



HISTOPATHOLOGICAL SPECTRUM OF SOFT TISSUE TUMORS: A THREE YEAR EXPERIENCE

Pathology

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ABSTRACT

Background: Soft tissue is composed of fibrous (connective) tissue, adipose tissue, skeletal muscle, blood and lymph vessels and peripheral nervous system. Soft tissue tumors are diverse group of lesions that arises from the supporting soft tissue of the body. This study aims to analyse the histopathological findings of soft tissue tumors and their distribution according to age, sex and site of occurrence in patients.

Materials and Methods: This retrospective and prospective comparative study was carried on soft tissue specimen received over the period of 3 years (March 2017 – February 2020) in department of pathology, Rajendra Institute of Medical Sciences, Ranchi, Jharkhand.

Results: A total of 226 specimens were received. Male constituted 128 cases (56.6 %) and females: 98 cases (43.4 %). Age ranges from 2 months to 79 years old. Majority of lesions belonged to benign category: 192 cases (85 %), malignant category: 27 cases (12 %) followed by 7 cases (3 %) of intermediate group. Among benign lesions, adipocytic tumours formed largest group consisting of 93 cases (41.2 %) followed by blood vessels tumors: 32 cases (14.2%) and peripheral nerve sheath and related tumors: 30 cases (13.3 %). Most common malignant lesion was fibrosarcoma: 10 cases (4.2 %)

Conclusion: Benign tumours are more common than malignant lesions. Majority of soft tissue tumors can be diagnosed by hematoxyline and eosin stained sections which are supplemented by immunohistochemistry.

KEYWORDS

Soft tissue tumors, benign, malignant, rhabdomyosarcoma, myxoid chondrosarcoma.

Introduction

Soft tissue is composed of fibrous (connective) tissue, adipose tissue, skeletal muscle, blood lymph vessels and peripheral nervous system. Soft tissue tumors are defined as mesenchymal proliferation which occurs in the extraskeletal non-epithelial tissues of the body, excluding the viscera, coverings of brain and lymphoreticular system.^[1] This heterogenous group of tumors are classified on a histogenetic basis according to the adult tissue they resemble. Genetic factors, environment factors, irradiation, viral infections and immune deficiency have been found to be associated with development of unusually malignant soft tissue tumors. There has also been reports of sarcoma arising from surgical procedures or thermal or acid burns, a fracture site and vicinity of plastic or metal implant usually after latent period of several years.^[2] Carcinogens like asbestos, phenocacetic acid, chlorophenol and their contaminants can also develop sarcoma. Radiation induced soft tissue sarcomas are quite uncommon. The annual incidence of soft tissue tumor is 1.4 per 100000 population.^[3] Soft tissue sarcomas accounts for 15 % of all childhood cancers.^[4] which is the most common malignancy in children after hematopoietic neoplasm, neural tumors and wilms tumor.^[5] Benign soft tissue tumors outnumbered malignant tumors by a wide margin.

Material and Methods

This study was conducted in all soft tissue specimens received in duration of 3 years in the department of pathology, Rajendra Institute of Medical sciences, Ranchi, Jharkhand. It included 2 years of retrospective and 1 year of a prospective study. A total of 226 cases of soft tissue specimen were studied during this period. For the retrospective period (March 2017 to February 2019) all soft tissue cases were taken out from the records of the department and slides were reviewed. In the prospective period (March 2019 to February 2020), all resected specimen received were followed up. Each case was analysed with respect to age, clinical presentation and microscopic diagnosis. The tissue was processed as per standard procedure; 4- 5 microns thick sections were cut and stained by hematoxyline and eosin and special stains in few cases.

Result

During the period of study, we received a total of 226 soft tissue specimens. Number of male patients were 128 (56.6 %) and female patients were 98 (43.4 %); hence the male to female ratio was 1.3:1. (Table. 1) The youngest patient was 2 months old and the oldest being 79 years old. Most common affected age group was second to third decade consisting of 52 cases. (23 %) (Table. 2)

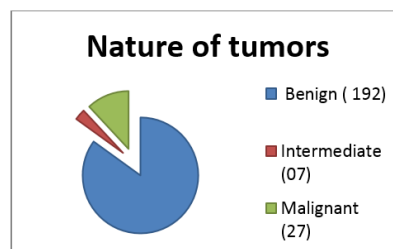
Table.1 : Sex wise distribution of soft tissue tumors

Lesions	Male	Female	Total
Benign	110	82	192 (85 %)
Malignant	04	03	07 (03%)
Intermediate	14	13	27 (12%)
Total	128	98	226 (100%)

Table.2: Age distribution of soft tissue tumours

Age group (Years)	Benign	Intermediate	Malignant
0 – 10	09	-	02
11 – 20	22	01	01
21 – 30	47	01	04
31 – 40	42	01	03
41 – 50	38	02	01
51 – 60	18	02	06
61 - 70	10	-	07
71 – 80	06	-	03
Total	192	07	27

Majority of cases belong to benign group: 192 cases (85 %) followed by 27 cases (12 %) of malignant lesions and 7 cases (03 %) belonged to intermediate group. (Figure. 1) Most common anatomical location of soft tissue tumors are head and neck region having 52 cases (23 %) (Table. 3)



(Figure. 1: Nature of tumors)

Table.3: Anatomical location of benign and malignant lesions

Location	Benign (%) n= 192	Inter-mediate (%) n=07	Malignant (%) n=27
Head and Neck	46 (23.9)	03 (42.8)	04(14.9)
Trunk	40(20.8)	-	01(3.7)
Chest and upper back	27(14.1)	-	03(11.1)

Upper Extremity	32(16.7)	-	03(11.1)
Lower Extremity	38(19.8)	02 (28.6)	12(44.4)
Retroperito-neum	08(4.2)	02 (28.6)	02(7.4)
Genital region	01(0.5)	-	02 (7.4)

Adipocytic tumours were most common lesion among benign tumors constituting 93 cases (41.2 %) followed by 32 cases of blood vessels tumors and 30 cases (13.3 %) of peripheral nerve sheath and related tumors. 7 cases of dermatofibrosarcoma protruberans was found in intermediate group of tumors. Among malignant tumours, fibrosarcoma was most common constituting 10 cases (4.2 %) followed by 3 cases (1.3 %) of undifferentiated pleomorphic sarcoma. (Table. 4)

Table.4: Histopathological subtype of soft tissue tumors

Type	Number of cases
BENIGN LESIONS	192 (85%)
Adipocytic tumors	93 (41.2%)
Classical lipoma	74
Fibrolipoma	16
Angiolipoma	02
Myxolipoma	01
Blood vessel tumors	32 (14.2%)
Hemangioma	18
Angiofibroma	14
Lymph vessel tumors	03 (1.3%)
Lymphangioma	03
Peripheral nerve sheath and related tumors	30 (13.3%)
Neurofibroma	22
Schwannoma	07
Traumatic neuroma	01
Fibrous tumors	06 (2.7%)
Fibroma	03
Fibromyxoma	02
Fibromatosis	01
Fibrohistiocytic tumors	16 (7.1%)
Benign fibrous histiocytoma	10
Dermatofibroma	03
Giant cell tumor of tendon sheath	03
Smooth muscle tumor	01 (0.4%)
Leiomyoma	01
Fibroblastic tumors	01 (0.4%)
Nodular fasciitis	01
Pericytic tumors	03 (1.3%)
Glomus tumor	03
Tumors of extragonal germ cells	05 (2.2%)
Soft tissue teratoma	05
Other tumor like conditions	02 (01%)
Tumoral calcinosis	02
INTERMEDIATE TUMORS	07 (03%)
Dermatofibroma Protuberans	07
MALIGNANT TUMORS	27 (12%)
Adipocytic tumors	
Liposarcoma	02 (01%)
Blood vessel tumors	
Angiosarcoma	02 (01%)
Hemanigiopericytoma	01 (0.4%)
Peripheral nerve sheath and related tumors(PNST)	
Malignant PNST	01 (0.4%)
Fibrous tumor	
Fibrosarcoma	10 (4.2%)
Fibrohistiocytic tumor	
Malignant fibrohistiosarcoma	01 (0.4%)
Striated muscle tumor	
Rhabdomyosarcoma	02 (01%)
Primitive neuro-ectodermal tumor and related lesion	
Extraskeletal Ewings sarcoma	02 (01%)
Undifferentiated sarcoma	
Undifferentiated pleomorphic sarcoma	03 (1.3%)
Cartilagenous tumor	
Myxoid chondrosarcoma	01 (0.4%)
Miscellaneous tumors & tumours of uncertain cell type	
Synovial sarcoma	01(0.4%)
Alvelar soft tissue sarcoma	01(0.4%)

Discussion

In the present study, 226 soft tissue tumors were analysed out of which

192(85%) were benign, 7(03%) were intermediate and 27 (12%) were malignant tumors. This is in concordance with the study conducted by Agravat et al. who analysed 100 cases of soft tissue tumors (n=94) and tumor like lesions (n=6), of which benign tumors formed the major group.[6] In another large scale study of 8686 cases done by Stout , 84.5% cases of benign lesions and 15.5% of malignant lesions. [7](Table. 5)

Table.5: Comparison with various studies

Study	Total cases	Benign (%)	Inter-mediate (%)	Malignant (%)
Stout (1953)	8686	7337 (84.5)	-	1349 (15.5)
Agravat et al (2010)	100	(86)	(02)	(06)
Present Study (2020)	226	192 (85)	7 (03)	27(12)

The age of patient ranged from 2 months to 79 years. Most common benign lesions were found in second decade of life followed by third and fourth decades which in correlation with study of Enzinger and Weiss.[8] Malignant tumors were common in 6th to 7th decade of life. The youngest patient was 2 months old female (soft tissue teratoma) and oldest patient being 79 years old male (angiosarcoma).

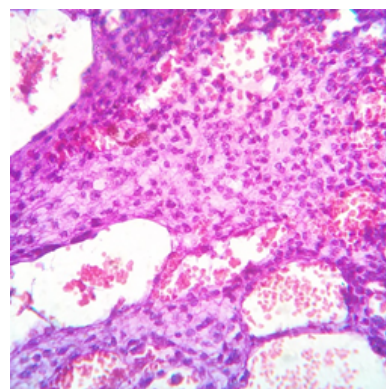
In our study, benign tumors were more common in head and neck region and malignant tumors more common in lower extremities, which were similar to studies conducted by Geeta Dev et al. [9,10] Average age of occurrence of benign tumor in the study of Mirza et al (2005) is 29.2 years, similar to the present study of 32.1 years whereas the average age of incidence of malignant tumors found highly increased in present study which is 47.7 years as compared to previous study of Mirza et al (2005) it was 37.8%. [5] (Table. 6)

Table. 6: Comparative analysis of age, sex and category of soft tissue tumors ; Benign lesions(B) and Malignant lesions(M)

Category	Mirza et al (2005)		Present study (2020)	
	B	M	B	M
Incidence (%)	82.5	17.5	85	12
Ratio (Benign: malignant)	4.7: 1		7.1 :1	
Average Age (Years)	29.2	37.8	32.1	47.7
M:F	1.13: 1	1.0: 1	1.3: 1	1.1: 1

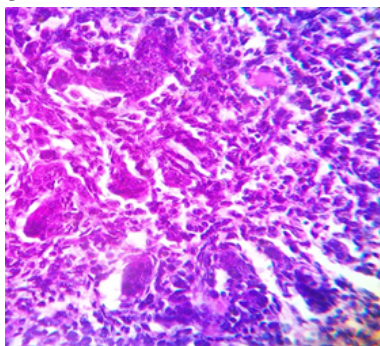
In present study, there were 192 benign soft tissue tumors, out of which 93 cases (41.2%) were reported as lipoma and its variants. Most common variant were classical lipoma and fibrolipoma similar to study conducted by Ndukwe et al. [11] Out of 27 malignant soft tissue tumors , 2 case were reported as liposarcoma which presented as thigh swelling. On microscopic examination, it is characterized by proliferating lipoblasts in different stages of differentiation, prominent anastomosing capillary network against mucoid background.(Fig. 2)

Benign blood vessel tumors constituted 32 cases, in which most common was hemangioma : 18 cases (7.9%) followed by 14 cases (6.2) of angiofibroma. Among malignant tumors, 2 caes of angiosarcoma were found and 1 case of hemangiopericytoma. 3 cases of lymphangioma were found. It composed of lymphatic channels lined by flattened endothelial lining and lymphocytic infiltration in stroma. In benign peripheral nerve sheath tumor, neurofibroma were 22 cases (9.7%), schwannoma 7 cases and one case of traumatic neuroma.



(Fig. 2: Atypical lipoblasts in liposarcoma, 40x)

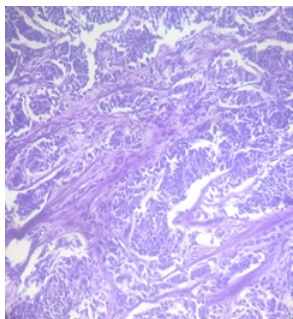
Neurofibromas were commonly seen in second decade with male preponderance and most common site being head and neck region was in accordance to study by Lin et al.^[12] The microscopic finding of neurofibroma revealed interlacing bundles of elongated cells with wavy nuclei. Neurofibromatosis (type 1 and 2) is associated with multiple benign nerve tumors. Near around 2% of the patients with neurofibromatosis type 1 develop in malignant peripheral nerve sheath.^[13] One case of malignant peripheral nerve sheath tumor was reported. In fibrous tumors, there were 3 cases of fibroma, 2 case of fibromyoma and 1 cases of fibromatosis. Out of 16 cases of fibrohistiocytic tumors, most common was benign fibrous histiocytoma : 10 cases (4.4%) followed by 3 cases of dermatofibroma and giant cell tumor of tendon sheath. Giant cell tumor of tendon sheath is common lesion seen on the distal extremities (hands more than toes) and is twice as common in women as men with a peak in the fourth through sixth decade of life.^[14,15] (Fig. 3) One case of leiomyoma and nodular fasciitis of hand was reported and 3 cases of glomus tumors. Soft tissue teratomas are more frequent in female and present either at birth or in early childhood.^[16,17] 5 Cases of soft tissue teratoma were found, all of which were present in sacrococcygeal area. The male : female ratio is 0.25 : 1. 2 cases of tumoral calcinosis were reported. Location being elbow and hips and both were in their third decade.



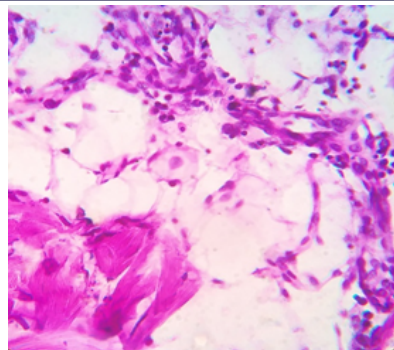
(Fig. 3: Linear arrays of mononuclear cells and multinucleated giant cells against collagen background in giant cell tumor of tendon sheath; 40x)

Out of 27 cases of malignant lesions, fibrosarcoma was the most common: 10 cases (4.2%) Microscopy showed infiltrative tumor composed cells arranged in fascicles and herring bone pattern 3 cases of undifferentiated pleomorphic sarcoma and 2 cases of rhabdomyosarcoma and extraskeletal Ewings sarcoma. Rhabdomyosarcoma have three major categories – pleomorphic, embryonal and alveolar. In present study, we found 2 cases of alveolar rhabdomyosarcoma in their 1st and 2nd decade of life. Microscopically, the section shows small round or oval tumor cells separated into nest by connected tissue septa. Some of the tumor cells detach from fibrous strands which results in a typical alveolar or pseudoglandular appearance. (Fig. 4)

1 case of synovial sarcoma, myxoid chondrosarcoma and alveolar soft tissue sarcoma were also found. Myxoid chondrosarcoma arises in the extremities of adult patients,^[18,19,20] but they have also been reported in the trunk and/or in children.^[21] Microscopically, it is characterized by multinodular growth pattern with strands and cords of relatively small cells with acidophilic cytoplasm embedded in abundant myxoid matrix. (Fig. 5) 7 cases of borderline dermatofibrosarcoma protuberans (DFSP). DFSP is typically centered in the dermis, but it can occur in deeper soft tissues.^[22]



(Fig. 4: Alignment of the tumor cells along fibrovascular septa in alveolar rhabdomyosarcoma; 40x)



(Fig. 5: Thin anastomosing strands of tumor cells surrounded by abundant myxoid matrix in myxoid chondrosarcoma; 40x)

CONCLUSION

Benign tumors were more common than malignant by a ratio of 7: 1. The most common benign tumors were lipoma followed by neurofibroma and hemangioma. The most common malignant tumor was fibrosarcoma. The most common site of benign lesion was head and neck region followed by trunk area and for malignant tumors it was lower extremities followed by head and neck region. Hematoxyline and eosin stained sections remain the best method for establishing the primary diagnosis of soft tissue tumors. Immunohistochemistry also plays an important role in diagnosis.

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