PEDIATRIC SOFT TISSUE SARCOMA CLINICO-EPIDEMIOLOGIC STUDY WITH REVIEW OF TREATMENT AND SURVIVAL

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INTRODUCTION

Multiple risk factors were found to affect the prognosis of soft tissue sarcoma. They include age, sex, histologic type and grade, tumour site and size (Rooser et al., 1988).

Rhabdomyosarcoma is the most common pediatric soft tissue sarcoma (Young and Miller, 1975). Overall survival rates have increased from less than 20 % in 1960 to as high as 70 % (Green and Jaffe, 1978). Appropriate therapy is developed by integrating surgery, radiation therapy and chemotherapy with supportive care (Ghavimi et al., 1975).

The potential for chemotherapy and radiation therapy to control gross and microscopic metastatic disease decreases the need of aggressive surgery which result in functional and

cosmotic morbidity. the optimum sequencing as well as the extent of surgery, radiation therapy and chemotherapy are continually evolving.

The aim of this retrospective work is to study and analyse the clinico-epidemiologic characters of pediatric soft tissue sarcoma with review of treatment and survival.

Patients and methods :

Twenty nine patients histologically proved to be soft tissue sarcoma (STS) in children (< 15 years old) were the subject of this study. They were refered and treated between 1981 and 1990 at Radiotherapy Department of Mansoura University Hospital. Age, sex, incidence, site. of the tumour, clinical presentation, histopathological type and treatment received were reviewed.

Surgical treatment was done in 22 patients which was either complete or partial excision of the tumour.

Radiotherapy was carried out in all patients as postoperative treatment,

as a primary line of treatment in patients presenting with tumours not

amenable for surgery or as a palliative measure in patients with recurrence

after surgery. The target volume was planned to include all areas of propa-

ble local spread. Extremities were usually treated with opposing fields.

Radiotherapy total tumour dose was 5000 - 6000 CGy / 5 - 6 weeks / 25 -

33 fractions using DXR 200 - 300 K. V., MA. 15, FSD 50, HVL 2.5 - 4

mmCu or cobalt 60 at SSD 80.

Chemotherapy was used in 24 pa-

tients as adjuvant treatment in rhabdomyosarcoma in recent years (VAC

protocol) For 12 months or in inoperable, metastatic and recurrent cases

(VACA regimen) every 3 - 4 weeks guided by haemogram.

Survival was calculated from the date of attendance to radiotherapy de-

partment. Response to therapy was assessed according to criteria of WHO

(Staquet, 1975) :

- Complete response : The disappearance of all known disease,

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determined by two observations not less than 4 weeks apart.

- Partial response : decrease of 50 % or more in the total size of the lesions with no appearance of new lesions nor progression of any lesion.

- No change : less than 50% decrease in tumour size or less than 25 % increase in the size of one or more measurable lesions.

- Progressive disease : A 25% or more increase in the size of one or more measurable lesions, or the appearance of new lesions.

RESULTS

Pediatric STS constituted 23. 8% of all STS patients seen during the period 1981 - 1990 at Radiotherapy Department and 6. 8% of pediatric solid malignant tumours recorded at the same period.

The mean age was 5.8 + 2 years. The male to female ratio was 1.6 : 1. Rhabdomyosarcoma was the commonest histopathological type (65. 5%). Angiosarcoma was the second common type (17. 2%) followed by undifferentiated sarcoma (13. 8%) then fibrosarcoma (3 - 4%) (Table I, Fig-

ures 1 and 2).

The commonest site of involvement was head and neck (37.9%) followed by the trunk and abdomen (34.5%). The extremities esp. upper limb was the least common site. The total response rate was 63.7% in head and neck STS, 60% in trunk and abdominopelvic, 100% in upper limb and 66.7% in the lower limb (Table 2).

A Swelling was the initial presentation in 89.6% of cases, pain was found in (37.9%), metastases and pressure symptoms were encountered in 20.7 and constitutional symptoms in 13.8% (Table, 3).

Table (4) showed that the immediate total response rate was 65.5% of patients. complete response occurred in 12 / 29 patients (41.4%) whereas 7/29 patients showed partial response (24.1%). Four patients had got stable disease (13.8%) and six patients showed progression of their disease (20.7%). The remission rate was the highest in patients treated with surgery, radiation therapy and chemotherapy (72.3%) total response and (64.7%) complete response followed by patients treated with surgery and radiation therapy (60%) total response and (20%) complete response then patients received radiotherapy plus

chemotherapy (28.6%) partial response. At the end of 3 years only 2 / 29 were survived (6.9%) and were free of the disease (2 / 12 = 16.7%) of the complete responders (Table, 5, Fig. 3.).

DISCUSSION

Pediatric STS accounts for 6.8% of childhood malignancy in the present work, A nearly similar incidence was reported by young and Miller (1975) where rhabdomyo sarcoma represents 4 - 8% of pediatric malignancy.

One of the most important prognostic parameters in pediatric STS is the pathological type of the tumour. Rhabdomyosarcoma provides ample evidence of its aggressive behaviour and its tendency to recur and metastasize (Franz, 1983) Embryonal rhabdomyosarcoma was the commonest histopathological subtype in childhood STS (62.1%) in the present study. Similar findings (64%) was reported by Mostafa et al (1989) at NEMROCK (Kasr El - Eini Centre of Radiation, Oncology and Nuclear Medicine) also by Rodary et al (1988).

Head and neck were the commonest site of involvement in this study (37.9%). nearly similar figure (38.5%)

was reported by Mostafa et al (1989). therapy (82.3%) and 64.7%) respectively.

We conform with Romadahi (1983)

that the best remission rate was seen

in children with STS of the extremities

(100%) in the upper limb and (66.7%)

in the lower limb in the present series,

this may be due to the facility of adequate surgical excision in the extremities and early diagnosis in these

parent sites.

Although STS are widely considered as radioresistant tumours as their response to radiation is slow (Edland, 1968), yet there is accumulating evidence that some of these tumours are radiocurable such as rhabdomyosarcoma, radical radiotherapy after limited surgery plays a main role in improving the control rate (Dewar and Duncan, 1985). The addition of effective cytotoxic chemotherapy may improve the prognosis by increasing the probability of local control and decreasing the risk of micrometastases (Rosenberg, 1984). The value of adjuvant chemotherapy is well established now for childhood STS especially in rhabdomyosarcoma (Maures et al., 1988 and Rodary et al., 1988). This is reconfirmed in the present work where the best total complete remission were obtained in patients who received post-operative radiotherapy and chemotherapy.

A total immediate response (complete and partial) of (65.5%) was obtained in the present series. Complete remission was achieved in (41.4%) of patients with 3 year actuarial survival of (6.9%). These figures are similar to those of Mostafa et al. (1989) which was (68%) but much lower than most of recently reported series for western countries (Dewar and Duncan, 1985 and Duncan, 1985 and Rodary et al., 1988). This difference can be explained by differences in clinicopathologic characteristics of each series, gross residual disease prior to radiation in the present series as well as suboptimum treatment given to some of our earlier patients who did not receive adjuvant chemotherapy.

In conclusion the successful management of pediatric cancer requires a carefully orchestrated combined modality team comprised of a pediatric oncologist, a surgeon, a radiotherapist, diagnostic specialists (radiology, nuclear medicine, pathology, clinical laboratories). In pediatric STS a combination of adequate surgery with post-operative irradiation and adjuvant chemotherapy can give a high remission

rate. A prospective randomised study with rhabdomyosarcoma has improved markedly with the addition of combination chemotherapy to surgery and radiation therapy, the long term sequelae of these therapies in young children must be studied.

Table (1) : - Age, sex and pathological type distribution in 29 cases of pediatric soft tissue sarcoma.

Type of STS	Patients		mean age	sex ratio
	No.	%		
Rhabdomyo sarcoma	19	65.5	6.3	2.1:1
Embryonal	18	62.1		
Alveolar	1	3.4		
Angiosarcoma	5	17.2	4.1	1 : 1
Fibrosarcoma	1	3.4	10.3	1.9 : 1
Undifferentiated sarcoma.	4	13.8	2.2	1.4 : 1
Total	29	100	5.8	1.6:1

STS : Soft tissue sarcoma.

Table (2) : Anatomical distribution of 29 patients with STS and response to treatment .

Site	Patients		C. R.		N. R.		P. D	
	No.	%	No.	%	No.	%	No.	%
Head and neck	11	37.9	5	45.5	1	9.1	3	27.3
Trunk and abdominopelvic.	10	34.5	3	30	2	20	2	20
Upper limbs	2	6.9	1	50	-	-	-	-
Lower limbs	6	20.7	3	50	1	16.7	1	16.7
	29	100	12	41.4	4	13.8	6	20.7

C. R. : Complete response.
P. R. : Partial response.
P. D. : Progressive disease.
N. R. : No response.

Table (3) : Clinical features in 29 Patients with pediatric STS at the time of presentation.

Clinical feature		Patients	
		No.	%
Swelling		26	89.6
Pain		11	37.9
Metastases		6	20.7
Pressure symptoms		6	20.7
Constitutional symptoms		4	13.8

Table (4) : Response to different treatment modalities of pediatric soft tissue sarcoma.

Treatment modality	Patients		Type of Response							
			C. R		PR		NR		PR	
	No.	%	No.	%	No.	%	No.	%	No.	%
S + R. T.	5	17.2	1	20	2	40	1	20	1	20
S + R. T. + C	17	58.6	11	64.7	3	17.6	1	5.9	2	11.8
R. T. + C. T.	7	24.1	-	-	2	28.6	2	28.6	3	42.9
Total	29	100	12	41.4	7	24.1	4	13.8	6	20.7

S : Surgery. R. T. : Radiation therapy.

C. T. : Chemotherapy.

Table (5) : Acturial survival rate in 29 patients with pediatric soft tissue sarcoma.

Duration in months	Survival r ate	
	No.	%
6	21	72.4
12	11	37.9
18	7	24.1
24	3	10.3
30	2	6.9
36	2	6.9

Fig. 1. Rhabdomyosarcoma of the

kidney

Fig. 2. Retroperitoneal angiosarcoma.

Fig. 3. Actuarial survival rate in 29 patients with pediatric soft tissue sarcoma.

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المختص المرضي

سرطان الأبنسجة الرخوة في الأطفال

دراسة أكاديمية، وراثية مع مراجعة العلاج ومعدل البقاء على الحياة

المشتركين في البحث

د. / ناظم شمس / أحمد حسين الشحات

(من أقسام علاج الأورام بالأشعاع والجراحة)

أخرى البحث على ٢٩ طفل يعاني من سرطان الأبنسجة الرخوة الذين تلقوا علاجاً شاملاً

بأشعاع والأشعاع في الفترة ما بين ١٩٨١، ١٩٩٠. وقد رجعت ملفات المرضى من حيث الصفات

الوراثية والأكلينيكية والعلاج ومعدل البقاء على الحياة.

- نتائج البحث كما يلي :-

سرطان الأبنسجة الرخوة في الأطفال ٨٣، ٨٪ من سرطان الأبنسجة الرخوة في

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وكانت ساركوما الغدد الصماء هي أكثر الأبنسجة الرخوة انتشاراً ٥، ٦٪ منها ساركوما

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وكان معدل البقاء على الحياة بعد ٣ سنوات ٩، ٩٪ (٩، ٩٪)

ومن هذا يتضح أن علاج سرطان الأبنسجة الرخوة في الأطفال يحتاج إلى تعاون كل من الجراحة

والعلاج الإشعاعي والكيميائي للحصول على أفضل النتائج.