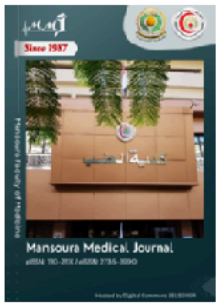
MANSOURA MEDICAL JOURNAL

ISSN - Print: 1110-211X - Online: 2735-3990

journal homepage: mmj.mans.edu.eg



Volume 19 | Issue 1 Article 19

PEDIATRIC SOFT TISSUE SARCOMA CLIN1COE-PIDEM10LOGIC STUDY WITH REVIEW OF TREATMENT AND SURVIVAL

A. H EL - Shahat

Departments of Radiation Oncology & Surgery Mansoura

N. M Shams

Departments of Radiation Oncology & Surgery Mansoura

PEDIATRIC SOFT TISSUE SARCOMA CLINICOEPIDEMIOLOGIC STUDY WITH REVIEW OF TREATMENT AND SURVIVAL

By

EL - Shahat, A. H. and Shams, N. M.

From

Departments of Radiation Oncology & Surgery Mansoura Egypt

Received for Puplication 3 / 4 / 1990

INTRODUCTION

Multiple risk factors ware 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 % (Creen 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 clinicoepidemiologic 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 propable 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. I5, FSD 50, HVL 2. 5 - 4 mmCu or cobalt 60 at SSD 80.

Chemotherapy was used in 24 patients 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 attendence to radiotherapy department. Response to therapy was assessed according to criteria of WHO (Staquet, 1975) :

- **Complete response :** The disappearance of all known disease, 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 measrable 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% 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 I.6 : I. 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 I 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 occured in 12 / 29 patients (4l. 4%) whearas 7/29 patients showed partial response (24.1%). Four patients had got stable disease (I3. 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 / I2 = 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 prognostric 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. Sinilar 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).

We conform with Romadahl (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 facillity of adequate surgical excision in the extremities and early diagnosis in these apparent sites.

Although STS are widely considered as radioresistent 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 Dunkan, 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 presont work where the best total complete remission were obtained in patients who received post - operative radiotherapy and chemotherapy (82.3%) and 64.7%) respectively.

A total immediale 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 acturial 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 characterestics 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 is essential using more effective drugs as anthracyclines and ifosfamide on neoadjuvant basis in bulky lesion may lead to better local control and survival. Although the survival of patients 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	I9	65.5	6.3	2.1:1
Embryonal	I8	62.I		
Alveolar	I	3.4		
Angiosarcoma	5	17.2	4.I	1 : 1
Fibrosarcoma	I	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 29patients 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.

N. R : No response. P. D. : Progressive disease

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

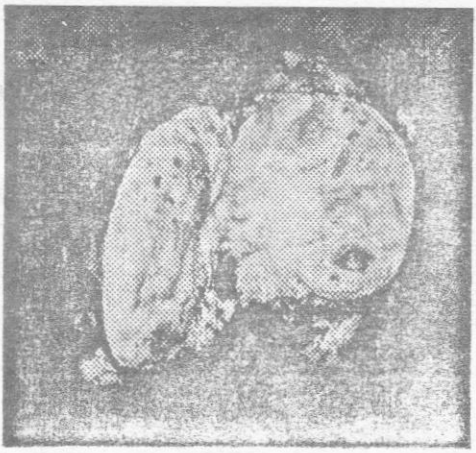


Fig. 1. Rhabdomyosarcoma of the kidney

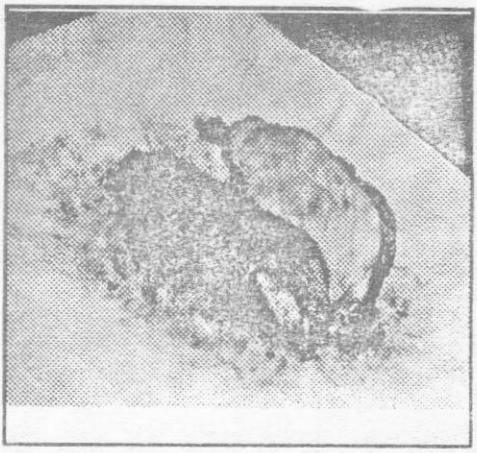


Fig. 2. Retroperitoneal angiosarcoma.

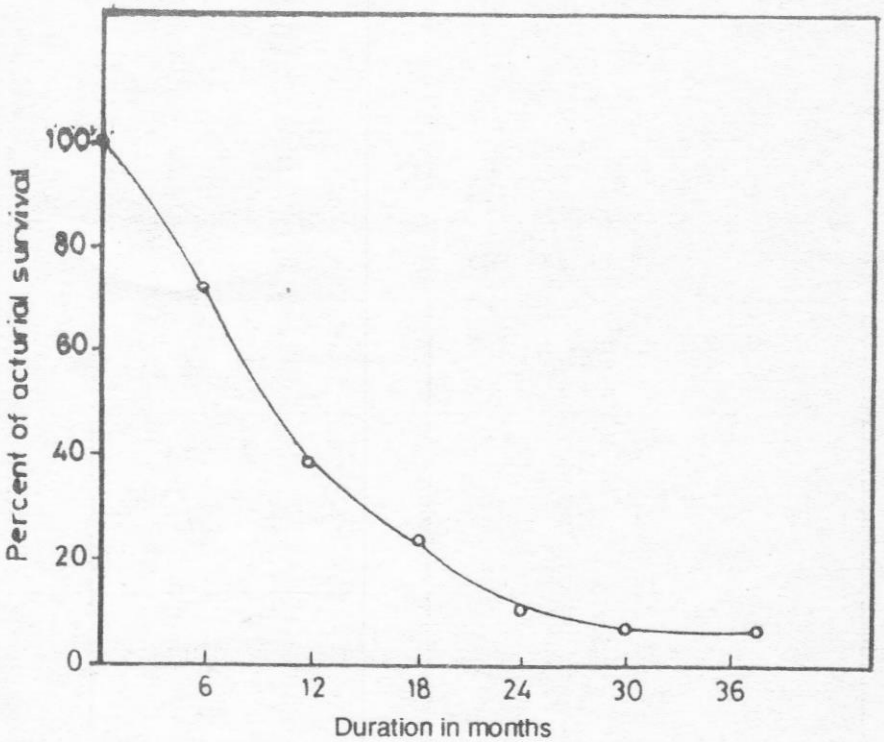


Fig. 3. Acturial survival rate in 29 patients with pediatric soft fissue sarcoma.

REFERENCES

EDLAND, R. W. (1985) : Am. J. Rontegen. Rad. Ther Nucl. Med. 103, 778 - 791.

DEWAR, J. A. and DUNCAN, W. (1985) : Clinical Radiol. 36 : 629 .

FRANZ, N., SHARON, W (1983) : Soft Tissue Tumours, S. T. Lowis, Toronto, London .

GHAVIMI, F.; EXELBY, R. R., DANGIO, G. J. et al. (1975): Cancer 35 : 677 - 686 .

GREEN, D. M. and JAFFE N. (1978): Cancer Treat. Rev. 5 : 7 - 27 .

MAURER, H. M.; MOHAN, B.; GEHEN, A. et al (1988) : Cancer 6l : 209 - 220

MOSTAFA, H; El - GHONIEMY, E.; ABDEEN, M.; MoSTAFA, A. and El - HADAD, SH. (1989) : EGYPT. J. Radiol. 20 (l): 79 - 86.

RODARY, C.; REY; A.; OLIVE, D. et al. (1988) : Med Ped. Oncol., 16 : 7l - 77.

ROMADAHL, M. H., LINDBERG, R. D. and MARTIN, R. G. (1978) : Cancer Treat. Symp., 2 : 25 l.

ROOSER, B.; ATTEZLL, R.; BERG, N. O. and RYDHOLM, A. (1988) : Cancer 6l : 817 - 823.

ROSENBERG, S. A., (1984) : Cancer Treat. Reports, 68 : 1067- l078.

STAQUET, M. J. (1975) : Cancer Therapy: Prognostic Factors and Criteria of Rasponse, Raver Press, New York.

YOUNG, J. L. MILLER, H. W. (1975): J. Pediatr. 86 : 254 - 258.

الملخص العربى

سرطان الأنسجة الرخوة فى الأطفال
دراسة أكلينيكية وبائية مع مراجعة العلاج ومعدل البقاء على الحياة

المشتركون فى البحث

د. / أحمد حسين الشحات د. / ناظم شمس

(من أقسام علاج الأورام بالأشعاع والجراحة)

أجرى البحث على ٢٩ طفل يعانى من سرطان الأنسجة الرخوة الذين عولجوا بقسم علاج الأورام بالجراحة والأشعاع فى الفترة مابين ١٩٨١ ، ١٩٩٠ . فقد روجعت ملفات المرضى من حيث الصفات الوبائية والأكلينيكية والعلاج ومعدل البقاء على الحياة .

وكانت نتائج البحث كما يلى : -

(١) تكون سرطان الأنسجة الرخوة فى الأطفال ٢٣,٨ ٪ من سرطان الأنسجة الرخوة الكلى، ٦,٨ ٪ من الأمراض الخبيثة فى الأطفال ومتوسط العمر ٨ ، ٥ سنة ونسبة الذكور للأناث ١,٦ : ١ وكانت ساركوما العضلات الأرادية هى أكثر الأنواع الباثولوجية أنتشارا ٦٥,٥ ٪ قلبها ساركوما الأوعية الدموية ١٧,٢ ٪ وأكثر الأماكن أصابة كان منطقة الرأس والعنق ٣٧,٩ ٪ وأقلها هى الأطراف وظهرت معظم الحالات على هيئة تورم فى (٨٩,٦ ٪) وكان معدل الأستجابة الكلى للعلاج ٦٥,٥ ٪ وأعلى معدل للأستجابة وجد فى المرضى الذين عولجوا بالجراحة والعلاج الأشعاعى والعلاج الكيمائى (٨٢,٣ ٪) ووجد أعلى معدل للأستجابة فى حالات سرطان الأنسجة الرخوة فى الأطراف، وكان معدل البقاء على الحياه بعد ٣ سنوات (٦,٩ ٪) .

ومن هذا يتضح أن علاج سرطان الأنسجة الرخوة فى الأطفال يحتاج الى تعاون كل من الجراحة والعلاج الأشعاعى والكيمائى للحصول على أفضل النتائج .