SOFT TISSUE SARCOMA IN CHILDHOOD

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SYNOPSIS

A review of soft tissue sarcomas at the University Hospital, Kuala Lumpur over a 10 year period reveals a similarity in incidence to that observed in developed countries. Rhadomyosarcoma is the commonest; there were also 2 rare cases of liposarcoma. Most of the cases had metastases at presentation and despite multimodal therapy only 2 patients have survived. Overall prognosis remains poor due to the late presentation and poor follow up.

INTRODUCTION

With improvements in socioeconomic conditions and control of infectious disease, malignancy is fast becoming an important paediatric problem in developing countries. One of the rarer tumours, namely the soft tissue sarcoma although once considered potentially fatal, can now be successfully treated (Wilbur, 1976). The International Union Against Cancer (Doll et al, 1972) has reported varying incidence and multiplicity of sites of origin of these tumours in different parts of the world, but no data is available in Malaysian children. This study reviews the epidemiological, clinical and pathological features and the outcome of treatment in children with soft tissue sarcomas seen at the University Hospital, Kuala Lumpur over a 10 year period.

MATERIALS AND METHODS

All children with soft tissue sarcomas admitted to the Paediatric Unit during the period 1968 through 1977 were studied. Details of the illness, physical findings, pathological classification, treatment and subsequent outcome were reviewed.

RESULTS

EPIDEMIOLOGY

During the index period, 15 children with soft tissue sarcomas were admitted. The total number of paediatric admissions during this period was 24,532 cases. Soft tissue sarcoma accounts for 0.61 per 1000 hospital paediatric admissions and 5.6 per cent of all childhood malignancies at the University Hospital.

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ETHNIC DISTRIBUTION

The 15 cases comprised 9 Chinese, 5 Malays and 1 Indian. There appears to be a lower incidence of soft tissue sarcomas in Indians compared to the other races. (see Table I)

TABLE I ETHNIC DISTRIBUTION OF SOFT TISSUE SARCOMAS

Race	No. of cases	No. of admission 1968 — 1977	Incidence per 1000 admissions
Chinese	9	12,040	0.75
Malay	5	5,298	0.94
Indian	1	6,509	0.15
Others	0	685	—
Total	15	24,532	0.61

SEX

There were 10 males and 5 females. There appears to be no significant association between sex and the incidence of soft tissue sarcomas when compared with the ratio of male: female admissions to the unit. (See Table II).

TABLE II SEX DISTRIBUTION OF SOFT TISSUE SARCOMAS

Sex	No. of cases	Admissions (1974 — 1977)
Male	10	8,353
Female	5	5,459

AGE

The mean age of the patients at presentation was 3 years with the range of 4 months to 8 years.

PATHOLOGICAL CLASSIFICATION

The pathological classification recommended by the World Health Organisation (Enzinger et al, 1969) was adopted. Rhabdomyosarcoma was found to be the commonest soft tissue sarcoma followed by fibrosarcoma, leiomyosarcoma, liposarcoma and haemangioendotheliosarcoma (Table III).

TABLE III RELATIVE FREQUENCY OF SOFT TISSUE SARCOMAS

Histological Type	No. of cases	Male	Female
Rhabdomyosarcoma	7	4	3
Fibrosarcoma	2*	1	1
Leiomyosarcoma	2	2	0
Liposarcoma	2	1	1
Haemangioendothelio sarcoma	2	2	0

*includes one neurofibrosarcoma

RHABDOMYOSARCOMA

Rhabdomyosarcoma comprised 2.7 per cent of all malignancies in childhood and is less common than brain tumour, neuroblastoma and Wilm's tumour. The primary site was the orbit in 2 patients and the nasopharynx, groin, calf, elbow and introitus in single cases respectively. The patients generally presented late with advanced disease. Two patients had stage IIA disease according to the criteria proposed by Pratt (1969); 2 had stage IIB disease and 3 had stage IIIA disease respectively. Despite multimodal therapy, 2 patients with stage III disease died within 8 months while the other defaulted follow up.

FIBROSARCOMA

Two patients had fibrosarcomas; the first had a recurrent left gluteal mass with pulmonary metastases and pleural effusion and defaulted follow up after radiotherapy. The second aged 3 years presented with a calcified posterior mediastinal tumour eroding the seventh rib and has remained well for 38 months following multimodal therapy.

LEIOMYOSARCOMA

Our experience is limited to 2 males, one with a tumour of the right thigh and pulmonary and spinal metastases and the other with a right parotid mass. The first patient died within 2 weeks while the second defaulted 2 months after surgery.

LIPOSARCOMA

This neoplasm was diagnosed in 2 cases. The first died of an extensive peritoneal liposarcoma and metastases to the liver. The second, had a well encapsulated mediastinal liposarcoma which was excised and she has remained well for the past 7½ years.

HAEMANGIOENDOTHELIOSARCOMA

Two males aged 4 years died of haemangioendotheliomas, one with a massive left sided haemothorax and metastic nodules in the lungs and brain and the second with tumour involving the common bile duct, liver, mesentery, adrenals and right kidney.

DISCUSSION

The incidence of soft tissue sarcomas in our series appears similar to that of other countries (Angio & Evans, 1975; Williams, 1975). Rhabdomyosarcoma is the most common and although three histological varieties are described:— embryonal, alveolar and pleomorphic types (Horn and Enterline, 1958), a mixed histological pattern is common, particularly in the embryonal group, in which wide variations are often present within the same tumour (Bale and Reye, 1975).

Prognosis varies with the extent of disease and staging in widely used (Pratt et al, 1972). Survival figures of 0 to 70 per cent have been variously reported for different stages of the disease with multimodal therapy (Jaffe et al, 1973; Clatworthy et al, 1973; Heyn et al, 1974). Wilbur et al (1975) have achieved 50%, 5 year disease free survival in patients with inoperable rhabdomyosarcoma and those

with distant metastases with intensive combination therapy programmes using vincristine, actinomycin and cyclophosphamide (VAC) over a period of 2 years.

We have used multimodal therapy, including VAC and methotrexate but have found it difficult to evaluate the response as most of our patients failed to attend follow-up. The other soft tissue sarcomas are uncommon and management along the same lines as rhabdomyosarcoma is generally adopted.

Included in our series are 2 exceedingly rare cases of liposarcoma in childhood of whom one had metastases in the liver. Only one previous case of liposarcoma with metastases has been reported in children (Knowles and Huggii, 1954).

The outcome in our 15 cases of soft tissue sarcomas is summarised in Table IV. There are only 2 long term survivors giving an overall survival rate of 13.3% which is comparable with the figures reported by Jaffe et al (1973) but is poor compared with other series (Heyn et al, 1974; Wilbur et al, 1975).

TABLE IV OUTCOME IN 15 CASES OF SOFT TISSUE SARCOMAS

	Lost to Follow up	Died	Alive and well
Rhabdomyosarcoma	5	2	—
Fibrosarcoma	1	—	1
Leiomyosarcoma	1	1	—
Liposarcoma	—	1	1
Haemangioendothelioma	—	2	—
	7	6	2

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