Original Article

Diversity in the spectrum of malignant soft tissue tumors

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Abstract

Soft tissue tumors are a highly heterogenous group of tumors that are classified on a histogenetic basis. Benign tumors far outnumber the malignant ones which form less than 1 % of all cancers. The most common sites are extremities, chest wall, mediastinum and retroperitoneum and as in the case of other malignancies, occur mainly in the older age group except rhabdomyo sarcoma. These tumors present a varied histomorphological pattern. Our study has included the different patterns, with specific site, gender and age distribution.

Keywords: Soft tissue tumors, malignant.

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INTRODUCTION

Most, if not all soft tissue sarcomas arise from partially commited primitive, multipotential mesenchymal precursor cells, which in the course of neoplastic transformation undergo differentiation along one or more lines. Traditionally they have been classified according to a histogenetic concept eg fibrosarcoma as a malignant tumor arising from the fibroblasts, liposarcoma as a tumor of lipoblasts and so on. This classification based on differentiation correlates with clinical parameters like location, pattern of growth, incidence, patient's age, therapeutic response and prognosis.

MATERIAL AND METHODS

All malignant soft tissue tumors received in the department of pathology, in two institutions over a 2 year period were included in the present study. The clinical details were recorded and the slides reviewed. The sections studied were from paraffin embedded blocks stained with haematoxylin and eosin.

RESULTS

A total of 106 soft tissue tumors were received during this period, of which 19 were malignant. The most common malignant tumors were rhabdomyosarcoma (6 cases), Liposarcoma (4 cases) and Pleomorphic undifferentiated sarcoma(malignant fibrous histiocytoma) (3 cases). Of the 20 cases, 11 cases were found in males and 9 case in female patients. Most of the malignant tumors were seen in middle aged patients (13 cases) and most of the cases of Rhabdomyosarcoma were in children. Lower extremity especially thigh was the most common site of malignant soft tissue tumors in this study. The gender specific, age specific and site specific distribution of tumors in our study is formulated in table I,II and III.

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Table 1: Gender distribution

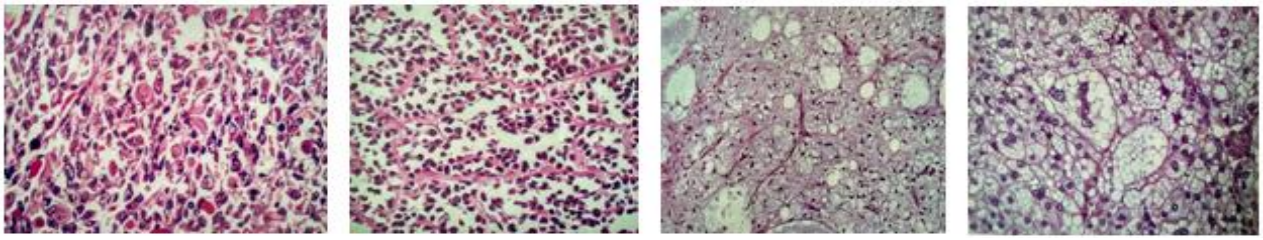
Sr. No	Tumour	Male	Female	Total
1	Embryonal rhabdomyosarcoma (rms)	1	1	2
2	Botryoid rhabdomyosarcoma	0	1	1
3	Alveolar rhabdomyosarcoma	1	2	3
4	Liposarcoma	2	2	4
5	Pleomorphic undifferentiated sarcoma/malignant fibrous histiocytoma (mfh)	3	0	3
6	Primitive neuroectodermal tumour (pnet)	2	0	2
7	Malignant peripheral nerve sheath tumour (mpnst)	0	2	2
8	Synovial sarcoma	1	0	1
9	Fibrosarcoma	0	1	1

Table 2: Age distribution

Pediatric (<15 years)	Rhabdomyosarcoma – Embryonal	2
	- Botryoid	1
	- Alveolar	2
	Malignant Peripheral Nerve Sheath Tumor (MPNST)	1
Young adults (16-30 years)	Liposarcoma	4
	Pleomorphic undifferentiated sarcoma/Malignant Fibrous Histiocytoma (MFH)	3
	Primitive neuroectodermal tumour (PNET)	2
	Fibrosarcoma	1
	Alveolar Rhabdomyosarcoma	1
Middle Age (31- 60 years)	Synovial sarcoma	1
	Malignant Peripheral Nerve Sheath Tumor (MPNST)	1

Table 3: site-specific distribution

UPPER LIMB	Pleomorphic undifferentiated sarcoma/Malignant Fibrous Histiocytoma (MFH)	1
	Synovial sarcoma	1
	Primitive neuroectodermal tumour (PNET)	1
	Malignant Peripheral Nerve Sheath Tumor (MPNST)	1
THIGH	Liposarcoma	4
	Pleomorphic undifferentiated sarcoma/Malignant Fibrous Histiocytoma (MFH)	1
	Malignant Peripheral Nerve Sheath Tumor (MPNST)	1
LEG	Fibrosarcoma	1
Chest Anterior chest wall	Primitive neuroectodermal tumour (PNET)	1
Back	Pleomorphic undifferentiated sarcoma/Malignant Fibrous Histiocytoma (MFH)	1
	Alveolar Rhabdomyosarcoma	1
Perineal region	Liposarcoma	1
	Botryoid Rhabdomyosarcoma	1
	Alveolar Rhabdomyosarcoma	1
Head and neck	Embryonal Rhabdomyosarcoma	1
	Alveolar Rhabdomyosarcoma	1
Gluteal region	Embryonal Rhabdomyosarcoma	1



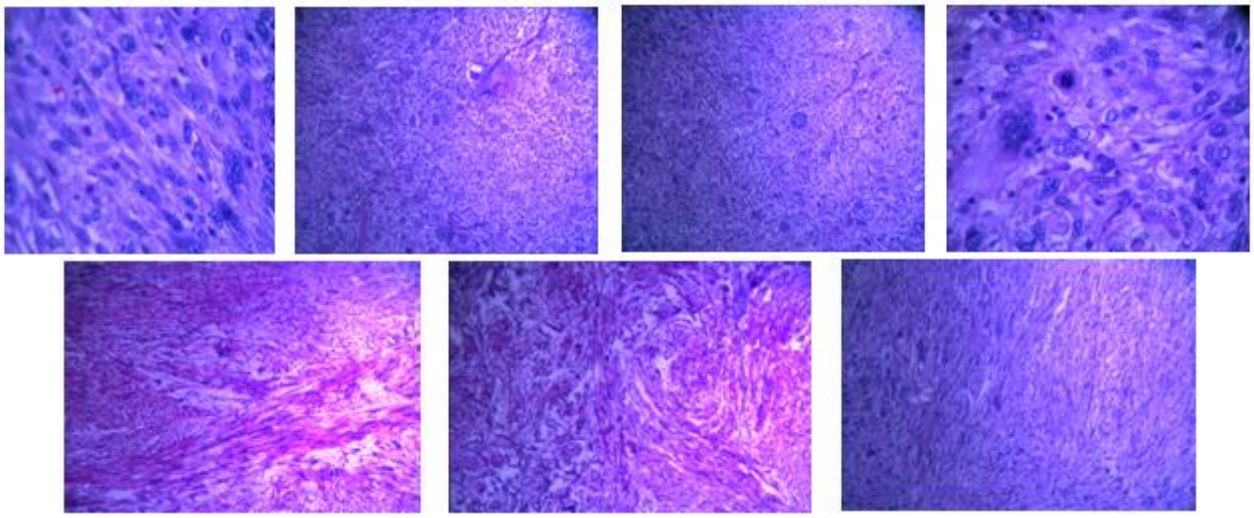


Figure 1: Embryonal Rhabdomyosarcoma-Tumor cells with eosinophilic cytoplasm.
Figure 2: Alveolar Rhabdomyosarcoma-Nests of small round tumor cells separated by connective tissue septae.
Figure 3: Myxoid Liposarcoma - Tumor cells in mucoid matrix with chicken wire appearance of capillary network.
Figure 4: Pleomorphic Liposarcoma.
Figure 5.0: Pleomorphic Undiffrentiated Sarcoma (MFH)- Pleomorphic spindle cells in vague storiform pattern. 40X
Figure 5.1: Pleomorphic Undiffrentiated Sarcoma (MFH) with Giant cells
Figure 5.2: Pleomorphic Undiffrentiated Sarcoma (MFH) with Giant cells.
Figure 5.3: Pleomorphic Undiffrentiated Sarcoma (MFH) with Giant cells and mitosis.
Figure 6.0: Malignant Peripheral Nerve Sheath Tumor (MPNST) - cellular fasicles of spindle cells.
Figure 6.1: Malignant Peripheral Nerve Sheath Tumor (MPNST) - cellular fasicles of spindle cells.
Figure 7: Fibrosarcoma- Intersecting fascicles of spindle cells (Herringbone appearance).

DISCUSSION

Soft tissue tumors constitute a large and heterogenous group of neoplasms.

A definite relationship exists between the type of tumor and the age of presentation. Embryonal Rhabdomyosarcoma is typically a tumor of infants and children. Pleomorphic undifferentiated sarcoma (malignant fibrous histocytoma) and liposarcoma are the most common soft tissue sarcomas of middle aged adults. Synovial sarcoma mainly affects adolescents and young adults.

The majority of soft tissue sarcomas arise de novo. Recognized causes include exposure to ionizing radiation, inherited or acquired immunologic defects. Trauma is frequently implicated in the development of sarcoma but no convincing evidence is provided. Exposure to phenoxy herbicides as the causative factor for some tumors has been suggested. There is a strong evidence suggesting the possibility of Human herpes virus (HHV8) as the causative agent of Kaposi sarcoma and Ebstein – Barr virus causing malignant smooth muscle tumors in patients with immuno deficiency syndromes or following therapeutic immunosuppression in the transplant setting. Rare soft tissue sarcomas have been reported as arising in scar tissue following surgical procedures or thermal or acid burns, at fracture sites and in the vicinity of plastic or metal implants, after a latent period of several years.

Light microscopic evaluation of heamatoxylin – eosin stained sections remains the standard technique for the diagnosis of these tumors and is sufficient in the majority of the cases. However there are special techniques like immuno histochemistry, molecular cytogenetic methods, conventional special stains to increase the diagnostic accuracy. The commonly used histochemical stains are Masson's trichrome for tumors of striated muscle, Periodic acid –Schiff (PAS) for demonstrating the intracytoplasmic crystals in alveolar rhabdomyosarcoma, Reticulin stain in vascular tumors, Mucin and reticulin stains in synovial sarcoma, to mention a few.

The two grading schemes that have been widely applied are the French Federation of Cancer Centres Sarcoma Group and NCI (National Cancer Institute).The grading is based on the evaluation of three separate parameters: tumor differentiation (score 1-3), mitotic count (score 1-3) and the amount of tumor necrosis (0-2). Grading is set up after summing up the scores. Score 2 or 3 is Grade I, Score 4 or 5- Grade II, Score 6 or 7 or 8 is Grade III.

Two main staging systems are proposed. American Joint committee (AJCC) based on TNM system- size of the primary tumor (T), status of lymph nodes (N), presence of distant metastasis (M) and tumor's histological grade (G).

Enneking system is suitable for lesions in the extremities. Staging is done based on anatomic settings (T1, intracompartmental or T2, extracompartmental) grades (G1- low or G2- high)and presence or absence of metastasis. The staging systems serve as valuable guide to therapy and provide useful prognostic information. Obviously staging soft tissue sarcomas require multidisciplinary approach with close cooperation among Clinician, Oncologist and Pathologist.

The prognosis of the soft tissue sarcomas depends on the tumor size and depth, location, the histological type, microscopic grade and the surgical margins. Evaluation of proliferation markers MIB-1 and P105 correlates with prognosis. DNA anuploidy correlates with a higher microscopic grade, higher rate of cell proliferation and decreased survival rates.

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