Case Report

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Pathology Section

Low Grade Fibromyxoid Sarcoma Presented with Local Recurrence and an Extensive Foot Deformity

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ABSTRACT

Low Grade Fibromyxoid Sarcoma (LGFMS) is a rare neoplasm commonly affects young adults and which typically arises in the deep soft tissues of the proximal extremities. This tumour has the potential for local recurrence or distant metastasis. We are presenting here, the case of a 70-year old male who developed a large ulceroproliferative nodular mass in the dorsum of the left foot, which led to an extensive foot deformity with local recurrence. Careful consideration of light microscopy in view of its bland histopathological findings and immunohistochemical features is important to distinguish LGFMS from a number of other benign and malignant soft tissue neoplasms, for the proper management of the patient.

Key Words: Low grade fibromyxoid sarcoma (LGFMS), Soft tissue tumour, Low grade myxofibrosarcoma

INTRODUCTION

Low Grade Fibromyxoid Sarcoma (LGFMS) is a distinctive variant of fibrosarcoma. LGFMS which was first described by Evans[1] in 1987, is a rare soft tissue sarcoma which exhibits bland histological features but a paradoxically aggressive behaviour by showing a high rate of local recurrence (33%) and metastasis(58%) [2].

Over the last two decades, roughly 150 cases have been documented with local recurrence, metastasis, and death [3]. A predominance in males and young adult groups which were between the ages of 25-46 years was evident, as was reported by Goodlad et al [4].

The tumour most commonly arises in the deep soft tissues of the proximal and lower extremities, particularly in the thigh [1,5].

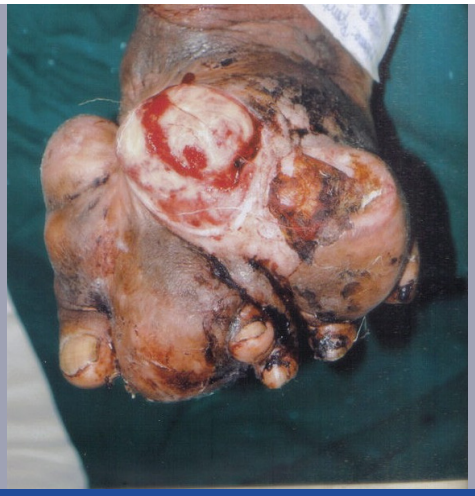
We are reporting here a case of a 70-old year male who presented with a superficial ulceroproliferative and nodular mass, with local pain, extensive foot deformity and local recurrence. The surgically Surgically excised mass on histopathology revealed features suggestive of LGFMS.

CASE REPORT

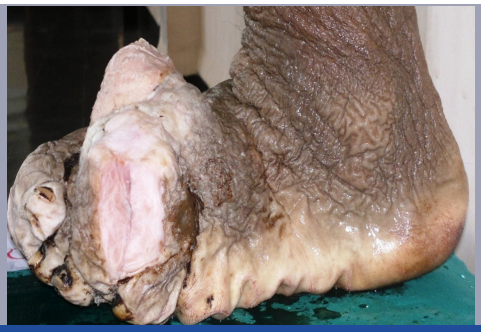
A 70-year-old male patient came to the surgical OPD of our hospital with the chief complaints of a non-healing wound over the dorsum of the left foot with a huge foot swelling of two and half years duration and difficulty in walking since three months. There was no history of trauma. His past history revealed the excision of the mass from the dorsum of the left foot one and half years back by a local practitioner. Since then, the mass had recurred after 6 months with nodular surface and skin ulceration, which slowly went on increasing. During the presentation to our hospital, the mass was found to have occupied the distal half of the foot. The local examination revealed a large, ulceroproliferative, nodular mass over the dorsum of the left foot, which totally measured 15x10 x9 cms, which involved the plantar aspect also. There were extensive deformities of all the toes and oedema on the distal one third of the leg and the foot. The left inguinal nodes were not palpable. The per abdominal examination revealed no abnormality. The X-ray of the chest and ultrasonogaphy of the abdomen showed no significant pathology. The X ray of the left foot revealed a soft tissue mass on the distal left foot with deformity of the bones. All the routine examinations were within normal limits. A small incisional biopsy was done, which was reported as suggestive of low grade fibromyxoid sarcoma. In view of its large size, the severe foot deformity, extensive surface ulceration and neurovascular defects, a below the knee amputation was done and the specimen was sent for histopathological examination. At present, the patient is on regular follow up.

Gross Features: We received the left below the knee amputation specimen with the large foot mass. The dorsal aspect of the foot showed a large, irregular, nodular, bosselated, grey white mass which extended from the lateral to the medial aspect, which involved the dorsum and the plantar aspect of the foot and which totally measured 15×10×9 cm [Table/Fig-1]. The lesion was composed of multiple, well circumscribed, nodular masses which extended all over the dorsum of the foot and those which extensively involved and deformed the dorsum of the foot. A large irregular surface ulcer was seen over the dorsum, which extended towards the great toe, which measured 5.2×4 cm. The surface of the ulcer was covered with necrotic slough. The cut surface of the mass was well circumscribed, with multiple, nodular grey white, fleshy areas with glistening surfaces [Table/Fig-2]. Multiple sections of it were taken for the histopathological diagnosis.

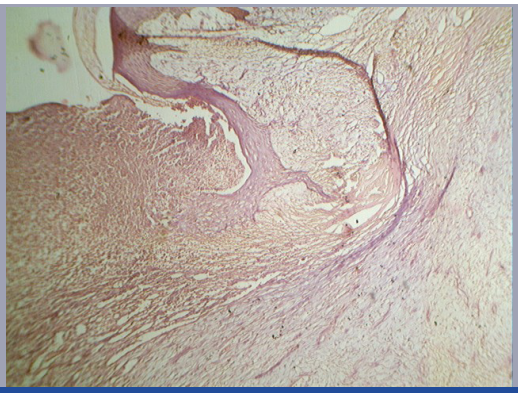
Light microscopy: The multiple sections showed a nodular tumour with sharp demarcation beneath the skin. The skin was thinned out and at places, it was ulcerated by the underlying tumour [Table/Fig-3]. The tumour was composed of moderate to low cellularity of a biphasic pattern, with alternate fibrous and myxoid areas. The tumour cells were arranged in fascicles and in swirling and whorled patterns. The individual tumour cells were bland and oval to spindle shaped, with small hyperchromatic, oval to tapering nuclei, which contained fine clumped chromatin and a pale, ill defined cytoplasm. The background matrix ranged from fibromyxoid to dense fibrous [Table/Fig-4 & 5]. At places, stellate cells were noted in the fibromyxoid areas [Table/Fig-6]. An occasional area of necrosis



[Table/Fig-1]: Gross specimen of left foot with tumor mass



[Table/Fig-2]: Cut surface of the mass showing multiple nodular gray white areas with glistening appearance



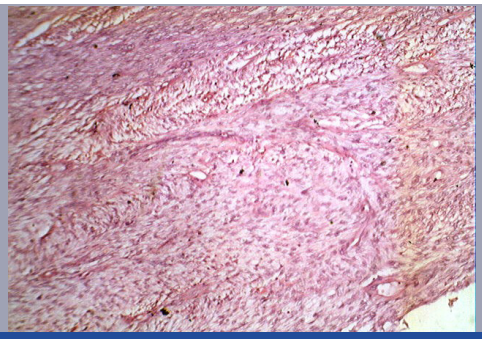
[Table/Fig-3]: Photomicrograph showing tumor composed of biphasic pattern of fibrous and myxoid areas with low to moderate cellularity with ulceration of overlying skin. (H&E stain, x100).

with a low mitotic activity was noted. Based on these features, the tumour was diagnosed as low grade fibromyxoid sarcoma. Immunohistochemistry was performed to confirm the diagnosis. The neoplasm stained strongly for vimentin and it stained negative for other markers like CD34, CD68, SM actin and S-100.

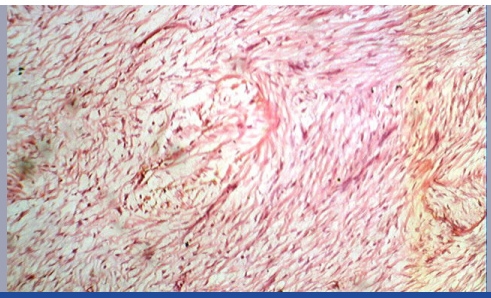
DISCUSSION

The Stanford School of Medicine defined low grade fibromyxoid sarcoma as a cytologically bland malignant neoplasm with alternate fibrous and myxoid stroma of a low grade/ low malignant potential [6].

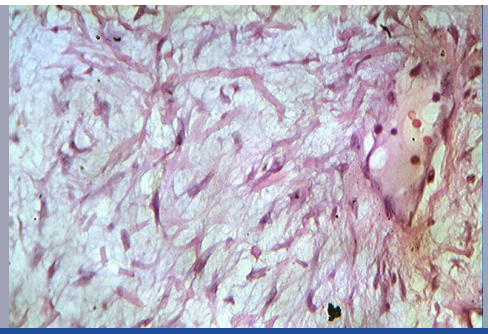
Low grade fibromyxoid sarcoma (LGFMS) was first described by Evans [1] as a slow growing, asymptomatic soft tissue tumour with apparently benign histological characteristics, which belied a high metastatic potential. The usual presentation of the tumour is



[Table/Fig-4]: Photomicrograph showing tumor cells arranged in fascicle, swirling and whorled patterns. (H&E stain, x100).



[Table/Fig-5]: Individual tumor cells are bland, oval to spindle shaped with small hyperchromatic, oval to tapering nuclei with pale ill defined cytoplasm. (H&Estain, x100).



[Table/Fig-6]: Photomicrograph showing few stellate cells in fibromyxoid background (H&E stain,x400).

a slow growing, painless, deep soft tissue mass that ranges from 1-18cm in greatest diameter, although most are about 8-10 cm [7]. The tumour most commonly arises in the deep soft tissues of the lower extremities, particularly the thigh. The following regions can be affected in the decreasing order of frequency: The chest wall/ axilla, shoulder, inguinal, buttock and the neck [5,8].

In recent years, several new entities of LGFMS have been described, which are: [2,5]

1. Low grade myxofibrosarcoma
2. Low grade fibromyxoid sarcoma
3. Hyalinising spindle cell tumour with giant collagen rosette (HST)- Evans tumour
4. Sclerosing epitheliod fibrosarcoma

Myxofibrosarcoma may show a progression to high grade sarcoma, but the remaining three types almost always remain as low grade fibrosarcoma [2,5].

Evans [1] and Goodlad et al [4] suggested that LGFMS were paradoxically aggressive tumours. In the retrospective early series, local recurrence was noted in 68%, metastasis in 41% and death from the disease in 18% of the tumours. So, Evans HL [9], in 1993, labeled LGFMS as a distinctive soft tissue sarcoma with the most

common affected location as the thigh, with predominance of local recurrence and distant metastasis to the lung according to his report on 12 cases.

We are reporting here, a case of a 70-year old male who presented with LGFMS as multiple local recurrence, foot deformities, surface ulcerations and pressure effects. Clinically, an extensive foot deformity was noted, which led to an inability to walk and to do the routine daily activities. In view of the above features, a below the knee amputation was done and the patient was advised regular follow up.

Regarding the differential diagnosis of LGFMS, it involves numerous benign and malignant soft tissue lesions which are characterized by fibrous and myxoid stroma, such as desmoids fibromatosis, perineuroma, myxoid neurofibroma, nodular fasciitis, low grade myxofibrosarcoma, myxoid liposarcoma and malignant peripheral nerve sheath tumour [6]. These lesions should be differentiated because their clinical behaviour and prognosis differs.

Desmoids fibromatosis and low grade myxofibrosarcoma are the most important differential diagnoses which should be discussed, as per the Stanford School of Medicine [6].

In extraintestinal desmoids fibromatosis–sweeping bundles of cells are seen instead of the whorled or swirling patterns. The nuclei are vesicular, with no rosettes. Frequent slit like vessels are noted.

Another differential diagnosis is low grade myxofibrosarcoma which is characterized by a lack of fibrous and myxoid alternate areas, with the presence of pleomorphic nuclei and an abnormal mitotic activity. These lesions are often subcutaneous and cellular. These features are lacking in LGFMS [6].

The chromosomal translocation, t (7;16) (q33;p11) is identified in up to 96% of the LGFMS cases. This genetic abnormality is not present in other myxoid or fibrous neoplasms [6].

CONCLUSION

Low grade fibromyxoid sarcoma is a rare entity with cytologically bland, malignant neoplasm and an aggressive nature. So, LGFMS should be considered in the differential diagnosis of spindle cell neoplasms while the histopathology of soft tissue tumours are studied. A regular follow up for local recurrence as paradoxically aggressive behaviour of tumour will be helpful for patient care.

We are presenting this case of LGFMS due to its unusual and unique presentation as local recurrence and an extensive foot deformity.

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