

Pattern of Soft Tissue Tumours- A Histopathological Study

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Abstract

Soft tissue is defined as the complex of non epithelial extraskeletal structures of the body exclusive of the supportive tissue of the various organs and the hematopoietic/lymphoid tissue. It is composed of fibrous tissue, adipose tissue, skeletal muscle, blood and lymph vessels and peripheral nervous system. Soft tissue tumours constitute a large and heterogenous group of neoplasms. WHO has classified soft tissue tumors in different categories like Adipocytic tumors, Fibroblastic, Fibro-histiocytic, Smooth muscle, Pericytic, Skeletal muscle, Vascular, Chondro-osseous & tumors of uncertain differentiation. These are further subdivided into Benign, Intermediate & Malignant. Pattern of a particular disease in any population is studied with the idea of obtaining information about the clinical presentation, the varied morphology and the etiological factors that can be assessed in relation to its prognosis. The present study was undertaken with a view to getting information about the rate of prevalence, relative frequency of various histopathological types of neoplasms and morphological patterns of soft tissue tumors in our institution.

Key Words

Soft Tissue Tumors, Histopathological, Benign Tumors, Malignant Tumors

Introduction

Soft tissue is defined as the complex of non epithelial extraskeletal structures of the body exclusive of the supportive tissue of the various organs and the hematopoietic/lymphoid tissue. It is composed of fibrous tissue, adipose tissue, skeletal muscle, blood and lymph vessels and peripheral nervous system.

Soft tissue tumours constitute a large and heterogenous group of neoplasms. These tumours may arise in any location, 40% occur in the lower extremities especially the thigh, 20% in the upper extremities, 10% in the head and neck and 30% in the trunk and retroperitoneum. WHO has classified soft tissue tumors in different categories like Adipocytic tumors, Fibroblastic, Fibro-histiocytic, Smooth muscle, Pericytic, Skeletal muscle, Vascular, Chondro-osseous & tumors of uncertain differentiation. These are further subdivided into Benign, Intermediate & Malignant. Majority of soft tissue tumours are benign. Benign mesenchymal tumours outnumber sarcomas by a factor of 100. Males constitute 61.5% cases as compared to females i.e 38.4% cases. Benign tumours are roughly equally distributed across all parts of the body with a slight predilection for the upper parts i.e. head

and neck and trunk region. Of the benign soft tissue tumours, 99% are superficial in location and 95% of tumours average less than 5cm in size. One-third are lipomas, one-third are fibrohistiocytic and fibrous, 10% vascular tumours and 5% nerve sheath tumours. Rosai and Ackerman.(1)

Soft tissue tumors may be further defined by the location of the lesion and the age of the patient like in the retroperitoneum, approximately half the benign lesions in the 16 to 25 age group are fibromatosis (20%), schwannoma (14%) and neurofibroma (13%). For the same location in children 5 years or younger, almost two thirds of the benign tumors are lipoblastoma (37%) or lymphangioma. Soft tissue tumors may be further defined by the location of the lesion and the age of the patient like in the retroperitoneum, approximately half the benign lesions in the 16 to 25 age group are fibromatosis (20%), schwannoma (14%) and neurofibroma (13%). For the same location in children 5 years or younger, almost two thirds of the benign tumors are lipoblastoma (37%) or lymphangioma. Soft tissue tumors may be further defined by the location of the lesion and the age of the patient

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like in the retroperitoneum, approximately half the benign lesions in the 16 to 25 age group are fibromatosis (20%), schwannoma (14%) and neurofibroma (13%). For the same location in children 5 years or younger, almost two thirds of the benign tumors are lipoblastoma (37%) or lymphangioma. Evans Histological Appearances of Tumours. (2)

Malignant soft tissue tumours are seen more commonly in males than in females in the ratio 1.4:1 and incidence increases with the increasing age. The commonest site of malignant soft tissue tumours is lower extremities (3). More than 50 histological types of malignant soft tissue tumors have been identified, the most common being malignant fibrous histiocytoma (28%) followed in the descending order of frequency by fibrosarcoma(25%), rhabdomyosarcoma (24%), leiomyosarcoma(12%), liposarcoma(15%) and synovial sarcoma(10%) .(4)

Soft tissue sarcomas also arise as a result of exposure to intense or high dose of certain chemicals such as vinyl chloride, arsenic, herbicides etc in the working environment. HIV infection in human leads to Kaposi sarcoma and the major risk factor or cause of this cancer is Kaposi Sarcoma-associated herpesvirus (KSHV) or human herpesvirus 8. (5)

Grading is the best predictor of metastasis outcome in adult soft tissue sarcomas and should be part of the pathologic report. Histological grade and tumor size are equally tant metastasis and survival. (6) The most commonly used grading systems for soft tissue tumors are the French grading and the National Cancer Institute grading. Both are 3-grade systems and are mainly based on histologic type and the subtype, tumour necrosis and mitotic activity. (7)

Material and Methods

The study done in Department of Pathology, Acharya Shri Chander College of Medical Sciences and Hospital, Jammu, deals primarily with soft tissue tumors located in the somatic soft tissues, it excludes those arising from the soft tissues of the mediastinum & visceral organs.

The present study was conducted in two parts: *Part 1-* For the retrospective study, detailed information was collected from the histopathology records of the Post Graduate Department of Pathology, Acharya Shri Chander College of Medical Sciences and Hospital, Sidhra, Jammu(J&K), of all the histologically diagnosed cases of soft tissue tumours for the last 5 years i.e from 1st November 2005 to 31st October 2010.

A) The relevant data was collected from both the clinical case sheets and histopathological records.

B) The H&E stained slides were retrieved and already

diagnosed cases were re-examined. Special stains were applied wherever required.

Part 2- For the prospective study, the study materials comprised of specimens of soft tissue tumours received prospectively in the Post Graduate Department of Pathology, Acharya Shri Chander College of Medical Sciences and Hospital, Sidhra, Jammu(J&K) from 1st Nov 2010 to 31st October 2011.

The clinical information of the patients of soft tissue tumours received by the department was obtained from histopathological requisition forms and deficient relevant information was procured from the clinical case sheets and the concerned clinician. The sections from the representative area of tumour were stained with H&E. Special stains such as Massons Trichrome, Reticulin, PTAH & PAS were carried out where ever necessary. (8)

Results

A total of 170 cases were studied. Out of a total of 170 cases detected during the said period, majority were benign tumours i.e 150 cases (88.2 %) , 17 were malignant tumours (10%) , whereas intermediate group constituted 3 cases (1.8 %) only. In the present study the overall prevalence of soft tissue tumours (benign as well as malignant) was observed to be higher in males (58.8%) as compared to females (41.2%) with a male to female ratio of 1.4 : 1 as shown in *table .1*

The ages of the patients as shown in *table . 2* ranged from 3 - 73 years. Maximum number of cases i.e 36 (21.2%) were seen in the age group 41-50 years , followed by 31-40 years age group i.e 33 cases (19.5%). Third in frequency was age group 21-30 years constituting 25 cases (14.7%). Least number of cases i.e 6 cases (3.5%) were observed in above 70 years of age. Maximum number of cases i.e.32 cases of benign soft tissue tumors were in 31-40 age group, followed by 41-50 age group i.e.29 cases. On the other hand, most of the malignant soft tissue tumors occurred after the age of 40 years, comprising 12 cases of the total 170 cases .

Amongst the benign tumours , Haemangioma(10 cases), lymphangioma (3 cases) were mostly confined to less than 10 years of age. Nerve sheath tumors like Schwannoma, Neurofibroma were seen most commonly in 31-40 age group, while Lipoma was distributed in all the age groups. Least number of cases i.e 6 cases were seen in greater than 70 years of age which included lipoma (1 case), Schwannoma (1 case), Fibromatosis (1 case), Neurofibroma(2cases). The malignant soft tissue tumors mostly occurred 40 years onwards. Only one case i.e rhabdomyosarcoma was

Table 1. Sex Wise Distribution Of Soft Tissue Tumors

	Lesion	Males		Females	
		No.	%	No.	%
Benign	Lipoma	32	18.8%	19	11.1%
	Schwanoma	20	11.7%	12	7.1 %
	Haemangioma	17	10 %	13	7.6 %
	Lymphangioma	2	1.2%	5	3 %
	Neurofibroma	8	4.7%	4	2.4%
	Granuloma pyo genicum	3	1.7%	2	1.2%
	Glomus tumor	1	0.6%	3	1.7%
	Fibroma	1	0.6%	1	0.6%
	Fibromatosis	2	1.2%	1	0.6%
	Traumatic neuroma	1	0.6%	-	-
	Leiomyoma	-	-	3	1.7%
Intermediate	Haemangioperi cytoma	2	1.2%	-	-
	DFP	1	0.6%	-	-
	Fibrosarcoma	-	-	2	1.2 %
Malignant	Rhabdomyo sarcoma	2	1.2%	-	-
	Angiosarcoma	2	1.2%	1	0.6%
	Liposarcoma	1	0.6%	1	0.6%
	Leiomyo sarcoma	1	0.6%	1	0.6%
	MFH	3	1.7%	2	1.2%
	MPNST	1	0.6%	-	-
	Total	100	58.8%	70	41.2%

Table 2. Age Wise Distribution of Soft Tissue Tumours

Age (Yrs)	Benign	Intermediate	Malignant	Total
Below 10	13	-	1	14 (8.2%)
11-20	22	-	1	23(13.6%)
21-30	24	-	1	25(14.7%)
31-40	32	-	1	33(19.5%)
41-50	29	3	4	36(21.1%)
51-60	19	-	4	23(13.5%)
61-70	6	-	4	10 (5.8%)
Above 70	5	-	1	6 (3.5 %)
Total	150	3	17	170

seen in a 6 year child. *Table .3* shows Upper extremities to be the most commonly encountered site for benign soft tissue tumours i.e 50 cases (29.4%) followed by head and neck i.e 37 cases (21.7%). Likewise the most common site observed for malignant soft tissue tumours was lower extremities i.e 7 cases (4.1%) followed by retroperitoneum accounting for 6 cases (3.5%). Least involved sites for malignant soft tissue tumours were upper extremities (0.6%), head and neck (0.6%), back (0.6%) and abdomen (0.6%). One case (0.6%) of intermediate soft tissue tumour was present each in upper extremity, lower extremity and abdomen. Lipoma, the most common of benign soft tissue tumor was mainly distributed in upper

extremities (19 cases). Similarly schwanoma (15cases), neurofibroma (3 cases), glomus tumor (3cases) were also observed mainly in upper extremities. Haemangioma (12 cases), lymphangioma (4cases) showed predominance in head & neck region. Two cases of schwanoma were located in spinal cord & 4 cases in brain. Retroperitoneum was an uncommon site for benign soft tissue tumors accounting for 3 cases only (one case of lipoma, one case of fibroma & one case of leiomyoma).

For malignant soft tissue tumors, lower extremity was the most common site accounting for 7 of the 17 cases. Amongst these 2 were of liposarcoma, 2 cases were of fibrosarcoma, one case was of MFH and one case was

Table 3 Site Wise Distribution of Soft Tissue Tumors

Site	Benign		Malignant		Intermediate	
	No	%	No	%	No	%
Upper extremities	50	29.4	1	0.6	1	0.6
Chest	13	7.6	-			
Back	16	9.4	1	0.6		
Lower extremities	16	9.4	7	4.1	1	0.6
Head&neck	37	21.7	1	0.6		
Retroperitoneum	3	1.7	6	3.5	1	0.6
Abdomen	5	3	1	0.6		
Spinal cord	5	3	-			
Brain	5	3	-			
TOTAL	150	88.2	17	10	3	1.8

of angiosarcoma. Second in frequency was the retroperitoneum accounting for 5 cases (two cases were of MFH, one case of angiosarcoma, one case of leiomyosarcoma & one case of rhabdomyosarcoma). One case of leiomyosarcoma was located in abdomen while one case of rhabdomyosarcoma was located in head & neck region. The overall prevalence rate of soft tissue tumors as per data obtained from the records available for the period of 6 years was 2.4/1000 surgical admissions for histologically proven cases. The prevalence rate of benign soft tissue tumours was 2.2/1000 surgical admissions while the prevalence rate of malignant soft tissue tumors was 0.25/1000 surgical admissions. Majority of benign soft tissue tumors i.e. 107 cases were between 2-5 cms in size, 35 cases varied between 6-10 cms, only 8 cases were >10 cms in size while Majority of malignant soft tissue tumours (14 cases) were > 10 cms in size and only 3 cases were between 6-10 cms in size. Amongst the 150 cases, most of Lipoma (47 cases) & Haemangioma (28 cases) were soft in consistency, 52 cases were firm in consistency which included schwannoma (24 cases), leiomyoma (3 cases) & glomus tumor (4 cases), while only one case of schwannoma was hard in consistency. whereas 11 malignant cases were hard in consistency, 5 malignant cases were firm in consistency & only one case of liposarcoma was soft in consistency. Out of 150 benign soft tissue tumours cases, capsule was appreciated in all cases of lipoma (51 cases) & schwannoma (32 cases) & in most cases of haemangioma (27 cases), glomus tumor (3 cases), leiomyoma (2 cases) whereas capsule was absent in all the malignant tumor cases. On microscopic examination, majority of the benign soft tissue tumours (146 cases) were well circumscribed, only 4 cases showed infiltrative growth pattern which included 2 cases of fibromatosis, 1 case of haemangioma & 1 case of granuloma pyogenicum. Out of 150 cases 143 were well differentiated and 7 cases showed moderate degree of

differentiation while all the malignant soft tissue tumours i.e 17 cases and 2 out of three intermediate cases showed infiltrative growth pattern. 14 cases were poorly differentiated and 3 cases showed moderate degree of differentiation. Most of the benign cases i.e 140 cases exhibited mild nuclear pleomorphism and 10 cases showed moderate nuclear pleomorphism. Conspicuous nucleoli were seen in 7 cases. Mitotic count of 0-9 per hpf were seen in 143 cases, 10-19 mitosis per high power field were observed in 7 cases only. In contrast to this 12 cases malignant tumors showed marked nuclear pleomorphism and 5 cases exhibited moderate nuclear pleomorphism. In intermediate group 2 cases exhibited moderate nuclear pleomorphism and 1 case showed mild nuclear pleomorphism. Mitotic count of > 20 per hpf was observed in 11 cases, 10-19 mitosis per hpf were seen in 6 of 17 malignant cases and 2 intermediate cases. Tumour necrosis was observed in 15 malignant cases and haemorrhage in 12 malignant cases while only 5 benign cases (2 cases of lipoma showing fat necrosis) showed necrosis and haemorrhage was seen in 13 cases. Tumour giant cells were seen in only 1 case of lipoma, while inflammatory cells were present in 95 cases. Tumour giant cells were seen in all the 17 malignant cases, vascular invasion in 8 malignant cases while 5 malignant cases showed presence of inflammatory cells most notably in MFH (2 cases), Angiosarcoma (1 case), Fibrosarcoma (1 case) & leiomyosarcoma (1 case).

Discussion

In the present study, out of a total of 170 cases, 150 cases (88.2%) were benign, 3 cases (1.8%) were of intermediate grade & 17 cases (10%) were malignant. This is comparable with a study carried out by Makino (9) on 651 patients, of which 628 (96%) were observed to be benign & only 23 (4%) to be malignant. Similarly in another study, conducted by Strauss (10), benign soft tissue tumors outnumbered malignant by a ratio of 100:1. In the present study males with soft tissue tumors slightly

outnumbered females, with a male to female ratio of 1.4:1. The sex distribution in the present series is comparable with findings of the study conducted by Sir Stanford Cade (11) who also observed a preponderance of soft tissue tumors in men. Of the total 153 patients, 86 were males & 67 were females with a male to female ratio of 1.2:1. In the present study benign soft tissue tumors were observed most frequently in 31-40 age group & malignant soft tissue tumors occurred after the age of 40 years with one case of rhabdomyosarcoma in a 6 year old child.

In children, the common tumors were haemangioma & lymphangioma. This is similar to study conducted by Coffin & Dehner (12) on soft tissue tumors of childhood & adolescence, who reported that more than 75% cases in this age group were haemangioma and lymphangioma. In the present study, all the cases of leiomyosarcoma were seen in the age group 51-60 years. This is concordant with the findings of study by Gustafson *et al* (13) in which the median age of 48 patients with leiomyosarcoma was observed to be 65 years.

In the present study benign soft tissue tumors were most frequently observed in upper extremities followed by head & neck. The common site for malignant soft tissue tumors was lower extremity, followed by retroperitoneum. Strauss (10) in his study concluded that malignant soft tissue sarcomas most commonly affect the lower limbs i.e approximately 45% of sarcomas occur in the lower extremities, 15% in the peritoneum and the abdominal wall, 15% in the abdominal and chest wall, 15% in the upper extremities and 10% in the head-and-neck region. In the present study 51 cases of lipoma were seen, most of them varying between 2-10 cms in size. Microscopically, most of the tumors were composed of mature adipose tissue. This was in agreement with study conducted by Furlong (14) who reviewed 125 cases of lipoma and concluded that apart from mature adipose tissue, lipoma variants like Fibrolipoma, Spindle or Pleomorphic variants could also be seen. In the present study 32 cases of schwannoma were seen. All were well circumscribed & encapsulated with distinctive microscopic growth patterns Antoni A & Antoni B areas. This is similar to study conducted by Bhatoe *et al* (15) who also confirmed schwannoma histologically by mixture of Antoni A & Antoni B areas. In the present study, 5 cases of MFH were seen, most of them were greater than 10 cms in size, unencapsulated & hard in consistency. Microscopically predominantly storiform pattern & tumor giant cell were seen. Enzinger (16) also observed microscopic pattern varying from storiform pattern to areas having pleomorphic appearance in 200 cases of

MFH studied by them. 3 cases of Fibromatosis were seen, which showed infiltrative growth pattern & moderate pleomorphism. Raymond *et al* (17) in their study on plantar fibromatosis, histologically observed proliferating fibroblasts with invasive properties & high cellularity.

Conclusion

The present study provides a fair insight into the varied histological patterns of soft tissue tumors in this region of the country.

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