Soft Tissue Sarcomas of The Limbs

An essay submitted in Partial
fulfiiment of Master degree in General Surgery

By

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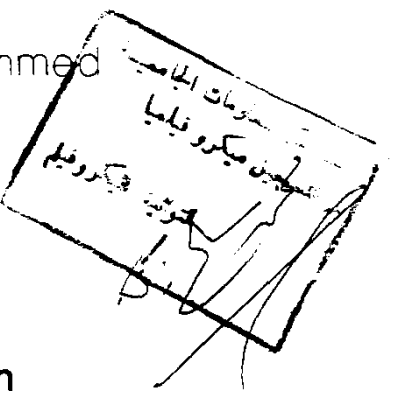

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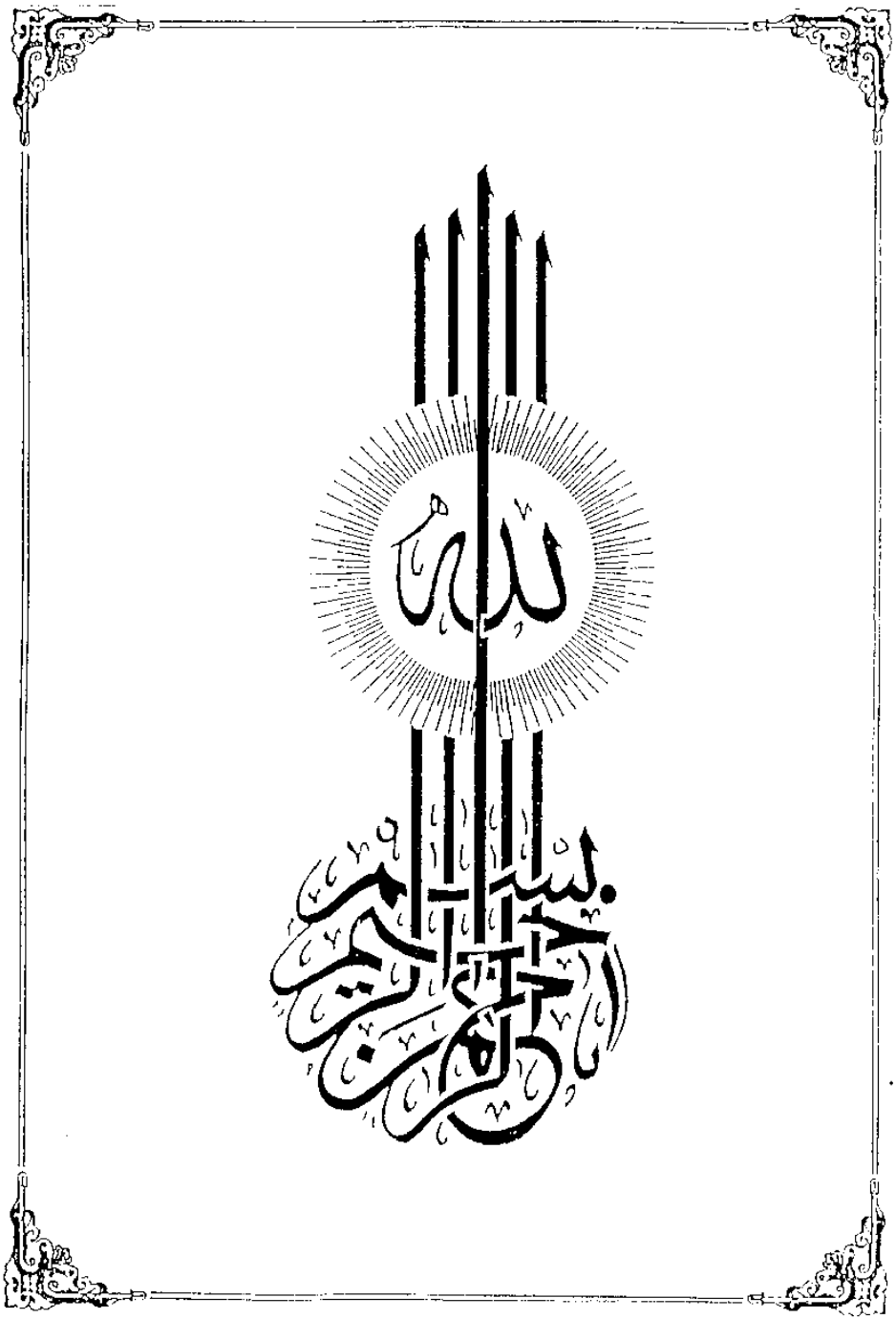

TO MY MOTHER

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INTRODUCTION

Soft tissue sarcomas of the limbs

Soft tissue refers to extraskeletal connective tissue of the body that connect, support, surround other discrete anatomic structures. ***[Rosenberg et al., 1985]***. These include fatty, fascial, muscle, fibrous, lymphatic and vascular tissues. Because of similarities in anatomic sities of origin, clinical presentation, clinical behavior, tumors arising in schwan cells "a class of cells surrounding peripheral nerves that arise from neural tube of primitive ectoderm" are also included in the category of soft tissue sacrcomas. Soft tissues comprises over 50% of the body weight. The over 400 muscles in the human body comprise about 40% of adult body weight. Soft tissue sarcomas refer to a large variety of malignant tumors arising in the soft tissue that are grouped because of similarities in the pathologic appearance, clinical presentation and behavior. ***[Rosenberg et al., 1989]***

Soft tissue sarcomas account for around 1% of all malignant tumors ***[Enzinger and Weiss, 1988]***, the majority arise on the extremities, the lower limb and pelvic girdle being the most frequent sites . Approximately 60% of sarcomas occurs in extremities, the ratio of lower extremity to upper extermity tumors is 3: 1, about 75% of the lower extremity sarcomas originate at or above the knee. ***[Torosian et al., 1988]*** **Table (1).**

Table (1) Sites of Soft Tissue Sarcoma

Study	Sites				
	Head and Neck	Trunk / Retroperitoneum	Upper Extremity	Lower Extremity	Total
Shieber et al (1961)	16	39	20	50	125
Hare et al (1963)	42	34/5	32	48	161
Ferrell et al (1972)	8	19	15	36	78
Sears et al (1980)	12	16	6	26	60
Abbas et al (1981)	24	66/38	42	81	251
Lindberg et al (1981)	26	74	63	137	300
Potter et al (1985)	12	48/36	59	152	307
Torosian et al (1987)	21	92/90	81	208	492
Acs Survey (1987)	406	872/568	594	2110	4550
Total	567	1997	912	2848	6324
%	(9)	(32)	(14)	(45)	(100)

Other sites include head and neck region, account for approximately 9% of these tumors, the trunk 31%, within the trunk approximately 40% of tumors are located in the retroperitoneum and the remaining tumors are located in the abdominal wall, chest wall, mediastinum and breast. ***[Rosenbrg et al., 1989]***.

The aim of the local treatment is complete eradication of the primary tumor with minimal functional disability, this requires multidisciplinary approach in which surgery, radiotherapy and chemotherapy are used in complementary fashion.

EPIDEMIOLOGY

Epidemiology

Soft tissue sarcomas are uncommon tumors. In 1989 there were about 5,600 cases diagnosed in U.S.A and approximately 3,000 patient died from this disease ***[Lawrence and Neifeld, 1989]***.

The annual age adjusted incidence of soft tissue sarcomas is roughly 2 per 100,000 population , that of extremity sarcomas is arround 1.4 per 100,000. ***[Rosenberg et al., 1985]***

Data from Newzeland suggests that both the incidence and mortality from soft tissue sarcomas has been increasing from 1.3 per 100,000 in 1955 to 2 per 100,000 in 1977, in that countery although no change in incidence was seen in Dennmark during the same period. ***[Lynge et al., 1987]***.

Soft tissue sarcomas are commonest in fifth and sixth decades of life but occur in all age groups including children in whom they rank fifth as a cause of death from malignancy behind leukaemia, central nervous system cancers, lymphomas, sympathetic nervous system cancers. There is no sex predilection. ***[Rosenberg et al., 1985]***.

Diagnosis of soft tissue sarcoma is difficult because of the multiplicity of histologic types and overlapping morphologic pattern, in almost every major study of specific sarcoma types a significant error rate is mentioned, the marked differences seen

Table (2) Relative incidence (%) of Histologic Types of Soft Tissue Sarcomas in Various Studies

	Study											
	Shieber and Graham (1962)	Hare and Cerny (1963)	Pack and Ariel (1964)	Martin et al (1965)	Ferrell and Frable (1972)	Shiu et al (1975)	Stimon and Enneking (1976	Russell et al (1977)	Lindberg et al (1977)	Suit (1983)	Potter et al (1985)	Collins et al (1985)
Sites	All	All	All	All	All	Extremity	Extremity	All	All	All	All	Extremity
Total No. of Cases	125	200	717	398	117	297	54	1215	166	315	307	315
	Types of Soft-Tissue Sarcomas											
Unclassified	0	28.5	36.4	14.8	0	7.1	5.6	10	6	16.5	9.5	1
Liposarcoma	16	11.5	14.5	26.3	17	27.6	18.5	18.2	12.7	15.2	18.3	33.9
Rhabdomyosarcoma	16	5	13.9	26.6	30	17.5	5.6	19.3	9.6	4.1	2.9	9.2
Synovialosarcoma	0.8	2.5	3.4	3	2.5	14.1	5.6	6.9	10.2	3.8	19.5	12.6
Neurofibrosarcoma	3.2	0	6.4	0	0	5.4	0	4.9	19.3	6	6.8	7.6
Fibrosarcoma	44	43	5.4	24.1	33	20.2	37	19	13.3	16.8	3.5	12.3
Angiosarcoma	4.8	0	2.6	0.3	0	2	0	2.7	1.2	3.2	1.6	1.2
Leiomyosarcoma	6.4	6.5	0	6.3	4	2.4	0	6.5	4.2	7.6	11.4	3.8
Mesenchymoma	0	0	0	0	0	0.3	0	0.3	0	0.9	0	0
Malignant fibrous histiocytoma	0	0	0	0	0	1	20.4	10.5	17.5	15.6	22.8	14.6
Other	8.8	3	12.1	4	13.5	2.4	7.4	1.7	6	10.2	3.6	3.4

in the relative frequences in **(Table II)** reflect difference in diagnostic criteria used by different pathologist. ***[Potter et al., 1986]***.

Epidemiologic factors

Little is known about epidemiologic factors of importance in patients with soft tissue sarcomas. There is no proven genetic predisposition to the development of soft tissue sarcomas though Fraumani have reported kindreds with pairs of young children with soft tissue sarcomas, this incidence exceeds that expected on chance basis (0.06). Howard and Casten reported two brothers who developed rhabdomyosarcomas of the orbit. ***[Howard and Casten, 1963]*** Only one in 100 - 400 cases of childhood rhabdomyosarcoma are associated with other relatives developing this cancer. ***[Fraumeni et al., , 1968]***.

There have been several reports of an association of childhood sarcomas with a small increased incidence of other familial cancer, especially breast cancer that tended to occur in mothers younger than 30 years of age. ***[Fraumeni, et al., 1968]*** the lymphangiosarcoma of the arm in women after mastectomy and axillary lymph nodes obtain almost certainly doesn't represent evidence of an etiologic correlation between mammary cancer and sarcoma of soft tissue but rather the development of lymphangiosarcoma in lymphedematous arm ***[Stewart and Treves, 1981]***.

Sloone and Hubbel have repoted an increased incidence of congenital defects in children with soft tissue sarcomas although this association was not seen by Li and Fraumeni. ***[Sloan and Hubbel, 1969]***

Soft tissue sarcomas are thought to occur with slightly increased frequency in patients with a variety of genetically transmitted disease such as the basal cell nervous syndrome, patients with multiple neurofibromatosis (Von ReckLinghousen's disease) have approximately 15% chance of developing neurofibro-sarcoma, ***[Heard, 1963]***.

Trauma

Although patients with soft tissue sarcomas may present with recent history of trauma, there is no known etiologic relation-ship, and it is likely that minor trauma nearly calls a pre - existing lesions to the patient attention ***[Rosenberg et al., 1989]***.

Chemicals

Chemical carcinogens such as 3-methylcholanthrene can pre-dispose to soft tissue sarcomas in experimental animals but there is no convincing link between these factors and sarcomas in humans. ***[Rosenberg et al., 1989]***.