CLINICO-EPIDEMIOLOGICAL STUDY OF SOFT TISSUE SARCOMAS

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of Radiotherapy

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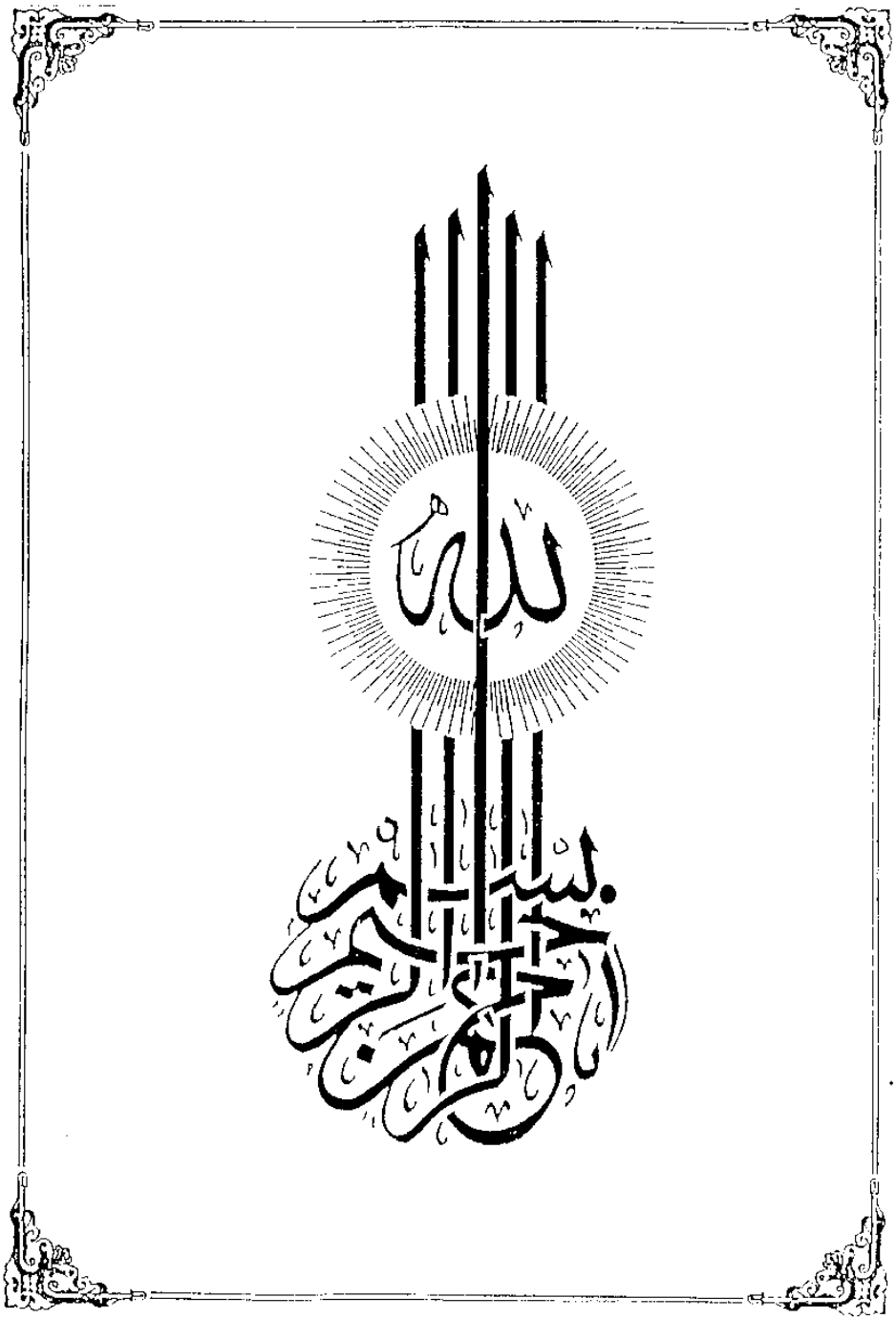

Contents

AIM OF THE WORK

The study of epidemiological pattern and clinical aspect of soft tissue sarcoma in Egyptian patients referes to Radiotherapy Department of Ain Shams University in the period from January 1980 to December 1985.

Also this study include the prelevence of varies type of soft tissue sarcoma in relation to age and sex, and evaluation of the treatment by surgery , radiotherapy and chemotherapy.

Introduction

Historical review :

The ward "Sarcoma" is derived from the Greek and means "Fleshy tumour" (Stout, 1961). Soft tissue sarcomas (S.T.S) can be defined as non-epithelial extra-skeletal tissue of the body exclusive of the reticuloendothelial system, glia, and supporting tissue of various parenchymal organs, (Enzinger , 1969). It is represented by the voluntary muscles, fat and fibrous tissue, along with the vessels serving these tissues. By convention it also includes the peripheral nervous system because tumours arising from nerves present as soft tissue masses. Embryologically, it is derived principally from mesoderm with some contribution from neuroectoderm. S.T.S. are classified on a histogenetic basis according to the adult tissue they resemble. Sarcomas are Locally aggressive tumours that are also capable of distant metastasis. Unfortunately , the term sarcoma does not indicate the likehood or rapidly of metastasis. Some sarcomas such as the myxoid Liposarcoma, metastasize infrequenty and at a relatively Late state of the disease, while others such as the malignant fibrous histocytoma, do so with alacrity. For these reasons it is important to qualify the term sarcoma with a statment concerning the degree of differentiation or the histological grade. The term was first introduced in 1963 to refer to a group of soft tissue tumours characterized by storiform or Cart wheel-like growth pattern (Franz, Sharon, 1983).

Haemangiopericytoma is a rather uncommon neoplasm that was first discribed and named by Stout and Murray, 1942.

In the original description the authors postulated that the tumour is composed mainly of pericytes, a specific cell

type first identified by Zimmermann in 1923.

In 1872 Kaposi, discribed five Cases of an unusual tumour that principally affect the skin of the Lower extremities in a multifocal and often symmetrical fashion. He considered the condition a round cell sarcoma that he termed "idiopathic multiple pigmented sarcoma" of the skin. Since his discription, others have embraced the belief that this unusual lesion, which often has an indolent waxing and waning course, is not a neoplasm but an infectious disease. It is now generally accepted that Kaposi's Sarcoma is a neoplasm, (Franz, Sharon, 1983).

Epidemiology of S.T.S.

Incidence:

S.T.S. are rare, they constitute only about 1% of all malignant tumours, yet they account for approximatly 2.1 % of all death from malignant disease, (James, 1982).

According to the National Cancer Survey, 4500 new cases occured in the United States in 1976 compared with 93,000 cases of lung cancer and 88,700 cases of breast cancer,(Franz,Sharon, 1983).

In Egypt, malignant tumours of connective tissue account for 7 % of total childhood reported cases and 3 % of total malignant cases in 1979 (Abou El Nesr, 1982).

Table (1) illustrates the relative frequency incidences of various pathological subtypes of S.T.S. Among 1215 Sarcomas reviewed and staged by Task Force of S.T.S. there were 234 (19%) rhabdomyosarcomas (R.M.S.) (Russel, 1977). Also Masson and Soule, 1965 had reported 19% among 2000 cases of S.T.S. to be R.M.S. According to Young and Millar, 1975, R.M.S. incidence in the general population is 4.5 per million per year for white children and 1.3 per million per year for black children, and at Least 350 new cases of R.M.S. occur in the United States every year.

Leiomyosarcomas account for about 7% of S.T.S (Russell, 1977).

Liposarcomas account for about 16% (Reszel, 1966), to 18 % (Russell, 1977), among S.T.S. but values as Low as 5% to 6% have also been given (Evans, 1979). Kindblom, et al , 1975,

reported an annual incidence of 2.5 per million in Sweden. It is very likely that future accounts of this neoplasm will reveal a slightly Lower incidence because a good number of tumours formerly diagnosed as Liposarcoma are naw classified as malignant fibrous histocytoma (France, Sharon, 1983).

Fibrosarcomas ranked third in frequency following liposarcoma and R.M.S. It account for 12% of 2310 S.T.S. This apparent decline in the incidence of fibrosarcoma may be accounted for by the introduction of malignant fibrous histocytoma as a specific tumours type, and by the separation of fibromatosis as a specific entity, intermediat in its behavior between a benign fibrous proliferation and a fibrosarcoma, (Franz , Sharon, 1983).

Malignant Fibroushistocytoma constitutes 10.5% of all S.T.S.

Malignant Schwannoma is the principle malgnancy of peripheral nerve, yet it remains one of the most elusive and poorly defined of all S.T.S. There is general agreement that if a sarcoma arises from a peripheral nerve or a neurofibroma it can usually be considered a malignant schwannoma. As a result the incidence of this disease is uncertain and varies considerably in the literature. Malignant Schwanoma accounts for approximatly 10% of all S.T.S., less than half occured with neurofibromatosis (Franz, Sharon,1983), and one fourth (Ghosh Bc, 1973) to over two thirds (WhiteHR, 1971), were reported to be associated with Von Reckilinghousen's disease.

The incidence of synovial Sarcoma observed by Pack, 195o at Memorial Hospital was 8.4 % among all malignant

tumours of the S.T.S, others report an incidence of 6.9 % (Russel Wo, 1977), 9.6 % (Bstch, Zentrable, 1959), and 10% (Cadman, 1965) of all S.T.S.

Extraskeletal osteosarcoma represented an incidence of 1.2 % of all S.T.S (Allan, 1971) and an incidence 4.6 % of all osteosareomas.

Extraskeletal Ewing's sarcomas is still of a limited information on it, at 1925 series of 39 cases only was reported by Kinsella, 1983 & Arnold J, 1985.

Extraskeletal mesenchymal chondrosarcoma is a rare tumour. It about three times more common in bone than in soft tissue (Dahlin, 1962).

Angiosarcoma are collectivelly one of the rarest forms of S.T.S. They account for a vanishingly small proportion of all vascular tumours and they comprise less than 1% of all sarcomas as estimated by a 20 year study at the M.D. Andrson Hospital (Bordwil, 1968). Angiosarcoma of the breast,accounting for 1 out every 1700 to 2000 primary malignancies of the breast (Mc Clanahan, 1954 and Mc Divitt,1966).

The geographical distribution of Kaposi's Sarcoma in Africa is similar, although not identical, to that of Burkitt's lymphoma. Although it is quite rare in most parts of the world, including the United States (where it accounts for 0.02 % of all malignancies (Lothef, 1963). It is relatively more common in Poland, Russia, Italy, and the central equatorial region of Africa. In central Africa it appears to be endemic and may

account for up to 9% of all reported malignancies (Niemi , 1965). In fact the incidence of the disease in the white population in the endemic region of Africa in general suggest that the epidemiology of the disease may be complex (Franz , Sharon, 1983).

Malignant endovascular papillary Angioendothelioma is a rare but distinctive vascular tumour discribed in 1969 by Dabska, on the basis of her six cases, it appeared to be a form of Low-grade angiosarcoma occuring in the skin or subcutis of infant and young children.

Alveolar soft part sarcoma is a uncommon neoplasm, its frequency is estimated as between 0.5 % and 1.0 % of all S.T.S. (Franz, Sharon, 1983).

The peripheral neuroepithelioma (peripheral neuroblastoma) is a primitive neuroectodermal tumour arising from peripheral nerve. This tumour exceedingly rare. The incidence of peripheral neuroblastoma is less than 1% of all malignant tumours of peripheral nerve (Joachim, 1975).

Malignant mesothelioma is a rare neoplastic disease arising in the mesothelium, mainly of the pleura and peritoneum. The disease accounts for less than 0.3 % of total number of cancer cases in Denmark (M. Andersson & J.H. Olsen, 1985)- 3:7.1/ million at 1970-1975 in USA (Hinds, 1978)- and 3.4/ million at 1969-1978 at England (Gardner, 1982).

Table (1)

Relative frequency incidence of S.T.S.

Type	%
R.M.S.	19
Leiomyosarcoma	7
Liposarcoma	16 - 18
Fibrosarcoma	12
Malignant fibrous histocytoma	10.5
Malignant schwannoma	10
Synovial sarcoma	8.4
Extra skeletal osteosarcoma	1.2
Angiosarcoma	1
Alveolar soft partsarcoma	o.5 - 1
Peripheral neuroepithelioma	< 1
Malignant mesothelioma	0.3
Kaposi's sarcoma	
* Central Africa	9
* United State	0.02

Age distribution:

Table (2) illustrates that,rhabdomyosarcoma has a fairly uniform age incidence. It is not only the most common S.T.S. of children under 15 years of age but to some what less frequently among adolescents and young adults. These tumours occurred mainly in the muscles of the Lower extremity and affected patients between 50 and 70 years of age. About 2 % or 3% of all cases are present at birth. R.M.S. are rare in adults beyond age 45. The median age varies slightly, depending on both the histological type and the anatomical location, this pleomorphic type affects the older age groups, the majority of cases occuring in patients over 30 years age. Alveolar R.M.S. occuring mostly in adolescents and young adults (Enzinger, 1969- Horn, 1864 - and Sutow, 1970). Embryonal type usually occurs in youngesters under the age of 10 years (Mahour, 1967), while Botryoidal type occurs in young children at the average age of 7 years over 30 years (James, 1981).

Table (2): Age distribution of S.T.S.

Type	Age incidence in years
. R.M.S.	
Embryonal	< 10
Pleomorphic	> 30
Botryoidal	7 & > 30
Alveolar	> 15
. Liposarcoma	40 - 70 10 - 15
. Fibrosarcoma	30 - 55
. Malignant fibrous histocytoma	18 - 84
. Malginant schwannoma	20 - 50
. Synovial sarcoma	15 - 35
. Extra skeletal osteosarcoma	20 - 71
. Ewing sarcoma	15 - 30
. Mesenchymal chondrosarcoma	15 - 35
. Hemangiopericytoma	20 - 70
. Kaposi's sarcoma	60 - 70
. Alveolar soft part sarcoma	15 - 35
. Mesothelioma	50 - 70
. Epitheliod sarcoma	15 - 35
. Clear cell sarcoma	20 - 40

Fig(I): Approximate relationship of age to incidence of various types of sarcomas. Continous line indicates peak incidence of tumour. Dotted line indicates reuced incidence of tumour.