

SPECTRUM OF SOFT TISSUE TUMORS IN A TERTIARY CARE CENTRE - A 5 YEAR STUDY

Pathology

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Abstract:

Soft tissue includes the supportive connective tissue of various organs and the other nonepithelial, extra skeletal structures excluding the lymphoreticular system, viscera and coverings of brain. Soft tissue tumors form a heterogeneous group of neoplasms which often cause diagnostic dilemma.

Objectives: In this present study we tried to find out the relative frequency of soft tissue tumors and age, sex & site distribution of different types of soft tissue tumors (benign/malignant/intermediate).

Materials and Methods: It is a retrospective study conducted in the department of Pathology, Sree Narayana Institute of Medical Sciences, Ernakulum, Kerala. We tried to evaluate all the soft tissue tumors received during a period of 5 years in our department (2011 to 2015). All the specimens were received in the laboratory in 10% formalin fixative and processed through paraffin wax embedding method. New sections were cut at 5micrometer thickness and stained by Haematoxylin and Eosin (H&E) stain and all tumors were re-examined and classified.

Results and Conclusions: During our study period of five years we received a total of 291 soft tissue tumors which formed 5.46% of whole specimen load in our department. Benign tumors formed 93.8% of all soft tissue tumors followed by intermediate tumor (3.4%) while malignant tumors constituted 2.8%. Soft tissue tumors showed a male preponderance with the male to female ratio of 1.7:1 and extremities (39.1%) and head and neck (29.8%) areas were the common location of these tumors. Adipose tumors (61.5%) formed single most common histological group and the commonest benign tumor was lipoma. Malignant tumors were more common in elderly males and were located commonly in extremities (50%).

Keywords: soft tissue, benign, vascular

Introduction

The term soft tissue includes the supportive connective tissue of various organs and the other nonepithelial, extra skeletal structures excluding the lymphoreticular system, viscera and coverings of brain. It includes fibrous connective tissue, skeletal muscle, adipose tissue, peripheral nervous system and blood/lymph vessels. Embryologically, soft tissues originate

from the mesoderm layer.

Soft tissue tumors constitute a heterogeneous and large group of neoplasm. Conventionally Soft tissue tumors are classified according to the histogenetic features. Lipoma, for example, is defined as a tumor arising from adipocytes.

Even though soft tissue tumors can appear at any age, there is a relation between

patient's age, gender, location and the type of tumor.^{1,2} Lipoma which form one-third of the benign tumors is very uncommon in children, while angioliipomas are most common in young men and painful angioleiomyomas are common in middle-aged women. Rhabdomyosarcoma is seen generally in children and young adults while malignant fibrous histiocytoma and liposarcoma occur frequently in older adults.³ The overall incidence of soft tissue sarcomas increases with age and it is higher in males compared to females

Most soft tissue tumors are classified as either benign or malignant, but still there is an intermediate group which typically shows locally aggressive behavior and a low-to-moderate chance for metastasis. Benign mesenchymal tumors outweigh soft tissue sarcomas by a factor of 100. The annual incidence benign soft tissue tumors is estimated up to 3000/million while that of sarcoma is only around 30/million.⁴

Soft tissue tumors, both benign and malignant, commonly present as a slowly enlarging painless mass. A sudden and rapidly increasing soft tissue mass should arouse suspicion that the lesion is malignant. One-third of the benign soft tissue tumors formed by lipomas and another one-third is formed by fibrohistiocytic and fibrous tumors. Rest is composed of vascular tumors (10%) and nerve sheath tumors (5%). Majorities (99%) of the benign tumors is superficial and are less than 5 cm in diameter.

Even though soft tissue tumors can arise everywhere in the body, the most common locations are the extremities, head & neck region trunk and abdominal cavity.⁵ Extremities form common location of soft tissue sarcomas as three fourth of tumors arise there. 10% each occur in retro peritoneum and trunk wall and two-third tumors are deep seated. In general, the prognosis in older patients with a diagnosis of high-grade sarcoma is poor.

A biopsy is indicated when any soft tissue tumor arises in a patient with no previous history of trauma.⁶ Another indication for open biopsy is a mass which persists even after 6 weeks of local trauma. Role of FNAC is significant in documenting local recurrences or metastasis in a previously diagnosed case of soft tissue sarcoma.⁷ Diagnostic accuracy of conventional light microscopy can be increased by using techniques like special stains, electron microscopy, immunohistochemical studies and molecular/ cytogenetic methods.

Objectives

1. To study the relative frequency of different types of soft tissue tumors (benign/ malignant/intermediate).
2. To study the histopathological features of soft tissue tumors and to know age, sex & site distribution of these tumors.

Materials and Methods

This study is conducted in the department of Pathology, Sree Narayana institute of medical Sciences, Ernakulum, Kerala. It is a retrospective study. We tried to evaluate all the soft tissue tumors (benign, intermediate and malignant) received during a period of 5 years in our department (2011 to 2015). All the specimens were received in the laboratory in 10% formalin fixative and processed through paraffin wax embedding method. We collected all the relevant history, clinical features, radiological and per operative findings from record section. New sections were cut at 5micrometer thickness and stained by Haematoxylin and Eosin (H&E) stain. Wherever necessary we used special stains like Geimsa, Periodic Acid Schiff, Reticulin, PTAH and Masson's Trichrome. All soft tissue tumors were further examined and categorized.

Figure 1:
Age and sex incidence in all soft tissue tumors

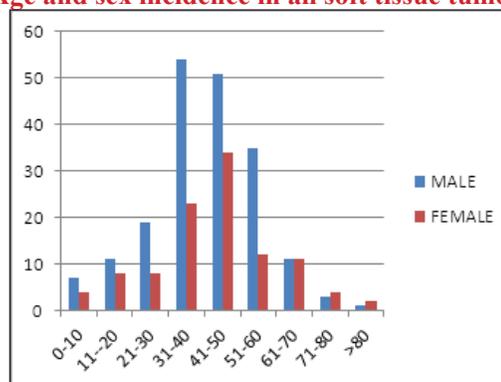


Figure 2: Sex incidence of all soft tissue tumors

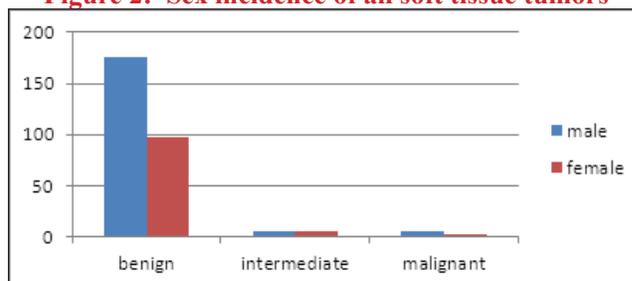


Table 1: Sex distribution of soft tissue tumors in various studies

	M:F ratio
Jain p ¹¹	1.2:1
Kransdorf ^{1,2}	1.2:1
Batra P ¹⁵	2.1:1
Mandong BM ¹⁰	2:1
Myhre –Jensen O ⁹	1:1
Beg S ⁷	1.8:1
Present study	1.7:1

Table 2: Site distribution of all soft tissue tumors

Site	Benign	Intermediate	Malignant	Total
Extremity	104	6	4	114
Head and neck	86		1	87
Shoulder and back	40		1	41
Trunk and abdomen	36	2	2	40
Others	7	2		9
Total	273	10	8	

Table 3: Distribution of cases

	Benign	Intermediate	Malignant
Adipocytic tumors	178	1	
Fibroblastic/myofibroblastic tumors	5	6	1
Fibrohistiocytic tumors	18	1	1
Vascular tumors	47	1	1
Pericytic/perivascular tumors	2		
Nerve sheath tumors	19		
Smooth muscle tumors	1		2
Tumors of uncertain differentiation	2	1	3
Chondro-osseous tumors	1		
Total Cases	273	10	8

Table 4: Distribution of soft tissue tumors in various studies

	Benign	Malignant
Jain p ¹¹	90.6%	9.4%
Kransdorf ^{1,2}	60.2%	39.8%
Batra P ¹⁵	89.2%	10.8%
Makino ¹⁶	96%	45%
Petersen ¹⁴	35%	49%
Present study	93.8%	2.8%

Table 5: Distribution of benign tumors

	No of cases
Adipocytic tumors	
Lipoma	157
Angiolipoma	7
Fibrolipoma	9
Myo lipoma	2
Spindle cell lipoma	3
Fibroblastic/myofibroblastic tumors	
Nodular fasciitis	2
Tendon sheath fibroma	1
Myositis ossificans	1
Nuchal type fibroma	1
Fibrohistiocytic tumors	
Benign fibrous histiocytoma	7
Giant cell tumor of tendon sheath	11
Vascular tumors	
Capillary hemangioma	40
Cavernous hemangioma	1
Arterio-venous hemangioma	3
Lymphangioma	3
Pericytic/perivascular tumors	
Glomus tumor	2
Nerve sheath tumors	
Schwannoma	12
Neurofibroma	7
Smooth muscle tumors	
Leiomyoma	1
Tumors of uncertain differentiation	
Myxoma	1
Deep angiomyxoma	1
Chondro-osseous tumors	
Soft tissue chondroma	1

Table 6: Distribution of malignant tumors

	No of cases
Fibroblastic/myofibroblastic tumors	
Fibrosarcoma	1
Fibrohistiocytic tumors	
Pleomorphic MFH	1
Vascular tumors	
Angiosarcoma	1
Smooth muscle tumors	
Leiomyosarcoma	2
Tumors of uncertain differentiation	
Synovial sarcoma	1
PNET	1
Clear cell sarcoma of soft tissue	1
	8

Table 7: Distribution of intermediate tumors

	No of cases
Adipocytic tumors	
Atypical lipomatous tumor	1
Fibroblastic/myofibroblastic tumors	
Superficial fibromatosis	4
Desmoid type fibromatosis	1
Inflammatory myofibroblastic tumor	1
Fibrohistiocytic tumors	
Giant cell tumor of soft tissue	1
Vascular tumors	
Kaposiform hemangioendothelioma	1
Tumors of uncertain differentiation	
Myoepithelioma	1
	10

Results

During our study period of five years we received a total of 291 specimens of soft tissue tumors specimens in the department. The results are concludes as follows. The soft tissue tumors formed 5.46% of whole specimen load in our department of pathology during the last 5 years. There were a total of 291 cases of soft tissue tumors. Soft tissue tumors were more commonly seen in 31-40yr and 41-50 yr old patients. Soft tissue tumors showed a male prepondence with the male to female ratio of 1.7:1. Extremities (39.1%) and head and neck (29.8%) areas were the common location of soft tissue tumors.

Benign tumors formed 93.8% of all soft tissue tumors followed by intermediate tumor (3.4%) while malignant tumors constituted 2.8%

While considering the whole soft tissue tumors, adipose tumors (61.5%) formed single most common histological group followed by vascular tumors (16.8%) and fibrohistiocytic tumors (6.8%)

65.2% of benign soft tissue tumors were adipocytic tumors, while vascular tumors (17.2%) formed the second largest category and then by nerve sheath tumors (6.9%)

The commonest benign tumor was lipoma

Most of fibroblastic tumors (46%) belonged to intermediate category and seen in extremities

Malignant tumors were more in elderly males and also located commonly in extremities (50%)

Discussion

Soft tissue tumors always have fascinated clinicians and pathologists for many years. This heterogeneous group include a large variety of tumors that show close histopathological similarities with only very few difference which is revealed only on accurate and careful microscopic examination.

By definition Soft tissue represents the nonepithelial-extra skeletal tissue of the body exclusive of glia, reticuloendothelial system and supporting connective tissue of parenchymal organs. Adipose tissue, voluntary muscles, vessels and fibrous tissue forms the soft tissue structures of the body.

In our study duration of five years soft tissue tumors formed 5.46% of the total specimen load. There were a total of 291 cases of soft tissue tumors. Even though such tumors can arise in any age group, most of the cases in our study belonged to 31-40 age group followed by 41-50yr group [fig 1]. Malignant soft tissue tumors/sarcomas were common in elderly patients belonging to 61-70 yr group. Soft tissue tumor were common in males with a male to female ratio of 1.7:1 which is equal to the results obtained by Lazim et al.⁸ He studied a total of 213 cases of soft tissue tumors in one year and reported a M:F ratio of 1.7:1. Our study is also comparable with studies of Beg et al while studies done by Mynes Jensen et al⁹ showed an equal sex distribution. In our study both benign and malignant tumors showed male preponderance, in contrast intermediate tumors showed an equal distribution among males and females [fig 2][Table 1].

When we categorized soft tissue tumors according to their original location, extremities are found to be the frequent location. 39.1% of all soft tissue tumors were located in the extremities. 38.1% of benign, 60% of intermediate and 50% of malignant tumors were also seen in the same site which is comparable with Beg et al and Madong et al¹⁰ studies. Next preferred locations were head and neck area (29.8%) and shoulder & back (14.1%) [Table 2].

Previous studies showed benign soft tissue tumors out number malignant tumors by a margin of 100. In our study 93.8% of soft tissue tumors were benign. There was only 8 cases (2.8%) of malignant tumors. [Table 3]. These results were also similar to previous studies done by Jain P et al.¹¹ In all previous studies done by Myher Jensen et al, Lazxim

et al and M.J. Kransdorf et al benign tumors predominated over malignant tumors [Table 4]. But in fact the relative frequency of benign to malignant tumors appear difficult to estimate clearly because most of the benign tumors do not cause much problems and so most of the benign lesions are not surgically removed.

In benign soft tissue tumors adipocytic tumors formed the largest group. 57.5% of benign tumors belonged to this group and lipoma formed the 88.2% of all adipocytic tumors. There were also a few cases of angiolipoma, fibrolipoma, myolipoma and spindle cell lipoma. Vascular tumors (17.2%) formed next largest group of soft tissue tumors followed by nerve sheath (6.9%) and fibrohistiocytic tumors (6.6%). [table 5] Most of the benign tumors were located in the extremities (38.1%) [Table 2] and were common in males (64.1%) [fig. 2].

Malignant soft tissue tumors were rare and formed just 2.8% of all soft tissue tumors [Table 6]. Majority (63.5%) of malignant tumors were seen in elderly males. 50% of these tumors were located in extremities. Trunk and abdomen (25%) seen to be the next preferred location. This predilection is also confirmed by the studies of Gebhard et al¹², Kransdorf² and Meis-Kindblom¹³. Accurate histological classification and grading are the two most important factors predicting the behavior and prognosis of soft tissue sarcoma. Grading of a soft tissue sarcoma is based mainly on the degree of differentiation, cellular pleomorphism, number of mitotic figures per high power field and presence/absence of necrosis. In addition to special stains we also needed help of immunohistochemistry for the diagnosis and confirmation of many cases of soft tissue sarcomas as in case of a small round cell tumor which on IHC appear to be Ewing's sarcoma.

According to the WHO classification, an intermediate group exists in soft tissue tumors which are known to cause repeated local recurrences and also show low to moderate risk for metastasis. There were 10 intermediate tumors in our study which formed 3.4% of all soft tissue tumors. Largest group in this category is formed by fibroblastic and myofibroblastic tumors like superficial and desmoids like fibromatosis, inflammatory myofibroblastic tumors [Table 7]. In a retrospective study done by Petersen et al¹⁴ who classified the tumors into intermediate group, 11.4% of soft tissue tumors belong to this group. An interesting tumor in this group was atypical lipomatous tumor / well differentiated liposarcoma. A 29 year old female patient

presented with slowly enlarging painless swelling in right thigh which. Microscopic examination of the sections showed a partially encapsulated cellular adipocytic tumor composed of adipocytes of differing sizes, and also stromal cells with hyperchromatic enlarged nucleus. Also seen scattered multivacuolated lipoblasts in between the adipocytes.

Soft tissue tumors form a vast and heterogeneous group of neoplasms. The diagnosis of soft tissue tumors requires a team approach. Careful examination of gross specimen and adequate sampling is very essential. Light microscopic examination of H&E stained sections are usually sufficient for the diagnosis. In doubtful cases other techniques like special stains, IHC and electron microscopy can also be used.

Conclusions

Soft tissue tumors form a significant specimen load in all histopathological laboratories. In our study 291 cases of soft tissue tumors formed 5.46% of whole specimen load during the last 5 years. Benign tumors formed major bulk of these heterogeneous neoplasms. Soft tissue tumors showed a male preponderance with common locations being Extremities and head and neck while considering the whole soft tissue tumors, adipose tumors formed single most common histological group followed by vascular tumors and fibrohistiocytic tumors. The commonest benign tumor was lipoma. Most of fibroblastic tumors belong to intermediate category and seen in extremities. Malignant tumors were more in elderly males and also located commonly in extremities. Even though the role of light microscopic examination is indispensable in the diagnosis of soft tissue tumors diagnostic accuracy can be increased by introduction of ancillary techniques.

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