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Low-Grade Fibromyxoid Sarcoma–A Case Report and Review of Literature

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ABSTRACT Low-grade fibromyxoid sarcoma (LGFMS) is a rare neoplasm with a tendency to develop in deep soft tissue of young adults. [1] It is also referred to as "Evans tumor", "Hyalinizing spindle cell tumor with giant rosettes" and "Low-grade fibrosarcoma with palisaded granuloma like bodies". [2,3] LGFMS is considered a diagnostic dilemma because of its innocuous and varied histological features that can be potentially confused with other benign or low-grade fibromyxoid lesions. This report is aimed at reinforcing the need to recognize LGFMS as a sarcoma despite its deceptively benign histological appearance.

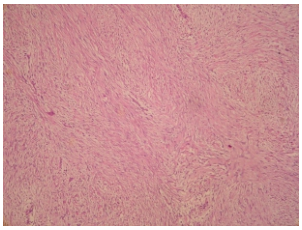
KEYWORDS : Sarcoma , Myxoid , Rosettes and Stellate

Case Report:

A 20-year-old female patient presented to the surgery Out Patient Department with a painless, deeply situated mass in the proximal extremity (right forearm), present for 5 years. Patient was referred to the pathology department for Fine Needle Aspiration Cytology (FNAC). FNAC was suggestive of a benign mesenchymal lesion of neural or fibroblastic origin. This was followed by excision.

Pathological examination of the excised swelling revealed a grossly circumscribed, Unencapsulated tumor measuring 8 x5 x3 cms in size. The cut surface varied from being gray white, firm and fibrous to gelatinous or myxoid. Microscopy revealed cellular areas constituted by bland spindle shaped fibroblasts, alternating with less cellular myxoid areas. A typical pattern of intermixed, sweeping bands of fibrous and myxoid tissue, focal areas of storiform architecture and concentric perivascular

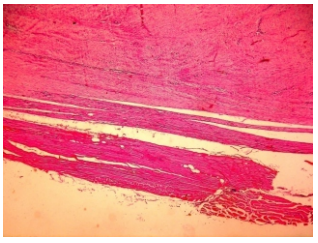
cuffs of slender spindle cells was seen (Figure 1).



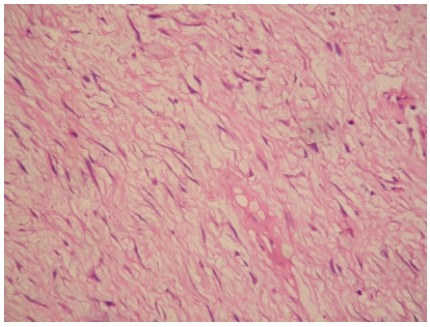
Low grade fibromyxoid sarcoma 100 X storiform pattern .(Figure 1)

Also noted was an occasional heavily collagenized area. The tumor lacked the nuclear anaplasia, mitotic activity, and necrosis generally associated with a sarcoma.

Microscopic infiltration into the surrounding muscle was however present (Figure 2 & 3).



Low grade fibromyxoid sarcoma 100 X surrounding muscle.(Figure 2)



Low grade fibromyxoid sarcoma 100 X myxoid areas.(Figure 3)

Based on the clinical history (swelling of prolonged duration) and histo-morphological features, a diagnosis of LGFMS was made.

Fewer than 150 cases of LGFMS have been reported so far. [3,4] Age of presentation varies from 6-54 years but most tumors arise between 25 to 46 years. Most common location is thigh, trunk & shoulder. It is rarely seen in the upper extremity.

LGFMS usually appears grossly circumscribed but microscopic infiltration is commonly observed. Characteristically seen are cellular fibrous areas abruptly transitioning into paucicellular myxoid areas with the former showing a swirling, whorled or storiform pattern. The myxoid areas are generally hypocellular, comprised of bland spindle or stellate cells having dark oval nuclei. Intranuclear inclusions may occasionally be seen. Pleomorphism is absent or slight. Mitotic figures are rarely seen. Collagen rosettes have been noted in some cases. Originally, these cases were reported as "hyalinizing spindle cell tumors with giant rosettes" and were thought to be different from LGFMS but now this entity is considered within the spectrum of LGFMS, as clinico-pathological features of the two lesions otherwise appear identical. [5]

A few histopathological deviations have been observed in some cases of LGFMS, namely:

- Focal hyper cellular areas with epithelioid cells
- Herringbone pattern

- Focal pleomorphism and necrosis
- Increased capillary vascularity with presence of curvilinear vessels
- Mitotic figures >1/10 HPF

Folpe et al [5] could not demonstrate a definite clinical significance of the above histological deviations on follow up of their patients; median duration of their follow up being 2 years.

LGFMS shows diffuse immuno positivity with Vimentin, rare positivity with Smooth Muscle Actin, & Desmin.[6] Entities to be considered in the differential diagnosis include **desmoid fibromatosis** (constituent cells of which are reactive fibroblasts arranged in a parallel/straighter alignment as compared to LGFMS), neurofibroma (differentiation is based on the presence of slender wavy nuclei and diffuse S100 Positivity in a **neurofibroma** as opposed to rare or absent S100 positivity in LGFMS) and low-grade myxoid MFH or **low-grade myxofibrosarcoma** which shows pleomorphic cells with bizarre nuclei & abnormal mitoses. **Intramuscular or juxta- articular myxoma** is differentiated from LGFMS based on lower cellularity, uniformly myxoid appearance, and lack of alternating fibrous & myxoid areas in the former.

REFERENCES:

1) Devaney DM, Dervan P, O'Neill S, Leader M. Low-grade fibromyxoid sarcoma. Histopathology. 1990;17:463-479.
2) Evans HL. Low-grade fibromyxoid sarcoma: a report of 12 cases. Am J Surg Pathol. 1993;17:595-600.
3) Goodland JR, Mentzel T, Fletcher CD. Low-grade fibromyxoid sarcoma: clinicopathological analysis of eleven new cases in support of a distinct entity. Histopathology. 1995;26:229-237.
4) Billings SD, Giblen G, Fanburg-Smith JC. Superficial low-grade fibromyxoid sarcoma (Evans tumor): a clinicopathological analysis of 19 cases with a unique observation in the pediatric population. Am J Surg Pathol. 2005;29:204-210.
5) Folpe AL, Lane KL, Paull G, Weiss SW. Low-grade fibromyxoid sarcoma and hyalinizing spindle cell tumor with giant rosettes: a clinicopathological study of 73 cases supporting their identity and assessing the impact of high grade areas. Am J Surg Pathol. 2000;24:1353-1360.
6) Fukunaga M, Ushigome S, Fukunaga N. Low-grade fibromyxoid sarcoma. Virchows Arch. 1996;429:301-303.