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Pathology

LOW GRADE FIBROMYXOID SARCOMA - AN UNUSUAL CLINICAL PRESENTATION

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ABSTRACT Low grade fibromyxoid sarcoma (LGFMS) is a rare soft tissue sarcoma (5% incidence), and its occurrence in the head and neck region is uncommon (0.5- 1.5%) , especially in adolescents. Despite its bland histology, comprising uniform spindle cells in alternating fibrous and myxoid areas; LGFMS carries significant recurrence and metastatic potential. It's important that it can arise in the neck and should be considered in the differential diagnosis of deep cervical masses. LGFMS shows strong MUC4 immunopositivity and FUS-CREB3L2 fusion gene. We report a rare case of cervical LGFMS in a 14-year-old female, emphasizing its unusual site and age of presentation.

KEYWORDS : Low grade fibromyxoid sarcoma, Evans tumor, soft tissue sarcoma, fibroblastic neoplasm.

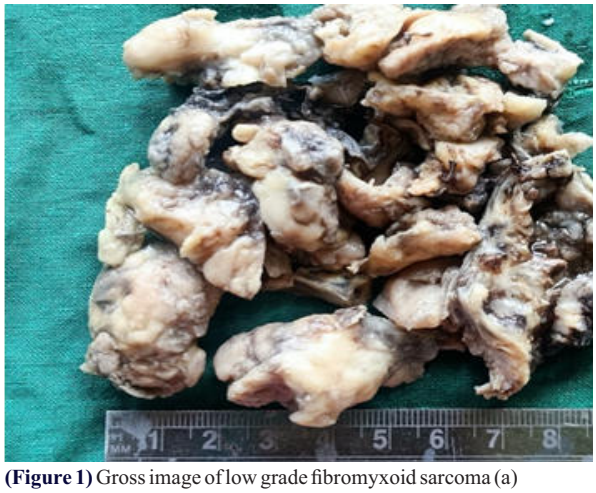
INTRODUCTION

Low grade fibromyxoid sarcoma (LGFMS) is an uncommon malignant fibroblastic tumor comprising about 5% of soft tissue sarcomas[2-3]. First described by Evans in 1987, it is therefore referred to as Evans tumor[3]. It most often affects young to middle-aged adults (mean age 34 years), although pediatric cases have been reported[10]. A slight male predominance is noted[3]. The tumor most frequently arises in the proximal extremities and trunk, and less commonly in the retroperitoneum, abdominal cavity, axilla, mediastinum, or head and neck region[1-7]. LGFMS typically exhibits the t(7;16)(q32–34;p11) translocation resulting in FUS-CREB3L2 fusion[4]. Rare alternative rearrangements include FUS-CREB3L1 and EWSR1-CREB3L1[4]. Although histologically bland, LGFMS shows high long-term recurrence (64%), metastasis (45%), and disease-related mortality (42%)[2-3-4], underscoring the need for early recognition and appropriate management.

CASE HISTORY

A 14-year-old female presented with a gradually progressive left-sided neck swelling since 6 months. Clinical examination revealed a soft, mobile, non-tender mass measuring approximately 10 × 8 cm in the left cervical region. Radiological findings showed an ill-defined, heterogeneous solid soft tissue mass measuring 8 × 7.5 × 2.5 cm. Fine-needle aspiration cytology (FNAC) revealed features of a malignant spindle cell lesion. A core biopsy demonstrated similar findings. The mass was subsequently excised in toto and submitted for detailed histopathological evaluation.

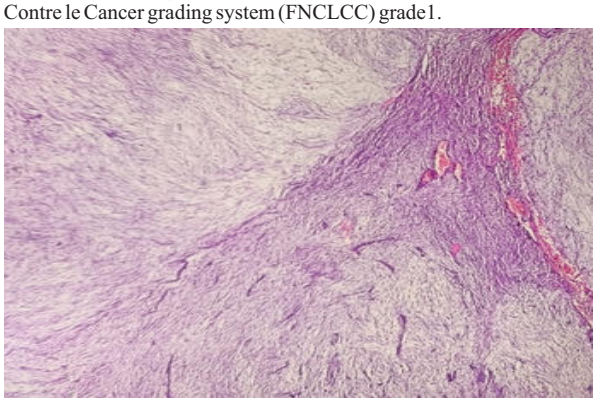
Gross examination consisted of multiple grey-white to grey-brown soft tissue fragments together measuring 8 × 8 × 3 cm. The cut surface was firm with mucoid areas **[Figure 1].**



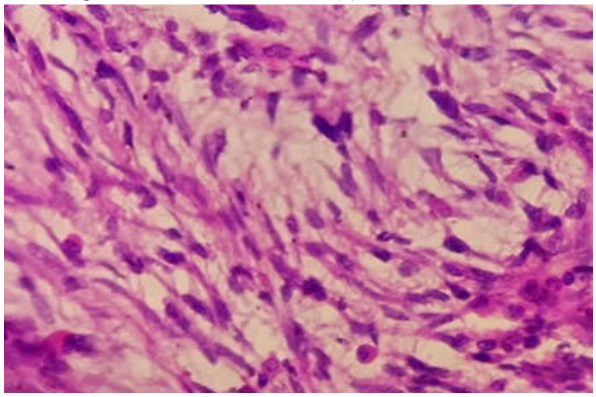
(Figure 1) Gross image of low grade fibromyxoid sarcoma (a)

Microscopically, the lesion comprised of alternating fibrous and myxoid areas **[Figure 2]** with tumor cells arranged in short fascicles which are spindle shaped having elongated nuclei with pointed ends and moderate eosinophilic cytoplasm **[Figure 3].** These cells are admixed with areas of hemorrhage, inflammatory cells and arcades of blood vessels. Mitotic figures 2–3/HPF.

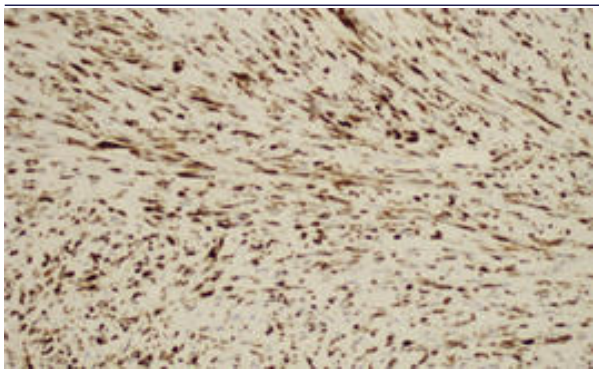
Differential diagnosis was Low grade Fibromyxoid sarcoma and Low grade myxofibrosarcoma. On Immunohistochemistry, tumor cells showed strong MUC4 cytoplasmic positivity **[Figure 4].** CD99 membrane positivity **[Figure 5],** focal EMA cytoplasmic positivity **[Figure 6].** Also negativity with markers like CD34 **[Figure 7],** SMA and S100 are seen. A final diagnosis of **Low grade fibromyxoid sarcoma (LGFMS)** with Federation Nationale des Centres de Lutte Contre le Cancer grading system (FNCLCC) grade1.



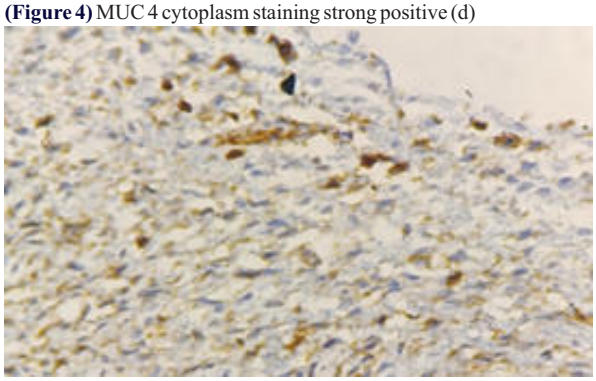
(Figure 2) H & E 4X shows alternating fibrous and myxoid areas along with elongated curvilinear blood vessel (b)



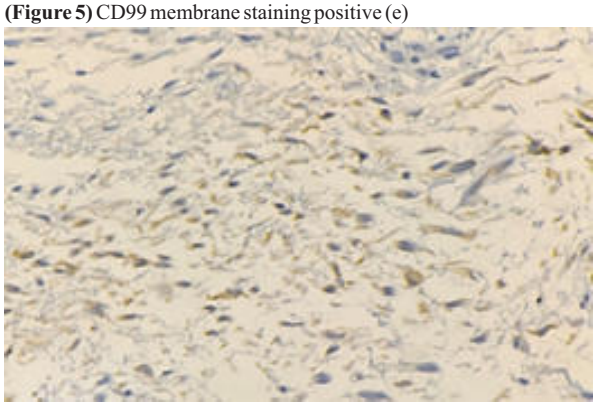
(Figure 3) H & E 10X shows spindle cells with elongated nuclei and pointed ends .(c)



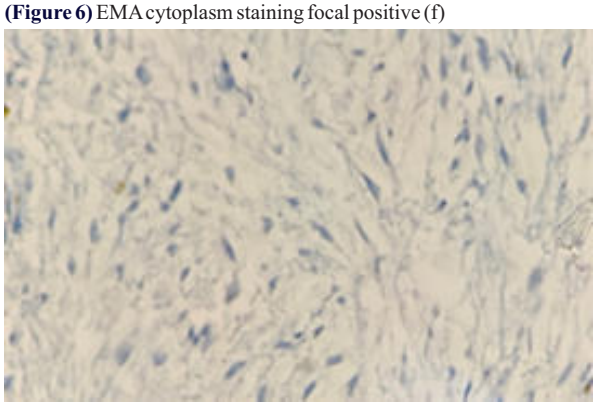
(Figure 4) MUC 4 cytoplasm staining strong positive (d)



(Figure 5) CD99 membrane staining positive (e)



(Figure 6) EMA cytoplasm staining focal positive (f)



(Figure 7) CD 34 membrane staining negative (g)

DISCUSSION

Low grade fibromyxoid sarcoma (LGFMS) is an uncommon soft tissue sarcoma characterized by deceptively bland morphology yet aggressive biological behavior. Although classically arising in the deep soft tissues of the trunk and proximal extremities, its occurrence in the head and neck region is distinctly rare, accounting for only 0.5–1.5% of all reported cases[1,7]. The typical age of presentation is young to middle adulthood; pediatric cases constitute a minority, with fewer than 20% occurring in individuals younger than 18 years. Therefore, the presence of LGFMS in an adolescent patient, particularly in the cervical region, represents an unusual clinical scenario and underscores the necessity of maintaining a broad differential diagnosis when evaluating deep neck masses.

Gross findings show well circumscribed or infiltrating lesion with lobulated or mild bosselated external surface. Cut surface – fibrous and myxoid areas , gelatinous with firm consistency **[Table 1].**

Histologically, LGFMS is composed of uniform spindle cells arranged in whorled, swirling, or linear fascicles embedded within alternating fibrous and myxoid zones **[Table 2].** Despite its bland appearance and the fact that most tumors fall within FNCLCC Grade 1, LGFMS has well-documented potential for late recurrence and distant metastasis. Historically, hyalinizing spindle cell tumor with giant rosettes (HSTGR) was described as a separate neoplasm; however, owing to shared histological patterns and identical molecular alterations, it is now recognized within the LGFMS spectrum[2]. Both subtypes exhibit similar immunohistochemical and genetic profiles, supporting their unified classification.

Immunohistochemistry plays a crucial role in diagnosis, particularly the **strong and diffuse expression of MUC4**, which is considered highly sensitive and specific for LGFMS[4]. Additional markers such as EMA and CD99 may show variable positivity. Negative staining for CD34, S100, SMA, desmin, and caldesmon **[Table 3]** is helpful in excluding other spindle-cell lesions, including dermatofibroma, peripheral nerve sheath tumors, and smooth muscle neoplasms[3]. On molecular testing, approximately **90%** of cases demonstrate the **characteristic FUS-CREB3L2 rearrangement**, while a smaller proportion harbor FUS-CREB3L1 or EWSR1-CREB3L1 gene fusions[4], all of which reinforce the diagnosis.

Clinically, LGFMS demonstrates a paradoxical course: although slow-growing and indolent in the early phase, it displays high rates of recurrence (up to 60%), metastasis (up to 45%), and disease-related mortality[2,3,4]. Notably, metastasize most commonly to lungs and pleura may manifest decades after excision, sometimes 40–50 years later, highlighting the tumor's unpredictable long-term behaviour [2]. Complete surgical excision with negative margins remains the mainstay of treatment. Given the propensity for late metastatic disease, long-term surveillance is essential, and chest CT imaging is recommended at diagnosis and during follow-up.[2]

Table 1 Gross Features With Other Differential Diagnosis Of LGFMS

GROSS	LGFMS	MYXOFIBROSARCOMA
External surface	Well circumscribed	Multinodular
Cut surface	fibrous and focally myxoid	Lobulated, gelatinous and myxoid, firm nodules Infiltrative Hemorrhage and necrosis +
Consistency	Firm	Firm

LGFMS – low grade fibromyxoid sarcoma

Table 2 Histopathological Features Of Differential Diagnosis Of LGFMS

LGFMS	MYXOFIBROSARCOMA
Alternating fibrous and myxoid areas with bland spindle cells in collagenized stroma.	Hypocellular and hypercellular areas with spindle shaped cells with abundant myxoid stroma. Elongated curvilinear blood vessels. Cell morphology ranges from low to high grade.

LGFMS – low grade fibromyxoid sarcoma

Table 3 Comparison Of IHC And Cytogenetics In Differential Diagnosis Of LGFMS

IHC and Cytogenetics	LGFMS	MYXOFIBROSARCOMA
MUC4	+	-
EMA	Focal +	-
CD99	+	-
CD34	-	Focal +
S100	-	-
SMA	-	Focal +
Cytokeratin	-	-
Cytogenetics	t(7;16)(q32-34;p11)	Gain of chromosome 5p

IHC – immunohistochemistry; LGFMS – low grade fibromyxoid sarcoma

CONCLUSION

LGFMS occurring in adolescents and in the head and neck region is exceptionally rare, contributing to diagnostic difficulty. Recognition of its characteristic histopathological, immunohistochemical (MUC4), and molecular features is critical for accurate diagnosis. Given its potential for late recurrence and metastasis, complete surgical excision and extended long-term follow-up remain essential components of management.

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