Original Article

Clinicopathological Profile of Myxoid Soft Tissue Tumors- A Retrospective Study in a Tertiary Care Hospital in South India

Karthik Sigamani, Karkuzhali Ponnuswamy[1]

Department of Pathology, Karpaga Vinayaga Institute of Medical Sciences and Research Centre, Chinnakolambakkam, Chengalpattu District, Tamil Nadu, [1]Pathology, Madras Medical College and Rajiv Gandhi Government General Hospital, Chennai, Tamil Nadu, India

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ABSTRACT

Background: Myxoid soft tissue tumors are rare and diagnostically challenging group of tumors with varied biological behavior ranging from benign, locally aggressive to distantly metastasizing malignant tumors. **Aims:** The objectives of the study are to identify the relative frequency and distribution of myxoid soft tissue tumors among patients in a tertiary care hospital and to study the clinicopathological features of these tumors. This was a retrospective cross-sectional study conducted in the department of pathology of a tertiary care hospital from January 2008 to December 2013. **Materials and Methods:** Clinical and pathological details of all the 80 myxoid soft tissue tumors reported during the study period were retrieved from the records of department of pathology. Corresponding Hematoxylin & Eosin (H & E) slides were reviewed, and Immunohistochemistry (IHC) was carried out for confirmation. The relationship among various prognostic variables was analyzed in case of myxoid sarcomas. **Results:** Myxoid soft tissue tumors accounted for 3.7% among the soft tissue tumors with a predominance of malignant myxoid sarcomas (71.25%) in contrast to the overall picture of sarcomas. Myxoid neurofibroma (34.78%) was the most common benign tumor, while myxofibrosarcoma (33.33%) was the frequent myxoid sarcoma. A statistically significant correlation was seen between tumor size and depth (*P*-value: 0.038) and also between presence of vascular invasion and histological grade (*P*-value: 0.012) of sarcomas. **Conclusion:** Light microscopic morphology, supplemented by ancillary techniques like IHC, remains the cornerstone for diagnosis of myxoid soft tissue tumors.

Keywords: *Clinical features, glycosaminoglycans, histomorphology, sarcomas*

Introduction

The term "Soft tissue" includes the nonepithelial extra-skeletal tissue of the body exclusive of the viscera, reticuloendothelial (lymphoreticular) system and coverings of the brain. Soft tissue tumors are a rare and heterogeneous group of tumors that arise from or show differentiation toward the mesodermally-derived connective tissue elements like fat, fibrous tissue, vessels, peripheral nerves, tendons, and fasciae. Myxoid soft tissue tumors constitute a group of lesions, both benign and malignant, with abundant extracellular myxoid matrix.[1] The myxoid matrix is composed of Glycosaminoglycans-sulphate (Chondroitin sulphate and Keratan sulphate) and non-sulphate (Hyaluronic acid).[2] Glycosaminoglycans (GAGs) are negatively charged and are highly hydrophilic forming a 'gel' state readily and result in the characteristic myxoid morphology when present in increased amount. The relative proportion of myxoid areas varies with some tumors being predominantly myxoid while others showing focal myxoid change. It appears on Hematoxylin & Eosin (H&E) sections as an amorphous basophilic substance. GAGs have high affinity for growth factors and cell adhesion molecules thereby

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Address for correspondence: Dr. Karthik Sigamani, F-3, Lakshmi Vilas, Plot No-87, Lalitha Nagar, Madambakkam, Guduvancherry – 603 202, Tamil Nadu, India.
E-mail: eversmile_kar@yahoo.com

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facilitating cell proliferation and cell to cell interaction.[3] The biophysical properties of GAGs favor tumor cell migration and the diffusion of metabolites, facilitating tumor growth.[4] Chondroitin sulphate modulates the fate of tumor cells by preventing apoptosis and is also involved in cell proliferation.[5]

Compared with other malignant tumors, soft tissue sarcomas are relatively rare constituting less than 1% of all cancers[6] and accounting for 1.15% of all cancer-related deaths.[7] Soft tissue sarcomas virtually can occur in any sites in the body with the most common sites being the extremities (59%), trunk (19%), retroperitoneum (15%), and head and neck (9%).[8] The prognosis of malignant myxoid soft tissue tumors depends on several pathological factors including tumor location, depth of the tumor, tumor size, histological type, histological grade, pathological stage, neurovascular and bony invasion.[9,10] The objectives of the study were to identify the relative frequency and distribution of myxoid soft tissue tumors among patients in a tertiary care hospital and to study the clinicopathological features of these tumors.

Materials and Methods

Study setting and design

This was a six-year retrospective cross-sectional study conducted in the department of pathology of a tertiary care hospital after approval by the institutional ethics committee. All the myxoid soft tissue tumors reported during the period of January 2008 to December 2013 in the department of pathology were included in the study. Analysis of the study was carried out in 2014.

Sample size

A total 80 myxoid soft tissue tumors were included in the study using purposive sampling technique.

Inclusion criteria

1. Patients of all age groups and gender.
2. Excision/Wide local excision specimens of myxoid soft tissue tumors.
3. Benign and malignant myxoid soft tissue tumors diagnosed by histopathological examination.

Exclusion criteria

1. Tumor-like conditions with myxoid change.
2. Chondromyxoid bone tumors with soft tissue extension.
3. Other resected non-myxoid soft tissue tumors.

Data collection

All the relevant clinical and pathological data including age, sex, site, tumor location, and tumor size were obtained for all the 80 myxoid soft tissue tumors from the surgical pathology records in the department. Corresponding Hematoxylin & Eosin (H&E)-stained slides were reviewed, and all the malignant myxoid soft tissue tumors were graded using the FNCLCC (Federation Nationale des Centres de Lutte Contre le Cancer) grading system. Other prognostic parameters like presence of vascular invasion, bone invasion, and presence of local recurrence were documented and analyzed. The relationship between various prognostic variables was also assessed. Immunohistochemistry (IHC) was carried out for confirmation based on the light microscopic features using a panel of markers that included vimentin, pancytokeratin (Pan CK), desmin, Smooth Muscle Actin (SMA), CD34, S100, and CD99.

Statistical analysis

- The statistical analysis was performed using statistical package for social science software version 11.5.
- Descriptive statistical measures like frequency and percentage were calculated.
- Pearson Chi-square test was used to investigate the relationship among various prognostic variables in case of myxoid sarcomas. A *P* value of 0.05 was taken as cut-off point to determine statistically significant results.

Results

In the study period of six years from January 2008 to December 2013, a total of 47,482 specimens were received in the department of pathology for histopathological examination. Out of them, the total number of soft tissue tumors was 2163 accounting for 4.56%. Among the soft tissue tumors, the total number of benign and malignant (including intermediate malignancy) tumors were 1,982 and 181, respectively. Thus, the distribution of benign tumors was 91.6% and that of malignant tumors was 8.37% among the soft tissue tumors. During this study period, the total number of myxoid soft tissue tumors was 80 with

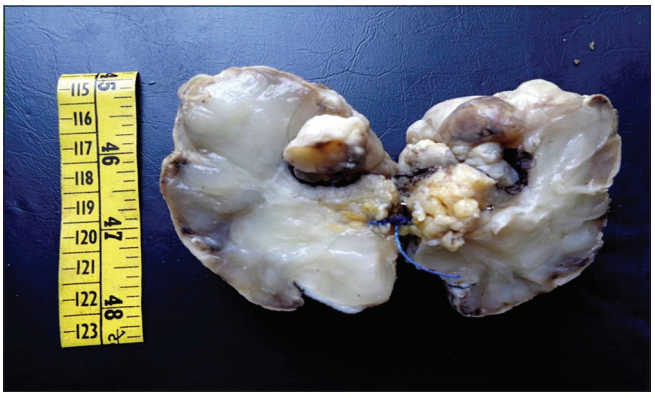

Figure 1: Gross-Aggressive angiomyxoma showing glistening grey white and focal brown areas

a relative percentage of 3.7% among the soft tissue tumors. Among the 80 myxoid soft tissue tumors, benign and malignant myxoid tumors numbered about 23 (28.75%) and 57 (71.25%), respectively. Thus, the ratio of malignant and benign myxoid soft tissue tumors was 2.5:1.

Table 1: Age-wise distribution of myxoid soft tissue tumors

Age in years	Benign tumors	Malignant tumors
<20	1 (4.35%)	3 (5.26%)
20-40	12 (52.17%)	17 (29.82%)
41-60	8 (34.78%)	26 (45.61%)
>60	2 (8.7%)	11 (19.31%)
Total	23 (100%)	57 (100%)

Table 2: Site distribution of myxoid soft tissue tumors

Site	Benign tumors	Malignant tumors
Upper extremities	8 (34.78%)	7 (12.28%)
Lower extremities	7 (30.43%)	29 (50.88%)
Trunk	2 (8.70%)	9 (15.79%)
Head & neck	3 (13.04%)	3 (5.26%)
Genital region	2 (8.70%)	1 (1.75%)
Inguinal region	1 (4.35%)	Nil
Retroperitoneum	Nil	7 (12.28%)
Mediastinum	Nil	1 (1.75%)
Total	23 (100%)	57 (100%)

Table 3: Size distribution of myxoid soft tissue tumors

Tumor size (cm)	Benign tumors	Malignant tumors
<5 cm	16 (69.57%)	5 (8.77%)
5--10 cm	4 (17.39%)	37 (64.91%)
>10 cm	3 (13.04%)	15 (26.32%)
Total	23 (100%)	57 (100%)

Table 4: Depth of myxoid soft tissue tumors

Depth	Benign tumors	Malignant tumors
Superficial	22 (95.65%)	25 (43.86%)
Deep	1 (4.35%)	32 (56.14%)
Total	23 (100%)	57 (100%)

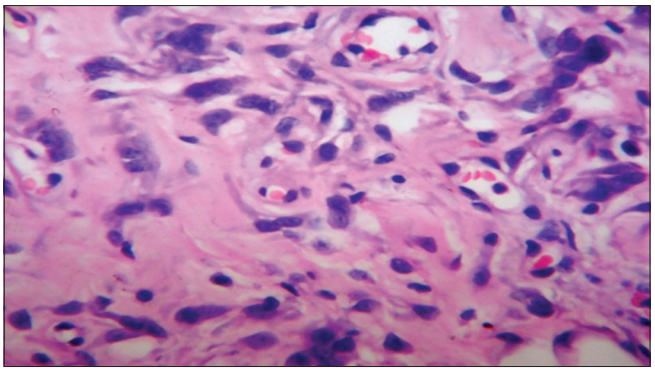

Figure 2: H and E, 40x- Aggressive angiomyxoma showing variable-sized blood vessels with perivascular hyalinization and stellate cells in a myxoid matrix

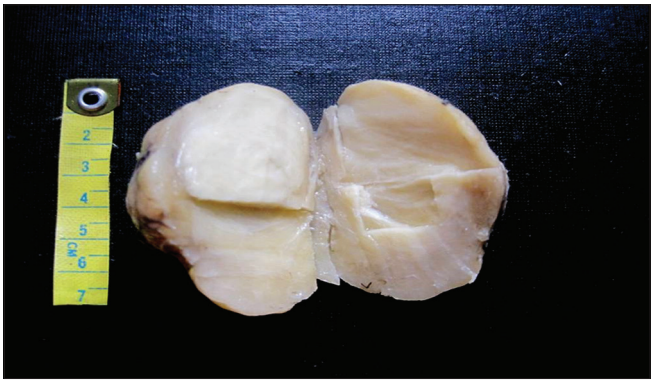

Figure 3: Gross- Myxolipoma showing yellowish and glistening grey, white cut surface

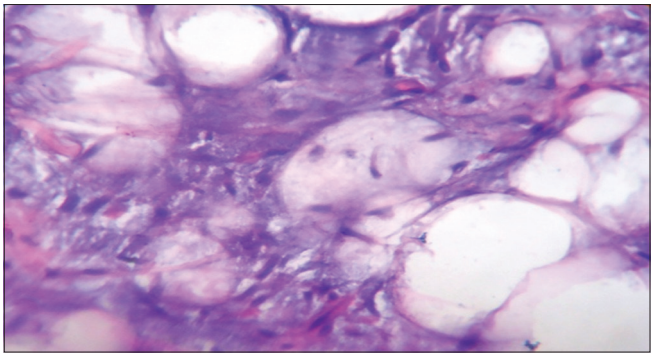

Figure 4: H and E, 40x- Myxolipoma showing mature adipocytes and spindle cells in a myxoid matrix

Table 5: Histomorphological distribution of myxoid soft tissue tumors

Benign (*n*=23)		Malignant (*n*=57)	
Histological Type	**No. of cases (%)**	**Histological Type**	**No. of cases (%)**
Myxoid neurofibroma	8 (34.78%)	Myxofibrosarcoma	19 (33.33%)
Cutaneous myxoma	6 (26.09%)	Myxoid Liposarcoma	15 (26.32%)
Aggressive angiomyxoma	3 (13.04%)	Myxoid Malignant Peripheral Nerve Sheath Tumor (MPNST)	8 (14.04%)
Myxoid neurothekeoma	2 (8.70%)	Myxoid Dermato fibrosarcoma protuberans (DFSP)	4 (7.02%)
Myxolipoma	2 (8.70%)	Low grade fibro myxoid sarcoma	4 (7.02%)
Intra-muscular myxoma	1 (4.35%)	Synovial sarcoma with myxoid change	3 (5.26%)
Schwannoma with myxoid change	1 (4.35%)	Extra-skeletal myxoid chondrosarcoma	3 (5.26%)
-	-	Myxoid Leiomyosarcoma	1 (1.75%)

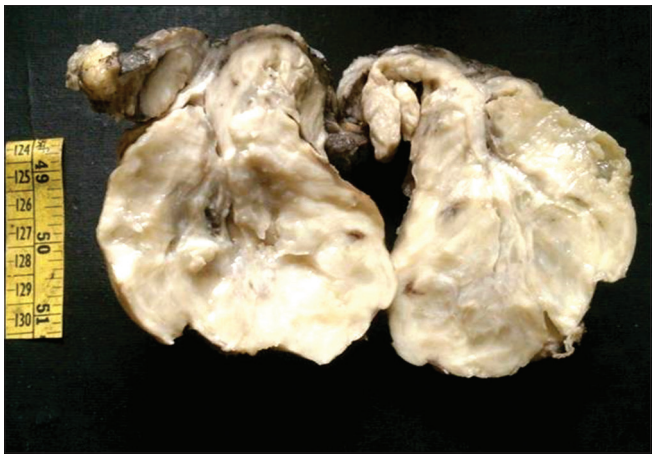

Figure 5: Gross- Myxofibrosarcoma showing glistening grey, white cut surface

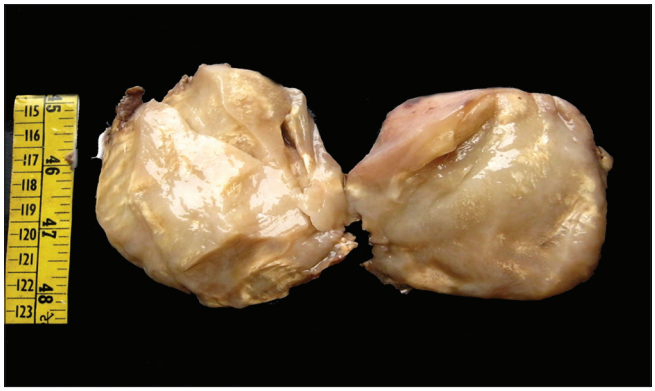

Figure 7: Gross- Low grade fibromyxoid sarcoma showing glistening grey, white cut surface

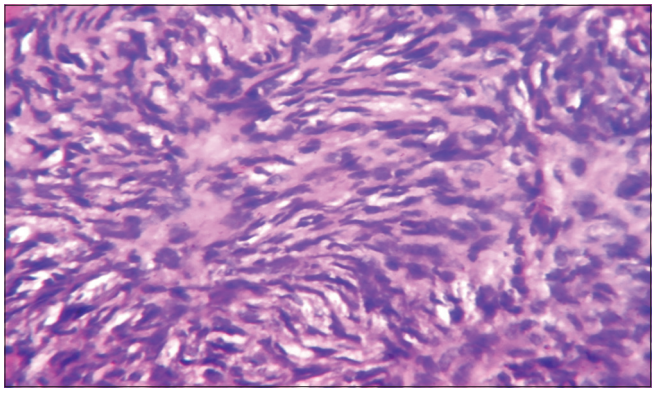

Figure 9: H and E, 40x- Myxoid DFSP showing spindle cells in storiform pattern in a myxoid stroma

In this study, 20--40 years was the most common age group for benign myxoid soft tissue tumors while majority (45.61%) of malignant myxoid soft tissue tumors were encountered in 41-60 years of age [Table 1]. Myxoid soft tissue tumors showed male sex predominance with a male: female ratio of 1.9:1 among the benign tumors and 2.8:1 among the malignant myxoid tumors of soft tissue.

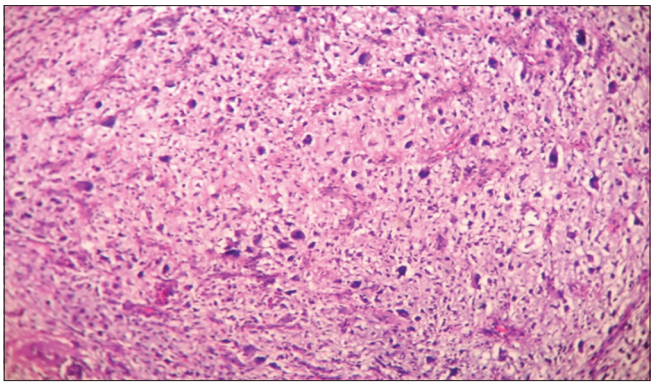

Figure 6: H and E, 10x- Myxofibrosarcoma showing atypical spindle cells, branching thick-walled capillaries and tumor giant cells in a myxoid matrix

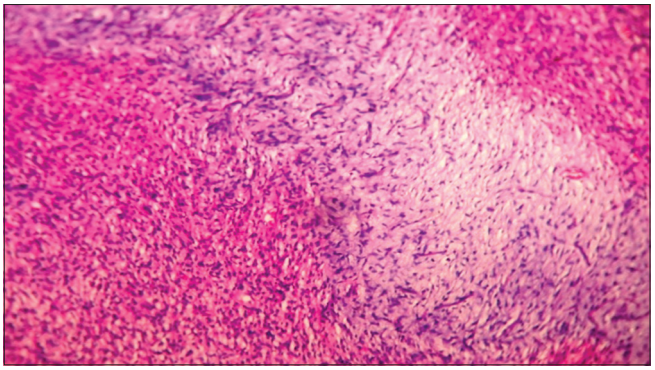

Figure 8: H and E, 10x- Low grade fibromyxoid sarcoma showing alternating fibrous and myxoid areas

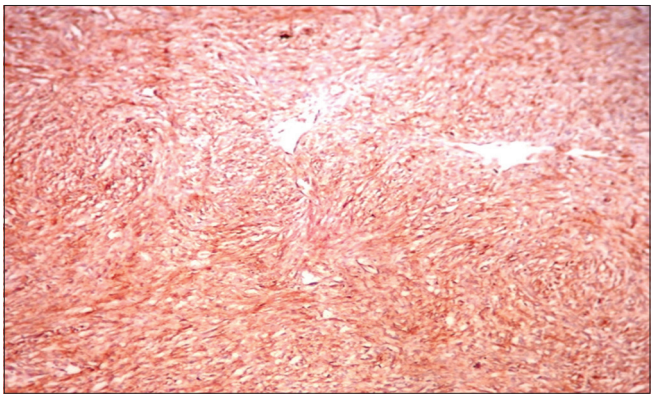

Figure 10: IHC, 10x Myxoid DFSP showing CD34 positivity in spindle cells

In this study, the most common site for benign myxoid soft tissue tumors was the upper extremities which constituted 8 cases with a relative percentage of about 34.78% among the benign myxoid tumors. Myxoid sarcomas showed a predilection for the lower extremities which constituted nearly half the number of cases among the 57 myxoid sarcomas with a relative percentage of 50.88% [Table 2].

Majority of the benign myxoid soft tissue tumors (69.57%) were less than 5 cm in size while 64.91%

Table 6: Distribution of histological grade among the various histological types of myxoid sarcomas

Histological type	Total	Grade-I	Grade-II	Grade-III
Myxofibrosarcoma	19	0	11 (57.9%)	8 (42.1%)
Myxoid liposarcoma	15	4 (26.7%)	11 (73.3%)	0
Myxoid MPNST	8	1 (12.5%)	5 (62.5%)	2 (25%)
Myxoid DFSP	4	4 (100%)	0	0
Low grade fibromyxoid sarcoma	4	1 (25%)	3 (75%)	0
Extra-skeletal myxoid chondrosarcoma	3	0	3 (100%)	0
Synovial sarcoma with myxoid change	3	0	0	3 (100%)
Myxoid leiomyosarcoma	1	0	1 (100%)	0
Total	57	10 (17.54%)	34 (59.65%)	13 (22.81%)

Table 7: Distribution of tumor depth among the various histological types of myxoid sarcomas

Histological type	Total	Superficial	Deep
Myxofibrosarcoma	19	18 (94.7%)	1 (5.3%)
Myxoid liposarcoma	15	0	15 (100%)
Myxoid MPNST	8	2 (25%)	6 (75%)
Low grade fibromyxoid sarcoma	4	0	4 (100%)
Myxoid DFSP	4	4 (100%)	0
Extra-skeletal myxoid chondrosarcoma	3	1 (33.3%)	2 (66.7%)
Synovial sarcoma with myxoid change	3	0	3 (100%)
Myxoid leiomyosarcoma	1	0	1 (100%)
Total	57	25 (43.9%)	32 (56.1%)

Table 8: Correlation of tumor size and depth of myxoid sarcomas

Tumor size	Superficial	Deep	Total	Pearson Chi-square test
≤ 5 cm	10 (66.7%)	5 (33.3%)	15 (100%)	Chi-square value:
>5 cm	15 (35.7%)	27 (64.3%)	42 (100%)	4.300
Total	25 (43.85%)	32 (56.14%)	57 (100%)	*P*: 0.038

Table 9: Correlation of tumor grade and vascular invasion among myxoid sarcomas

Histological grade	Vascular invasion		Total	Pearson Chi-square test
	Present	Absent		
Grade-I	0	10 (100%)	10 (100%)	Chi-square
Grade-II	3 (8.8%)	31 (91.2%)	34 (100%)	value: 8.827
Grade-III	5 (38.5%)	8 (61.5%)	13 (100%)	
Total	8 (14%)	49 (86%)	57 (100%)	*P*: 0.012

of myxoid sarcomas were 5--10 cm in size and 26.32% of myxoid sarcomas were more than 10 cm in size [Table 3]. In this study, location of the tumors with respect to the fascia and skeletal muscles varied considerably between the benign and malignant myxoid soft tissue tumors. Almost all the benign myxoid soft tissue tumors except a single case of intramuscular myxoma were superficial in location with a relative percentage of 95.65% among the benign tumors. Out

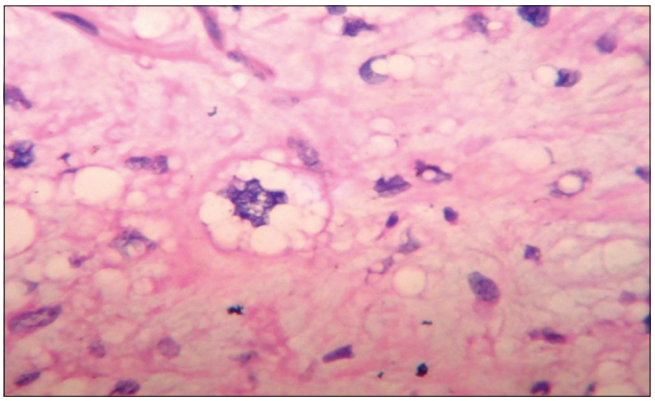

Figure 11: H and E, 40x- Myxoid liposarcoma showing lipoblasts and spindle cells in a myxoid stroma

of the total 57 myxoid sarcomas, 32 (56.14%) were deeper in location while 25 (43.86%) were superficial in location [Table 4].

Histomorphologically, seven types of benign myxoid soft tissue tumors and eight types of malignant tumors were recognized in this study. Among the benign myxoid soft tissue tumors, myxoid neurofibroma constituted eight cases with a relative percentage of 34.78% followed by cutaneous myxoma which accounted for six cases (26.09%). Out of the eight histological types of myxoid sarcomas encountered in this study, the most frequent histological type was myxofibrosarcoma with a total of 19 cases and a relative percentage of 33.33%. The next most common histological type was myxoid liposarcoma which constituted 15 cases with a relative percentage of 26.32% among the myxoid sarcomas [Table 5].

All the 57 myxoid sarcomas in the study were graded according to the FNCLCC grading system, out of which 10 cases (17.54%) were in grade I, 34 cases (59.65%) were in grade II and 13 cases (22.81%) were in grade III category. Vascular invasion was present in 14.04% of myxoid sarcomas while bone invasion was seen in 1.75% of myxoid sarcomas. Local recurrence at presentation was observed in 14.04% of myxoid sarcomas.

Table 10: Comparison of distribution of histological types of benign myxoid soft tissue tumors among other studies

Turkish study[13]	Kransdorf M J[16]	Current study
Benign neural tumors (14%)	Neurofibroma (5%)	Myxoid neurofibroma (34.78%)
		Myxoid neurothekeoma (8.70%)
	Schwannoma (5%)	Schwannoma with myxoid change (4.35%)
Myxoma (1.5%)	-	Cutaneous myxoma (26.09%)
		Intra-muscular myxoma (4.35%)
		Aggressive angiomyxoma (13.04%)
Lipoma (21.5%)	Lipoma & variants (16%)	Myxolipoma (8.70%)
Vascular lesions (37.5%)	Hemangioma (8%)	-
Benign Fibrous Histiocytoma (2%)	Benign Fibrous Histiocytoma (13%)	-

Table 11: Comparison of distribution of histological types of myxoid sarcomas among other studies

Turkish study[13]	Coindre *et al*[9]	Current study
Pleomorphic sarcoma (24.5%)	Malignant Fibrous Histiocytoma (28.2%)	Myxofibrosarcoma (33.33%)
Liposarcoma (16.4%)	Liposarcoma (15.2%)	Myxoid liposarcoma (26.32%)
MPNST (6.6%)	MPNST (5.8%)	Myxoid MPNST (14.04%)
DFSP (1.8%)	-	Myxoid DFSP (7.02%)
Synovial sarcoma (13%)	Synovial sarcoma (10%)	Synovial sarcoma with myxoid change (5.26%)
Leiomyosarcoma (4.9%)	Leiomyosarcoma (12%)	Myxoid leiomyosarcoma (1.75%)
-	-	Low grade fibromyxoid sarcoma (7.02%)
-	-	Extra-skeletal myxoid chondrosarcoma (5.26%)

The distribution of histological grade and tumor depth among the various histological types of myxoid sarcomas are given in Tables 6 and 7, respectively. Majority of the myxofibrosarcomas and myxoid DFSP were superficial in location with a relative percentage of 94.7% and 100%, respectively, as against the other myxoid sarcomas which were predominantly deep-seated in this study.

In this study, tumor size was correlated with the depth of myxoid sarcomas. Around 66.7% of tumors ≤5 cm in size were superficially located and 64.3% of tumors more than 5 cm in size were deep-seated. The correlation was statistically significant with a *P* value of 0.038 [Table 8]. In this study of 57 myxoid sarcomas, tumor location (Depth) with respect to the fascia and skeletal muscles was compared with the histological grade (FNCLCC grade). Pearson Chi-square test gave a *P* value of 0.245 which was statistically insignificant. Thus, there was no significant correlation between depth of myxoid sarcomas and histological grade (FNCLCC grade) in this study.

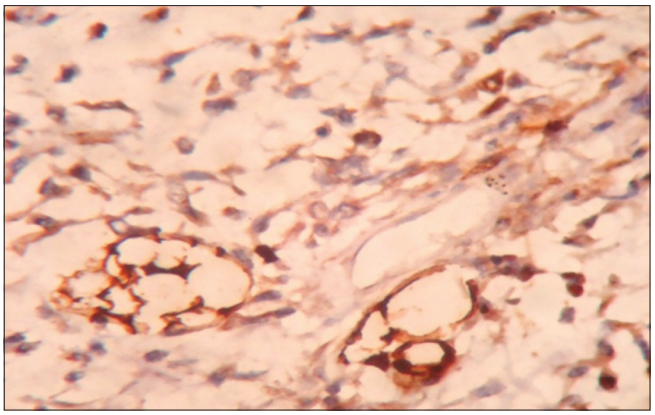

Figure 12: IHC, 40x- Myxoid liposarcoma showing S100 positivity in lipoblasts

In this study, 8.8% of Grade-II sarcomas and 38.5% of Grade-III sarcomas showed vascular invasion while none of the Grade-I sarcomas had vascular invasion. There was a significant correlation (*P*-value: 0.012) between FNCLCC grade of myxoid sarcomas and presence of vascular invasion [Table 9].

In this study, bone invasion was present in only one case (2.9%) of Grade-II sarcomas while none of the Grade-I and Grade-III sarcomas showed bone invasion. The correlation between histological grade (FNCLCC grade) of myxoid sarcomas and presence of bone invasion was statistically insignificant in this study with a *P* value of 0.709. Of the 57 myxoid sarcomas, 11.8% of Grade-II sarcomas and 30.8% of Grade-III sarcomas had local recurrence at presentation. None of the Grade-I sarcomas had local recurrence at presentation in this study. The correlation of histological grade (FNCLCC grade) of myxoid sarcomas and local recurrence at presentation was statistically insignificant with a *P* value of 0.091.

Immunohistochemical analysis of benign myxoid soft tissue tumors revealed CD34 positivity in cutaneous myxoma, intramuscular myxoma and aggressive angiomyxoma, while S100 was positive in myxolipoma, myxoid schwannoma, myxoid neurofibroma, and neurothekeoma. Among the myxoid sarcomas, consistent S100 positivity was observed in myxoid liposarcoma, myxoid MPNST and extra-skeletal myxoid chondrosarcoma. Myxoid DFSP showed CD34 positivity,

while synovial sarcoma with myxoid change showed Pan CK positivity. Myxoid leiomyosarcoma was positive for SMA, desmin and Pan CK. Myxofibrosarcoma and Low grade fibromyxoid sarcomas showed no consistent positivity for immunohistochemical markers except for vimentin. The histopathological and IHC images of the various myxoid soft tissue tumors encountered in this study are given in Figures 1 to 12.

Discussion

The relative percentage of soft tissue tumors was 4.56% in the present study and the myxoid soft tissue tumors accounted for 3.7% among the soft tissue tumors. In this study, benign soft tissue tumors (91.6%) vastly outnumbered the malignant soft tissue tumors (8.37%) which was almost similar to the WHO statistics.[11] However the relative percentage of benign and malignant myxoid soft tissue tumors was 28.8% and 71.2%, respectively. Thus, malignant myxoid soft tissue tumors were more frequent than benign myxoid soft tissue tumors unlike the overall distribution of soft tissue tumors.

In the present study, 52.17% of benign myxoid soft tissue tumors occurred in young adults (20--40 years) while 50.9% of myxoid sarcomas occurred in patients ≥50 years of age. As per WHO statistics,[11] there was a relationship between the histological type of benign soft tissue tumors and patient's age with majority of them occurring in young to middle-aged adults while the median age for sarcomas was 65 years. In a study of 1,331 benign soft tissue tumors by Myhre-Jensen *et al*,[12] the mean patient age was 44.5 years. In a study of 1,240 sarcomas by Coindre *et al*,[9] 52.7% of cases occurred in patients ≥50 years of age which was similar to the present study.

In the current study, a male sex preponderance was observed among the individuals diagnosed with benign (M:F—1.9:1) and malignant (M:F—2.8:1) myxoid tumors of soft tissue which was similar to the WHO population based statistics.[11] Coindre *et al.*[9] reported a Male: Female ratio of 1.06:1 for sarcomas. Another study from Turkey[13] also showed predominance of sarcomas among males (M:F—1.22:1).

The most common site for benign myxoid soft tissue tumors was upper extremities (34.78%) while lower extremities (50.88%) was the most common site for myxoid sarcomas in this study. The SEER program of the National Cancer Institute reported 45% of sarcomas involving the lower extremities in their study of 6883 sarcomas.[14]

Around 95.65% of benign myxoid soft tissue tumors were superficially located and 69.57% of cases had dimensions less than 5 cm. As per WHO,[11] 99% of benign soft tissue tumors were superficial and 95% of cases were less than 5 cm in size. Among the 57 myxoid sarcomas, 32 (56.14%) cases were deep-seated, and 42 (73.68%) cases were more than 5 cm in size in this study. Coindre *et al*.[9] reported almost 76% of sarcomas with dimensions more than 5 cm and nearly 86% of sarcomas were deeply situated in their study. Trovik *et al*.[15] reported an increase in the relative risk of metastasis by 1.5% for every 5 cm increase in tumor size.

Of the total 23 benign myxoid soft tissue tumors, myxoid neurofibroma (34.78%) was the most common histological type followed by cutaneous myxoma (26.09%). Though there was no available literature reference on the exact prevalence of benign myxoid soft tissue tumors as such, one study from Turkey[13] and another by Kransdorf M J[16] had reported the general prevalence of benign soft tissue tumors which included the myxoid tumors as well [Table 10]. In both these studies, the bulk of the tumors belonged to the vascular and adipocytic category, thus highlighting the relative rarity of benign myxoid soft tissue tumors.

Of the 57 myxoid sarcomas, myxofibrosarcoma (33.33%) was the most common histological type followed by myxoid liposarcoma (26.32%). One Turkish study[13] and another study by Coindre *et al*.[9] reported that the most prevalent sarcomas belonged to the category of Malignant Fibrous Histiocytoma (MFH) followed by liposarcoma [Table 11].

Of the 57 myxoid sarcomas in this study, 10 (17.54%) were Grade-I, 34 (59.65%) were Grade-II and 13 (22.81%) were Grade-III sarcomas. This was not concurrent with Coindre *et al*.[9] observations of 12.7% of Grade-I, 41.2% of Grade-II and 46.1% of Grade-III sarcomas. Thus, myxoid sarcomas showed lower histological grades when compared with grades of sarcomas in general. Invasion of bone or vascular structures was present in 18.5% of cases in the study by Coindre *et al*.[9] while 15.8% of sarcomas showed vascular or bone invasion in the present study.

The correlation between tumor size and depth of myxoid sarcomas was statistically significant in the present study with a *P* value of 0.038. This was in concurrence with the study of 490 sarcomas by Anders Rydholm and Pelle Gustafson[17] (*P*-value: 0.000). Histological grade was correlated with the depth of myxoid sarcomas, and the correlation was found to be statistically insignificant (*P*-value: 0.245) in the present study. This was almost similar to the study by Anders Rydholm and Pelle Gustafson[17] (*P* value: 0.889).

In this study, vascular invasion was frequent in high grade sarcomas and this correlation was statistically significant in the present study (P-value: 0.012). As per Lack *et al*,[18] vascular invasion (*P*-value: 0.003) was associated with malignant behavior and higher histological grade of sarcomas concordant to the present study. Another study by Merimsky *et al*.[19] also showed that presence of vascular invasion was always associated with locally advanced sarcoma of higher grade.

There was no significant correlation between histological grade and presence of bone invasion (*P*-value: 0.709) in this study. This was similar to the analysis of 874 sarcomas by Ferguson *et al*.[20] (*P*-value: 0.150). No significant association between locally recurrent myxoid sarcomas and histological grade was found in this study (*P*-value: 0.091) which was concurrent with the study of *P* Gustafson[21] (*P*-value: 0.08). Immunohistochemical results were concordant to the observations of Weiss SW and Goldblum JR.[6]

Limitations

This study had few limitations like lack of follow-up data and absence of community-based analysis. Future studies directed upon the community with longer follow-up period would throw light on the exact prevalence and behavior of this relatively rare group of tumors in the community. Further research on determining the exact tumor-specific biological composition of the myxoid matrix in various myxoid neoplasms could lead to the identification of tumor-specific markers and eventually the development of a more feasible detection method, thereby ensuring accurate histopathological diagnosis.

Conclusion

Myxoid soft tissue tumors are rare and exhibit overlapping clinical and histomorphological features. The biological behavior of this group of tumors varies considerably including benign, locally aggressive and distantly metastasizing malignant tumors, thus highlighting the importance of a precise diagnosis in this challenging group of tumors. A multimodality approach that includes careful clinical examination and radiological evaluation along with diligent light microscopic histopathological examination aided by ancillary techniques like IHC and molecular studies will help in arriving at the correct diagnosis. In case of myxoid sarcomas, a thorough assessment of the various prognostic variables like tumor size, depth, grade, and stage is of utmost importance for effective management of the patients.

Ethical approval

Obtained

Key messages

Malignant myxoid soft tissue tumors were more frequent than benign myxoid soft tissue tumors. Majority of large sized myxoid sarcomas were deep-seated and vascular invasion was frequent in myxoid sarcomas of higher histological grade.

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Conflicts of interest

There are no conflicts of interest.

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