutter *er al*[3] in 1965 as primitive multipotential primary sacroma of bone were undoubtedly mesenchymal chondrosarcoma. Hutter *er al*[3] also noted that Wirth and Shimkin[4] in 1943 had already described a similar neoplasm. This was described by them as a 'chondrosarcoma of the nasopharynx simulating juvenile angiofibroma'. Jacobson believes that mesenchymal chondrosarcoma represents one morphologic type of the bone tumour which he calls ''polyhistioma''.[5]

Data from 85 of the recognized cases in the English literature [1-4, 6-24] were traced and the results reviewed. 45 (53%) patients had skeletal neoplasms while 40 (47%) had the extraskeletal variant. An additional 7 patients[25-31] had to be excluded because their ages and sexes were not stated. All these 7 patients had extraskeletal neoplasms; *6* had orbital lesions and the seventh a lesion involving the nail bed of the thumb. The patient reported here is not included in the tables.

Tables 1, 2 and 3 show the distribution of cases by age and sex. An overall predilection for the second and third decades is apparent with 48 patients between the ages of 10 and 30 years. This is predominantly due to those with skeletal lesions in which 30 (68%) out of 44 patients belonged in this age group. The distribution of extraskeletal lesions by age is much more uniform with 19 patients (48%) between the age of 10 and 30 years and 17 (43%) patients older than 30 years. The oldest patient with a skeletal lesion was a 58 year old male and the youngest a 2 year old female. The corresponding ages for the extraskeletal variant were 84 years (female) and 5 years (male).

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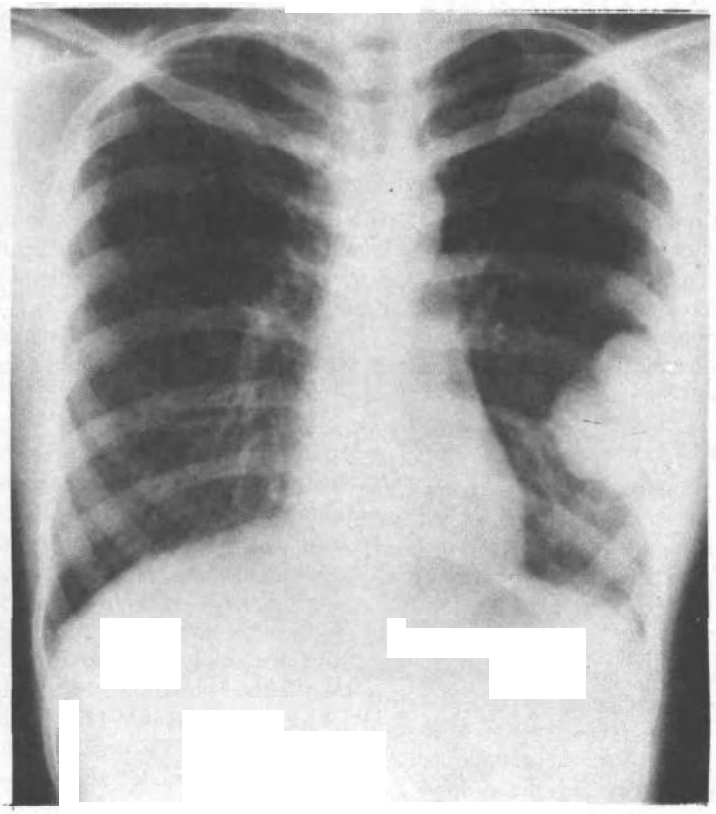

Fig. 1. Chest radiograph showing lobulated mass arising from left sixth rib.

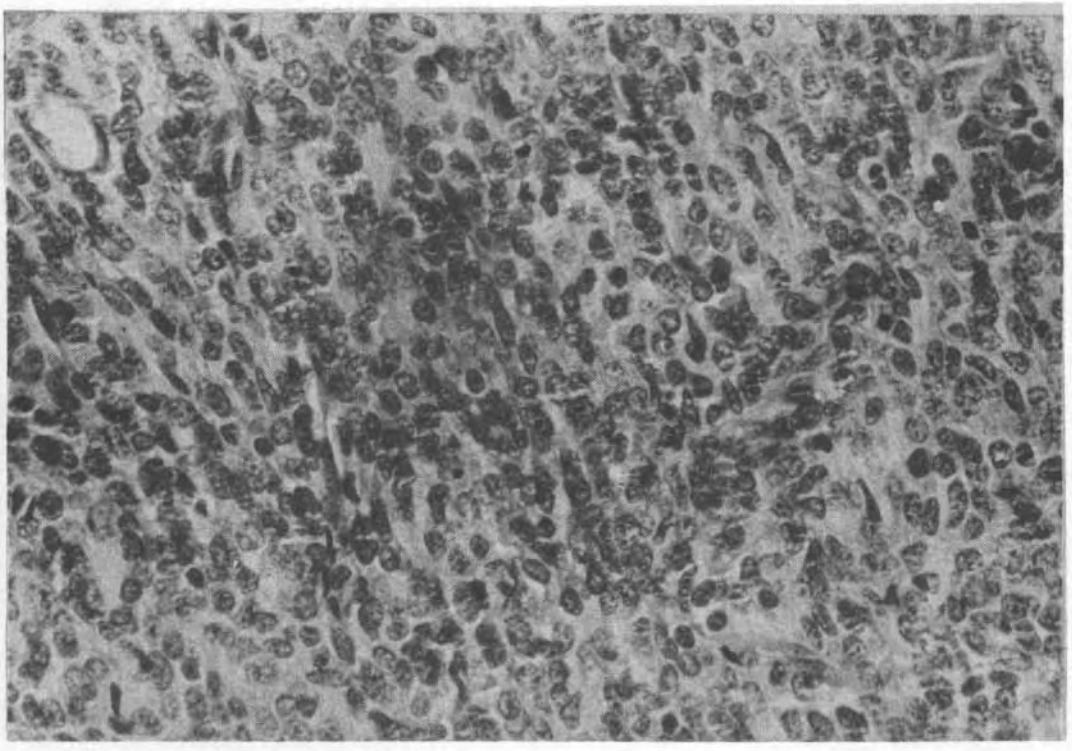

Fig. 2. **Sheets of spindle cells. H&E x 400**

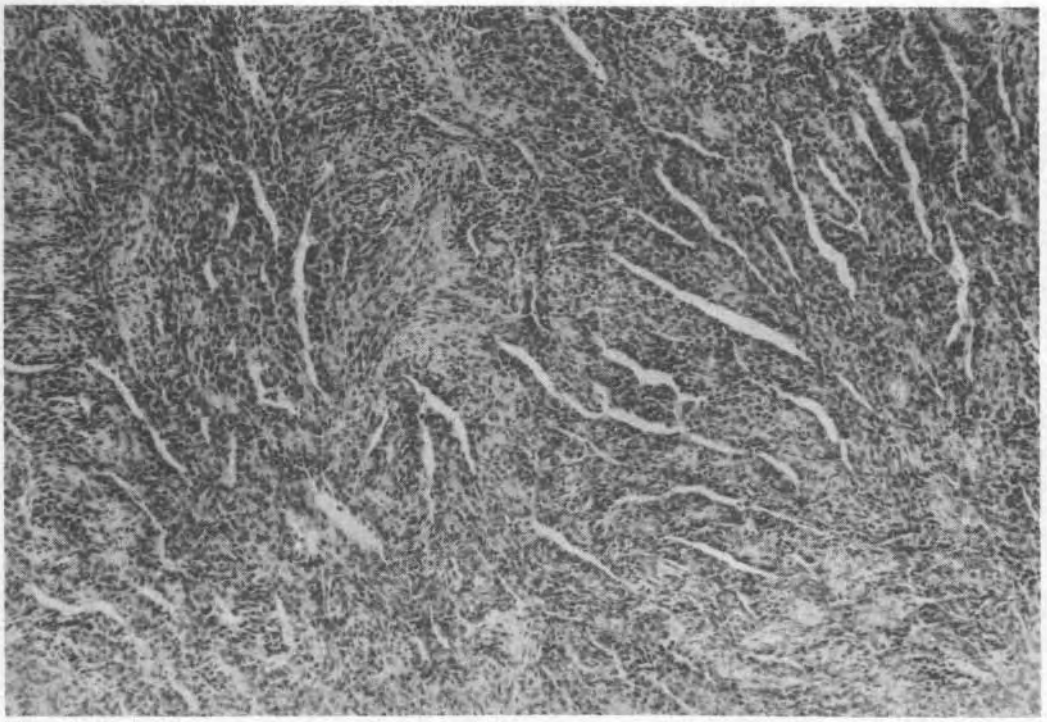

Fig. 3. **Capillary spaces in a spindle-cell strorna. H&E x 100.**

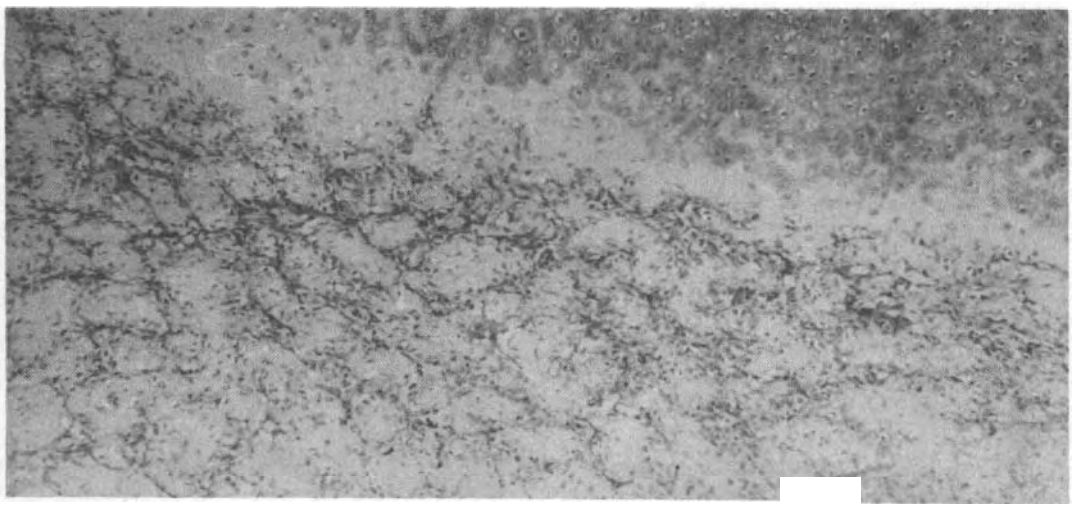

Fig. 4. **Cartilage and haernangiopericytornatous area. H&E x 100.**

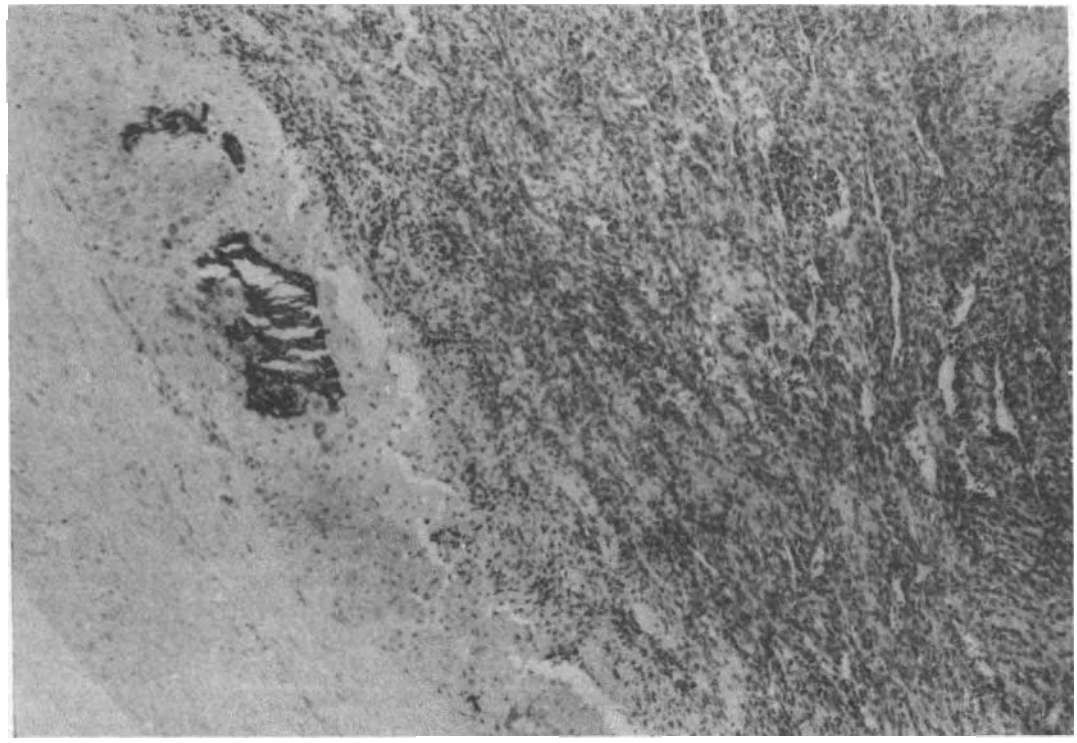

Fig. 5. Calcified cartilage and haemangiopericytomatous area. H&E x 100.

TABLE 1
DISTRIBUTION OF MESENCHYMAL CHONDROSARCOMA
BY AGE AND SEX

Age (years)	No. of Patients		
	Male	Female	Total
Under 10	3	2	5
10 – 19	9	15	24
20 – 29	11	13	24
30 – 39	4	6	10
40 – 49	4	7	11
50 and above	4	5	9
TOTAL	35	48	83

(1 male and 1 female of unknown age not included)

TABLE 2
DISTRIBUTION OF SKELETAL MESENCHYMAL CHONDROSARCOMA BY AGE AND SEX

Age (years)	No. of Patients		
	Male	Female	Total
Under 10	0	2	2
10 — 19	5	10	15
20 — 29	7	8	15
30 — 39	3	3	6
40 — 49	2	2	4
50 and above	1	1	2
TOTAL	18	26	44

(1 female of unknown age excluded)

TABLE 3
DISTRIBUTION OF EXTRASKELETAL MESENCHYMAL CHONDROSARCOMA BY AGE AND SEX

Age (years)	No. of Patients		
	Male	Female	Total
Under 10	3	0	3
10 — 19	4	6	10
20 — 29	4	5	9
30 — 39	1	3	4
40 — 49	2	5	7
50 and above	3	3	6
TOTAL	17	22	39

(1 male of unknown age excluded)

There is a slight female preponderence, the female: male ratio being 1.4;1. This feature is present in both the skeletal as well as the extraskeletal neoplasms.

In contrast, the usual chondrosarcoma is commoner in males (male: female ratio — 1.5: I) and is commoner after the age of 30 years with a maximum incidence in the fifth decade.

The distribution of lesions by site is shown in Tables 4 and 5. Overall, there is an apparent preference for the head and neck by both skeletal as well as extraskeletal neoplasms. Among the skeletal lesions, the maxilla, mandible and ribs are by far the commonest bones affected. Lesions of long bones are uncommon. Among the extraskeletal lesions, the commonest sites of occurence are the craniospinal meninges, the orbit and the soft tissues of the lower limb.

The aetiology of mesenchymal chondrosarcoma remains unknown. Sears et al[9] described a patient who received therapeutic irradiation to the retroperitoneal area during infancy for an adrenal carcinoma. Twelve years later, she developed a mesenchymal chondrosarcoma in the lower thoracic vertebrae.

Some authors[1,2,17] have commented on the possible multicentric origin of the skeletal lesions because of the development of multiple osseous lesions in the same patient over a long time period. In their original paper, Lichtenstein and

TABLE 4
DISTRIBUTION OF SKELETAL MESENCHYMAL CHONDROSARCOMA BY SITE

Site	No. of Patients
Skull	3
Facial bones and hard palate	11
Vertebra	5
Ribs	10
Sacrum and pelvic bones	4
Scapula	1
Humerus	1
Femur	5
Tibia	2
Fibula	2
Metatarsal	1
TOTAL	45

TABLE 5
DISTRIBUTION OF EXTRASKELETAL MESENCHYMAL CHONDROSARCOMA BY SITE

Site	No. of Patients
Cranial dura	8
Spinal dura	5
Orbit	7
Remainder of head and face	4
Neck	1
Chest wall	2
Paraspinal muscles	1
Abdomen & Retroperitoneum	1
Pelvis	2
Lower limb	9
TOTAL	40

Bernstein[1] named the tumour 'mesenchymal chondrosarcoma developing in multicentric foci'. One of their patients developed 3 histologically similar neoplasms in different sites over a 12 year period; the other developed lesions in the vertebrae, multiple skull bones, sternum, sacroiliac bones and scapula over a 1 year period. Subsequently, Dahlin and Henderson[2] and Pepe *et al*[17] have raised the same possibility.

Ultrastructural investigations[12,13] suggest that the tumour is derived from primitive mesenchyme with the potential towards chondroid differentiation. The vascular pattern is due to proliferation of undifferentiated mesenchymal cells, rather than pericytes, around vascular spaces.

Follow-up data were available in 35 of the patients with skeletal lesions and 32 of those with extraskeletal lesions. These data are summarised in

TABLE 6
STATUS OF PATIENTS WITH MESENCHYMAL CHONDROSARCOMA

Type of Lesion	Dead	Alive with Recurrence/ Metastasis	Alive and Well	Total
Skeletal	22	5	7	34
Extraskeletal	15	6*	11	32
TOTAL	37	11	18	66

*Includes 1 patient with questionable pulmonary metastasis

Table 6. Out of 67 patients, 37 (55%) were dead, 11 (16%) were alive with recurrences or metastases and only 18 were still alive and well at the time of reporting. The longest follow-up period in this last group of patients was 10.5 years. Nevertheless. their prognosis must remain guarded as delayed deaths have been recorded. Dahlin and Henderson* reported a patient who was free of tumour for 22 years before developing a secondary lesion (or perhaps another primary) that killed her 2 years later, 24 years after excision of the original lesion! This propensity for developing late secondaries (or new primaries) is noted in all the large series.

Although the histological features are unique, mesenchymal chondrosarcoma may be mistaken for i) malignant haemangiopericytoma ii) synovial sarcoma and iii) chondrosarcoma. The occurrence of foci of cartilage in haemangiopericytoma and synovial sarcoma is exceedingly rare. The latter lesion also has a characteristic biphasic epithelial and spindle cell pattern. Finally, ordinary chondrosarcoma lacks undifferentiated spindle cell and haemangiopericytorna-like areas.

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