

Bone and Soft Tissue Sarcoma: An Epidemiological Study at the National University of Malaysia, 1992-97

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ABSTRACT

Seventy cases of primary malignant bone and soft tissue sarcomas from 1992 to 1997, treated at the Department of Orthopaedics and Traumatology, National University of Malaysia were analyzed retrospectively.

There were 40 (57%) bone sarcomas and 30 (43%) soft tissue sarcomas. The male to female ratio for bone and soft tissue sarcomas was 1:0.9 and 1:1.7 respectively. Sarcomas of bone occurred predominantly during the first and second decades of life compared to the third and fourth decades in soft tissue sarcomas. The youngest patient was 16 months old and the oldest, 69 years.

The commonest bone sarcomas were osteosarcoma, giant cell tumour and Ewing's tumour. Liposarcoma, fibrosarcoma and malignant fibrous histiocytoma were the commonest soft tissue sarcomas. Twelve patients (17%) had secondaries at presentation.

INTRODUCTION

In Malaysia very few cases of bone and soft tissue sarcoma are detected early in the course of their disease. This may be due to ignorance among the local population and possibly also to the low index of suspicion of the primary physician in the initial stage of the disease.

Treatment by traditional healers is commonly sought after, further delaying orthopaedic treatment.

There is scanty and incomplete information on the pattern of primary malignant bone and soft tissue sarcomas in Malaysia. Previous local studies on bone tumors were by Bovil et al¹ on osteosarcoma and Peh et al² on tumour and tumour-like lesions of bone, from University Hospital, Kuala Lumpur.

In order to address this paucity of local data, the aim of our study is to determine the type of sarcomas seen at our institution and provide additional local data on sarcomas involving the musculoskeletal system.

MATERIAL AND METHODS

We reviewed the medical records retrospectively from January 1992 to December 1997 of all cases of bone and soft tissue sarcoma that were treated in our institution. All cases were confirmed by histopathological examination.

Seventy case notes were available for review. Only bone and soft tissue sarcomas involving the musculoskeletal system were included. Metastatic disease and marrow tumors including plasmacytoma and lymphoma were excluded in this study.

The histopathological specimens were prepared and examined by the staff from the pathology department. The cases were classified according to their histological diagnosis. The age, gender, ethnic group, as well as the site of the bone and soft tissue sarcomas were recorded.

RESULTS

Table 1 shows the types of tumour, gender and ethnic groups. There were 40 cases of bone (57%) and 30 cases (43%) of soft tissue sarcomas. Of the 40 bone sarcomas, there were 19 (47.5%) osteosarcomas, 11 (27.5%), giant cell tumours, 6 (15%) Ewing's sarcomas and 4 (10%) chondrosarcomas.

Of the 30 cases of soft tissue sarcomas, there were 11 (37%) liposarcomas, 4 (13%) malignant fibrous histiocytomas (MFH), 4 (13%) fibrosarcomas and 3 (10%) synovial sarcomas.

The other soft tissue sarcomas were malignant hemangiopericytoma (2), primary sarcoma of unknown origin (2), rhabdomyosarcoma (2), leiomyosarcoma (1) and neurofibrosarcoma (1).

There were 22 males and 18 females with bone sarcoma. Eleven males and 19 females had soft tissue sarcoma.

The age distribution is shown in Figure 1. Sarcomas of bone predominantly occurred during the first and second decades of life compared to the third, fourth and fifth decades in soft tissue sarcomas. The youngest patient with bone sarcoma was 16 months while the eldest was 64 years with chondrosarcoma. The youngest patient with soft tissue sarcoma was a 10-year old with rhabdomyosarcoma and the eldest was a 69-year old with liposarcoma.

The sites of involvement of bone and soft tissue are shown in Figures 2 and 3 respectively. The majority of patients with bone and soft tissue sarcoma presented with a localized swelling over the affected region. Most of the tumours were extra-compartmental. Out of 70 cases of bone and soft tissue sarcoma, 12 (17%) had a secondary deposit at presentation. Secondary deposits at presentation were found in 7 (59%) osteosarcomas, 3 (25%) liposarcomas, 1 (8%) malignant fibrous histiocytoma and 1 (8%) primary sarcoma of unknown origin. The commonest site of secondaries was the lungs in 7 cases (58%), followed by osseous deposits in 5 cases (42%).

Fourteen patients (20%) requested for discharge against medical advice to seek alternative (traditional) treatment after the diagnosis was confirmed and surgery planned. This

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Table 1. Types, gender and ethnic distribution of bone and soft tissue sarcomas [n=70]

Type of sarcoma	No. of cases	Gender		Ethnic group		
		Male	Female	Malay	Chinese	Indian
Osteosarcoma	19	10	9	12	4	3
Giant cell tumor	11	7	4	8	1	2
Ewing's sarcoma	6	2	4	3	0	3
Chondrosarcoma	4	3	1	1	2	1
Liposarcoma	11	3	8	7	4	0
Fibrosarcoma	4	1	3	3	0	1
Malignant fibrous histiocytoma	4	3	1	4	0	0
Synovial sarcoma	3	1	2	2	1	0
Rhabdomyosarcoma	2	1	1	2	0	0
Primary sarcoma of unknown origin	2	2	0	1	1	0
Malignant haemangiosarcoma	2	0	2	1	1	0
Neurofibrosarcoma	1	0	1	1	0	0
Leiomyosarcoma	1	0	1	1	0	0

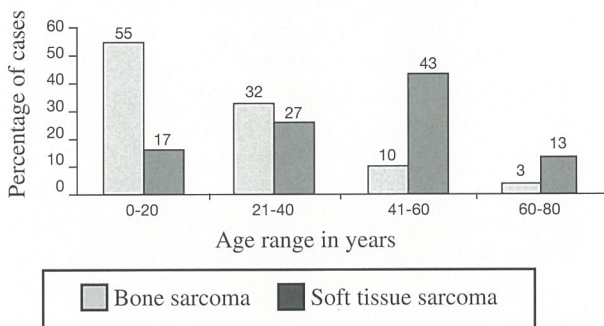


Fig. 1. Age distribution of bone and soft tissue sarcomas (n=70)

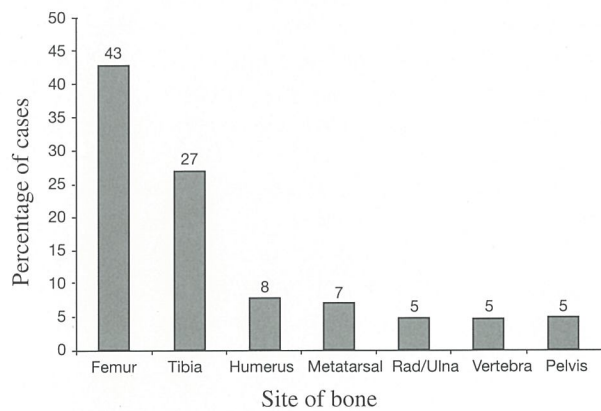


Fig. 2. Site of bone involvement (n=40).



Fig. 3. Site of soft tissue involvement (n=30).

was frequently for cases that were planned for amputation. Definitive surgery depended on the site of the tumours. This included above-knee amputation, below-knee amputation, hemipelvectomy, shoulder disarticulation, elbow amputation, debulking surgery, wide excision and other limb preservation surgery.

Out of the remaining 56 patients (80%) who agreed for definitive surgery, 20 had amputations.

The amputation included wide or radical amputation while limb salvage was in the form of wide en-bloc excision or radical en-bloc resection. Two patients had limb preservation surgery using allograft. The types and number of amputation performed are as follows: 11 above-knee amputations, 4 below-knee amputations, 3 disarticulations and 2 ray amputations.

DISCUSSION

The commonest tumours in our study were osteosarcomas (47.5%), giant cell tumours (27.5%) and Ewing's sarcomas (15%).

Peh² from University Hospital, Kuala Lumpur, showed a similar pattern. Osteosarcoma was the commonest (41%), followed by giant cell tumor (33%), Ewing's sarcoma (11%), multiple myeloma (8%) and chondrosarcoma (7%). Our findings were similar to other regional and Western studies^{3,4}. To the best of our knowledge, there are no previous local publication on soft tissue sarcomas. In Scandinavia⁵, the commonest soft tissue sarcomas were malignant fibrous histiocytoma, leiomyosarcoma and liposarcoma. In contrast, the commonest soft tissue sarcomas in our study were liposarcoma, fibrosarcoma and malignant fibrous histiocytoma.

The small number of patients in our study precludes a meaningful statistical analysis. Malignant fibrous histiocytoma may have been underdiagnosed previously as histopathologic classifications have evolved considerably over the last few decades with the aid of new diagnostic techniques, such as immunohistochemistry, electron microscopy and cytogenic analysis. These advances and refinements have resulted in the reclassification of several tumours⁶. Fibrosarcoma, a common histotype 30 years ago, is today considered rare. The sex distribution of osteosarcoma, giant cell tumor and chondrosarcoma showed

a slight male predominance with the exception of Ewing's sarcoma where males are twice as common as females. The small number of soft tissue sarcomas in our study showed a female preponderance, especially liposarcoma and fibrosarcoma. Gustafson⁵ in a large series of 508 cases of soft tissue sarcomas, demonstrated a slight male preponderance. Schajowicz⁴ noted that the highest incidence of osteosarcoma occurred in the second decade of life followed by the first and third decade. Isolated observations of congenital osteosarcoma and familial osteosarcoma have been reported⁷.

Our study showed that most of the bone and soft tissue sarcoma occurred in the first two decades of life. The youngest patient in our series was a sixteen month old boy with osteosarcoma. The incidence of osteosarcoma in Malaysia (cases per 100,000 population per year) was 0.11 in Malays, 0.23 in the Chinese, and 0.23 in Indians¹. The observed difference between the three racial groups was postulated to be not necessarily genetic since there were many differences in life-style between them.

The racial distribution of Ewing's sarcoma in Africa and the United States have suggested a genetic resistance among the Black population⁸. Li⁹ and Peh² showed that Ewing's sarcoma was rare in the Chinese community. There were 6 patients with Ewing's sarcoma in our series. None were Chinese. It is possible that the Chinese may have a similar genetic resistance. Further research in the field of molecular biology may determine whether there is a genetic basis for the rare occurrence amongst the Chinese.

The youngest patient with soft tissue sarcoma was ten years old with rhabdomyosarcoma and the eldest was sixty-nine with liposarcoma. In the Western population, osteosarcomas, developing after middle age are usually secondary to Paget's disease of the bone¹⁰ or irradiation. None of our patients had a history of irradiation or Paget's disease.

Although any bone may be affected, osteosarcoma shows a preference for the long bones, with a predilection for the

metaphyseal areas of the lower end of the femur, the upper end of the tibia and, less frequently, the upper end of the humerus. These are sites of the most actively growing epiphyses. Tumours of the first two sites, represent 58 percent of lesions located around the knee⁴. Our study showed that about 50 percent of bone sarcomas occur around the knee involving the distal femur and proximal tibia. The diaphyses of the long bones and also short and flat bones were infrequently affected. We found only 2 cases (5%) involving flat bones of the pelvis.

Gustafson⁵ reported 75 percent of soft tissue sarcomas, occurred over the proximal part of the lower extremity. Fifty-four percent of our cases involved the proximal part of the femur and gluteal region.

It is now generally accepted that pulmonary micrometastases already exist when the primary tumour is discovered. At present, use of scintigraphy with technetium has demonstrated that bone metastases are more frequent than previously reported. Uribe-Botero et al¹¹, in a clinicopathologic investigation of 243 cases of primary osteosarcoma of bone, found metastases in the following organs: lung (98%), bone (37%), pleura (33%), heart (20%), kidney and liver (11%) and other organs (less than 10%). We were able to detect secondary deposits in 17% of all our cases at presentation. Secondaries to the lung accounted for 60% of cases.

Pisters et al¹² reported a 5-year survival rate of 76% for localised soft tissue sarcoma of the extremities. The retrospective nature of our study has its limitations. We were unable to determine the 5-year survival rate of our patients as a significant number were lost to follow-up.

In conclusion, many of our patients presented late, requiring amputation of the affected limb. An education programme to raise the level of awareness is necessary to reduce the number of patients presenting in the late stage of the disease. None of our patients with Ewing's tumour were Chinese. Further research to determine genetic resistance is required.

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