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Original Research Article

Histo-Morphological Study of Soft Tissue Tumors

Meeta Thomas[1], Sharmila Annadurai[2]

[1,2]Assistant Professor, Department of Pathology, Pushpagiri Institute of Medical Sciences & Research Center, Thiruvalla, Kerala 689101, India.

Corresponding Author:
Sharmila Annadurai, Assistant Professor, Department of Pathology, Pushpagiri Institute of Medical Sciences & Research Center, Thiruvalla, Kerala 689101, India.

E-mail: drsharmila.medico@gmail.com

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Abstract

Background: Soft tissue tumours (STT) represent a heterogeneous and complex group of mesenchymal lesions that may show a broad range of differentiation. Inspite of their pathological diversity, there is a significant overlap in their clinical and radiological features which highlights the importance of an accurate histopathological diagnosis. *Objective:* The present work aims to study the histomorphological patterns of soft tissue tumors and to correlate it with clinical presentation and epidemiological distribution according to age, sex and site of occurrence. *Methods:* 190 cases of patients with soft tissue tumors attending the surgery department in Government Medical College Kottayam were included in the study. Clinical features including symptoms and duration of the swelling were documented. Gross as well as microscopic features (after routine H and E stain) were studied in detail. *Results:* Soft tissue tumours were seen to comprise 2.78% of all biopsies received. The most common mode of presentation was a painless swelling of prolonged duration. Of the 190 cases studied, 162 cases (85.26%) were benign, 18 cases (9.47%) were malignant and 10 cases (5.26%) were those of intermediate category.

Keywords: Soft tissue tumours; histomorphology.

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Introduction

Soft tissue tumours (STT) represent a heterogeneous and complex group of mesenchymal lesions that may show a broad range of differentiation. The large majority of STTs are benign, with a high cure rate after surgical excision. Malignant mesenchymal neoplasms amount to less than 1% of the overall burden of STTs but recent data has shown an upward trend in the incidence of soft tissue sarcomas. These soft tissue sarcomas are life threatening and pose a significant diagnostic and therapeutic challenge since there are more than 50 histological subtypes of ST sarcomas which are associated with some unique clinical, prognostic and therapeutic features.

In comparison with other forms of cancer, soft tissue tumors are relatively rare, hence most

pathologists lack adequate experience with the histopathological and cytopathological features of these tumors. Also, soft tissue tumors are associated with a bewildering spectrum of morphological appearances and there is a considerable overlap in the morphology of benign and malignant lesions.

The close interaction of pathologists, surgeons and radiologists is essential and this approach will contribute to a significant increase in disease free survival for tumors which were previously almost invariably fatal.

The present work aims to study the histomorphological patterns of soft tissue tumors and to correlate it with clinical presentation and epidemiological factors.

Materials and Methods

One hundred ninety (190) cases of patients with soft tissue tumors attending the surgery department in Government Medical College Kottayam were included in the study. Gross features including size, appearance, presence of areas of necrosis and hemorrhage were documented. Representative sections were taken, routine H & E stain was performed and subjected to histopathological examination. The tumors were subtyped on the basis of WHO classification.

Observations & Results

One hundred ninety (190) cases of soft tissue tumors were studied over a period of 18 months. Soft tissue tumors were seen to comprise ~2.78% of all biopsies received.

Of the 190 cases of soft tissue tumors studied 98 were males (52%), showing a slight male preponderance.

The age range of the group varied from 3 yrs to 83 yrs with a mean age of 40.8 years. Majority of the soft tissue tumors occurred in the fifth decade (29%).

The most common site involved was the extremity 79 cases (41.58%), followed by trunk 68 cases (35.79%) and head and neck 43 (22.63%).

Of the 190 soft tissue tumors studied, 162 cases (85.26%) were benign, 18 cases (9.47%) were malignant and 10 cases (5.26%) were those of intermediate category. Thus, the benign tumors outnumbered the malignant tumors in a ratio of 9: 1.

Of the 190 cases studied, a vast majority 84 (44.21%) were of lipomatous origin, followed by neural tumors which constituted 45 (23.68%) cases, vascular tumors constituted 24 (12.63%) cases, fibrous and fibrohistiocytic tumors constituted 26 (13.7%) cases, skeletal and smooth muscle tumors constituted 5 (2.63%) cases and tumors of uncertain differentiation which constituted 6 (3.16%) of all cases.

Of the adipocytic tumors, 81 were benign tumours including histological variants like fibrolipoma, angiolipoma, pleomorphic lipoma and lipoblastoma. Of the 3 malignant tumours, 2 were well differentiated liposarcomas and were retroperitoneal in location while 1 was a myxoid liposarcoma and was situated in the deep soft tissue of the leg. Both benign and malignant tumours were seen to occur predominantly in the 5th and 6th decades of life.

The mean age for presentation of benign lipomatous tumors was 43 yrs with an almost equal incidence in males and females. Most of the lipomas presented as slow growing painless masses and multiplicity was seen in only a minority of cases (i.e., 3 cases). Lipomas were seen to have a ubiquitous distribution in the body. The size of the tumors was seen to range from 2 cm to 30 cms. FNAC was done in 23 cases with a positive correlation in 19 cases with the rest being inconclusive.

Lipocarcomas were seen to have a median age of occurrence of 46 yrs and were seen to occur most commonly in the retroperitoneum followed by the deep soft tissue of the extremities. Also, well differentiated liposarcoma was found to be the most common histological subtype. The size of the tumor was seen to vary from 5 to 35 cms based on the location i.e., the tumors of the extremities being detected earlier were found to be smaller in size. The tumors were assessed preoperatively by radiological studies which accurately assessed the malignant nature of the lesion. The tumors were not subjected to FNAC possibly due to their deep location.

The next most common tumours in order of frequency were those of peripheral nerve sheath origin, which constituted 45 cases of which 44 cases were benign (24 neurofibromas and 20 schwannomas) and 1 was a malignant peripheral nerve sheath tumour. The neurofibromas included the diffuse and plexiform variants and the plexiform variant was found to be strongly associated with NF–1. The schwannomas included the classical and cellular types. The cellular type was found to be firmer with fewer myxoid areas grossly than the classical type. The most common presenting

symptom in decreasing order of frequency were: mass lesion (62.5%), pain with or without a mass lesion(22.5%), and neurological deficit(15%). The most common site of involvement was the head and neck (37%), followed by the upper limb (25.9%), lower limb (14.8%) and trunk (7.4%). Multiple tumours were seen in 14.8% cases.

The vascular tumours were the 3rd most common soft tissue tumours, comprising about 24 cases, 3 of which were of pericytic origin. About one half of the cases were seen in the first 2 decades of life with the rest being distributed from the 4th to 6th decades of life. The pericytic tumours were seen mainly in the older age group (5th and 6th decades). The single malignant vascular tumour was an epitheloid hemangioendothelioma.

The vascular tumours were seen to occur predominantly in the distal extremities. Most of them presented as painful swellings of prolonged duration. The mean age of presentation was 27 yrs. Because of their vascular nature, FNAC in most cases proved to be inconclusive due to the bloody aspirate obscuring the cells.

Of the 20 fibrous tumours included in the study, 7 cases were benign, 10 were of intermediate category and 3 were malignant in nature. The benign cases were predominantly fibromas including nuchal type, sclerotic and collagenous fibromas and 1 was a case of nodular fasciitis. The intermediate category lesions included 5 cases of fibromatosis, 3 cases of desmoid tumour and 2 cases of solitary fibrous tumour. The malignant tumours were 1 case each of Low grade fibromyxoid sarcoma, myxofibrosarcoma and myxoinflammatory fibroblastic sarcoma. The benign as well as malignant fibrous tumours showed a wide age of distribution from the 2nd to 7th decades of life.

The mean age of presentation was 36 yrs with onset of a painless swelling and a history of fairly rapid increase in size. FNAC in most cases was paucicellular possibly because of the fibrous nature of the lesion and could only suggest a spindle cell lesion. Abdominal fibromatosis was seen to occur mostly in females while superficial fibromatosis was seen to occur predominantly in males. Low grade fibromyxoid sarcoma was seen to occur in the deep soft tissues of young adults while myxofibrosarcoma was seen to occur in the superficial soft tissue of elderly individuals.

The fibrohistiocytic tumours comprised 6 cases all of which were benign, including 4 cases of benign fibrous histiocytoma and 2 cases of giant cell tumour of tendon sheath.

The tumours of smooth and skeletal muscle origin were relatively rare with majority of them being malignant. There were 3 cases of leiomyosarcoma of which 1 was seen originating from the wall of the IVC. The only tumour of skeletal muscle differentiation was a rhabdomyosarcoma occurring in the upper limb of a 3 year old child.

Among the tumours of uncertain differentiation were 4 cases of synovial sarcoma and 1 clear cell sarcoma. All 4 cases of synovial sarcoma were seen to occur between the 2nd to 4th decades of life with the site of occurrence of all being the lower limb.

The synovial sarcomas were seen to present as painless swellings with a rapid increase in size. FNAC was done in one of the cases and was reported as a spindle cell sarcoma. One of our patients who underwent an above knee amputation, presented with metastases to the lung which was resected and following which he developed abdominal wall metastases (diagnosed cytologically). Clear cell sarcomas were also seen to occur in the same age group and with a similar presentation.

Mode of Presentation

The most common mode of presentation was presence of a painless swelling of prolonged duration with a history of gradual increase in size.

Associated pain was present in about 11% of cases and was especially seen in the vascular and some neural tumors.

Other associated symptoms were non-specific (Constipation, abdominal distension, loss of weight, loss of appetite) and varied according to the site of the lesion.

Duration of symptoms

The mean duration of symptoms before the patient sought medical attention was :

27 months for lipomatous tumors

23 months for neural tumors

8 months for fibrous tumors

12 months for vascular tumors

15 months for myogenic tumors

8 months for tumors of uncertain origin

Discussion

The current study included a total of 190 cases of soft tissue tumours of which 162 cases (85.26%)

were benign, 18 cases (9.47%) were malignant and 10 cases(5.26%) were those of intermediate category. Thus, the benign tumors outnumbered the malignant tumors in a ratio of 9:1. These findings are in concordance with the studies by Harpal *et al*.[1] and Agravat *et al*.[2] however M.J. Kransdorf *et al*.[3,4]

Table 1:

S. No.	Study	Total cases	Benign	Intermediate	Malignant
1.	Agravat *et al.*	100	86%	2%	6%
2.	Harpal *et al.*	200	84.5%	5.5%	10%
3.	Present study	190	85.26%	5.26%	9.47%

reported 60.2% benign and 39.8% malignant soft tissue tumour in their study (Table 1).

Of the 190 cases of soft tissue tumors studied 98 were in males (52%), showing a slight male preponderance (1.06:1). This observation is in concordance with the studies by Harpal *et al.*[1] (M:F = 1.1:1) and Jemal *et al.*[11] (M:F = 1.2:1).

The age range of the group varied from 3 yrs to 83 yrs with a mean age of 40.8 years. Majority of the soft tissue tumors occurred in the fifth decade (29%). Benign tumours were seen more commonly in younger individuals while malignant tumours were more common in the older individuals with a few exceptions including rhabdomyosarcoma, synovial sarcoma and low grade fibromyxoid sarcoma which were seen to occur in children and young adults.

The most common site involved was the extremity in 79 cases (41.58%), followed by trunk 68 cases (35.79%) and head and neck 43 cases (22.63%). This was in concordance with the study by Abbas *et al*.,[10] in which 49% cases were located over extremities, 26% cases over trunk, 10% cases over head and neck region, and rest in the retroperitoneum.

Majority of the benign soft tissue were found to be superficial in location, while malignant tumors were deep.

Also the soft tissue sarcomas were seen to have a strong predilection for the limbs (50%) especially the lower limbs (44%) followed by trunk (38%). This is in concordance with the studies by Kransdorf[3] and Pramila *et al*.[7]

Of the 190 cases studied, a vast majority 84 (44.21%) were of lipomatous origin, followed by neural tumors which constituted 45 (23.68%) cases, vascular tumors constituted 24 (12.63%) cases, fibrous and fibrohistiocytic tumors constituted 26 (13.7%) cases, skeletal and smooth muscle tumors constituted 5 (2.63%) cases and tumors of uncertain differentiation which constituted 6 (3.16%) of all cases. This is in concordance with the study by Baste *et al*.,[8] however, in the studies by Harpal *et al*.[1] and Agravat *et al*.[2]% the 2nd most common tumours were of vascular origin and were followed by the neural tumours.

Among the different histological subtypes, liposarcomas were seen to occur most commonly in the retroperitoneum followed by the deep soft tissue of the extremities which is in contrast to the study by D Nemanqani, WA Mourad *et al*.[12] Also in contrast to the same study, well differentiated liposarcoma was found to be the most common histological subtype.

Among the neural tumours, the plexiform variant was found to be strongly associated with NF-1 as in the study by Sushma Gabhane *et al*.[13]

Among the tumors of fibrous origin, abdominal fibromatosis was seen to occur mostly in females while superficial fibromatoses were seen to occur predominantly in males as in the study by JS Wang, HC Liu.[15] Also as in the study by Oda Y, Takahira T *et al*.,[16] low grade fibromyxoid sarcomas were seen to occur in the deep soft tissues of young adults while myxofibrosarcomas were seen to occur in the superficial soft tissue of elderly individuals.

FNAC was performed in about one fourth of the cases with diagnostic yield being maximum in benign lipomatous tumours[21] and some histological variants like pleomorphic lipoma could also be diagnosed cytologically. Also some of the malignant lesions could be identified as sarcomas cytologically enabling preoperative triaging of these lesions. However, the precise cytological diagnosis of fibrous and neural lesions was limited by factors including low cellularity of the aspirate and overlapping cytomorphological features as a result of which only a diagnosis of a spindle cell lesion could be rendered in most of the cases. The diagnosis of vascular lesions was limited by the bloody nature of the aspirate.

FNAC was found to be especially effective tool in diagnosing recurrent and metastatic lesions[22] in patients with a known primary.

Conclusion

Soft tissue tumors form a varied and complex group which may show a wide range of differentiation. They are rare compared to epithelial neoplasms. Diagnosis and classification of soft tissue tumors is one of the most challenging areas in surgical pathology.

All diagnostic modalities for evaluation of soft tissue tumors are complementary and an ideal diagnostic pathway must involve a multidisciplinary approach with inputs from the clinical history and radiological findings.

A total of 190 biopsy samples were included in the present study. The occurrence of soft tissue tumors was found to show a slight male preponderance. Benign soft tissue tumors constituted 162 of the total cases, thus outnumbering the malignant cases by a ratio of about 9:1.

Maximum number of cases were observed in the 5th decade and the majority of the cases were seen to occur in the extremities.

In our study the bulk of soft tissue tumours comprised of lipomatous tumours followed by neural tumours, vascular tumours, fibrous tumours and myogenic tumours in descending order of frequency.

The clinical diagnosis was found to correlate with the histology in 134 cases, i.e., 70.53% of all cases.

The diagnostic yield of FNAC was found to be maximum in benign lipomatous tumours and aspiration cytology was seen to be especially useful in the diagnosis of recurrent and metastatic lesions.

Soft tissue tumors are an area with relative paucity of literature due to multiple factors, however they are not uncommon and an accurate diagnosis involves a multidisciplinary approach. This study emphasizes the need for such studies where extensive clinical history, findings of physical examination are correlated with detailed cytological and biopsy findings.

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