Myxofibrosarcoma – An enigma in diagnosis: Case report

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ABSTRACT

Myxofibrosarcoma is a distinct malignant neoplasm of fibroblastic origin with variable clinical and histopathologic features. It is common in elderly, and up to two thirds present as dermal tumors, frequently with infiltrative borders making complete resection difficult. We present here a case, which initially on clinical and radiological grounds appeared as a benign hemangioma, but turned out to be myxofibrosarcoma on histopathological examination, to highlight the diagnostic difficulties it may present. This patient was treated according to standard protocol including radiation therapy and followed for 1 year without any evidence of local recurrence or metastasis.

Key words: myxofibrosarcoma, sarcoma, hemangioma

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INTRODUCTION

Myxofibrosarcoma is one of the common histologic types of malignant soft tissue sarcoma presenting usually in adult life, peaking at seventh decade, and frequently involving the extremities, trunk, retroperitoneum, mediastinum and head and neck region [1, 2]. We present this case which clinico-radiologically disguised as a benign hemangioma, but on histopathological examination was diagnosed as Myxofibrosarcoma.

CASE REPORT

A 65-year-old Indian female presented with a large swelling measuring 15 cm × 8 cm × 4 cm over the anterolateral aspect of right leg of 8 months duration (Fig.1). The lesion was painful, and patient reported an increase in size during the last 6 months. There were no similar swellings in other parts of the body or any constitutional symptoms. Systemic examination was normal and there was no significant lymphadenopathy or distal neurovascular deficits.
Plain radiographs were normal except for the soft tissue shadow. The magnetic resonance imaging showed a well-defined lobulated soft tissue lesion of 17.6 x 6.6 x 2.8 cm. It was predominantly hyperintense in T2 and hypointense in T1 with multiple hypointense septae.

Lesion showed calcifications and there was intense post contrast enhancement. However, there was no evidence of extension into muscle or bones (Fig.2). These features were suggestive of vascular malformation like hemangioma. The doppler study done to supplement our diagnosis also showed solid components with arterial (predominantly) and venous waveform, pointing towards hemangioma.

Her haematological parameter including ESR was within normal range. Clinical and radiological investigations revealed no evidence of distant metastasis. Excision biopsy was done. The tumor was seen in the subcutaneous plane on the anterior aspect of proximal and middle third of right leg (Fig.3).

On microscopy, the tissue showed a relatively hypocellular tumor with vaguely nodular contour composed of myxoid material and scattered spindle cells with hyperchromatic nucleus. Moderate degree of pleomorphism, few binucleate cells, slender capillary channels with lymphoid aggregates and areas of haemorrhage, bands of dense fibrocollagenous tissue infiltrated by occasional atypical mitotic figures and focal areas of hyalinisation was evident (Fig.5). These features were suggestive of myxofibrosarcoma.

Post operatively the patient was given adjuvant radiation therapy. The postoperative External beam radiation therapy (EBRT) was given at a dose of 63 Gy at 1.8Gy/fraction. The initial target volume included the tumor bed plus 5–10 cm margins to a dose of 45Gy. This was followed by two cone downs to bring the median total dose to 63 Gy. The patient was followed for 1 year with clinical examination, serial radiographs and ultrasound abdomen. No evidence of local recurrence or distant metastasis was noted in our case.

Fig : 1- Clinical Picture

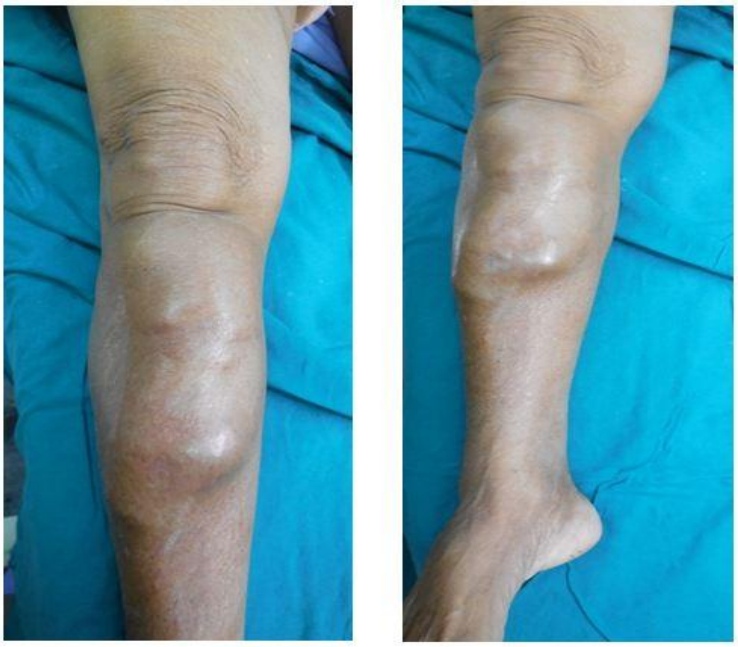

Fig.1- Clinical picture

Fig: 2- MRI showing tumor in axial, coronal and sagittal sections

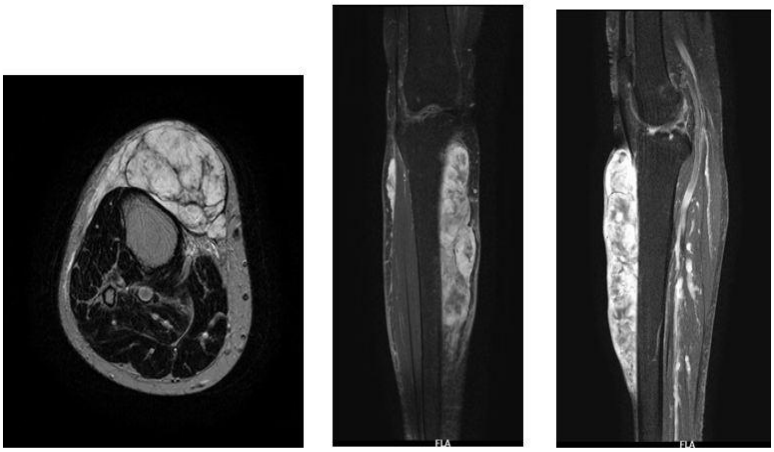

Fig.2- MRI showing tumor in axial, coronal and sagital section

Fig: 3- Intraoperative picture showing tumor tissue

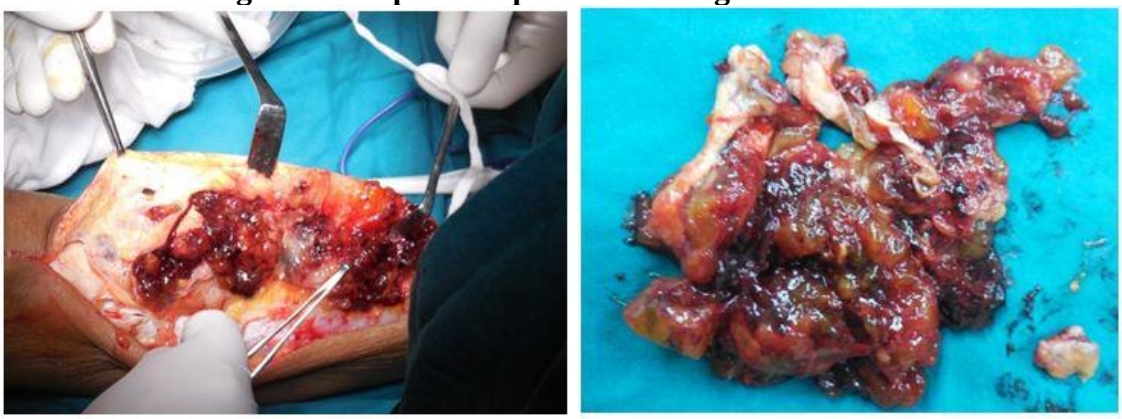

Fig.3- Intraoperative picture showing tumor

Fig: 4- Gross appearance of tumor

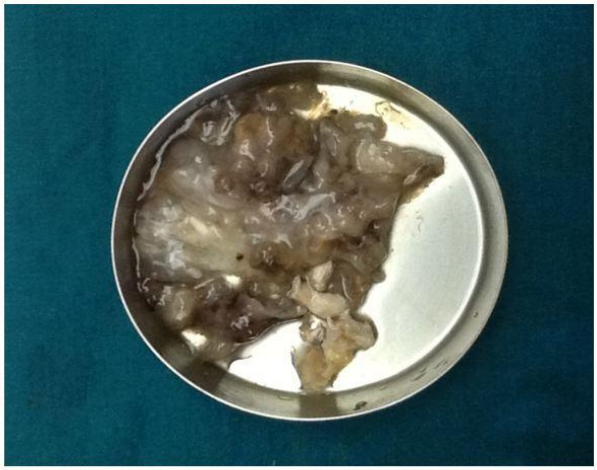

Fig.4- Gross appearance of tumor

Fig: 5- Histopathological appearance of Myxofibrosarcoma- Low power and high power

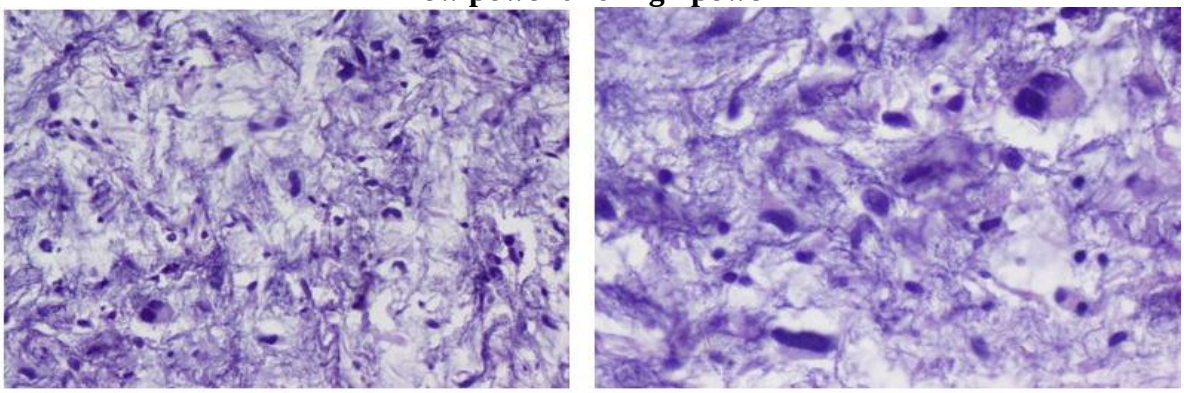

Fig.5-Histo-pathological appearance of myxofibrosarcoma- low power and high power

DISCUSSION

Myxofibrosarcoma refers to a spectrum of malignant myxoid tumors of fibroblastic origin that exhibit varied clinical and histo-pathologic features [3]. They are most commonly seen in older adults, and two thirds of these cases develop within the dermis or subcutis which has brought interest from dermatologists and dermatopathologists[4-6].

There is no significant gender predilection and tend to affect patients in the sixth to eighth decades [7]. However, the overall age range is wide, and cases have been reported in patients as young as 15 years [8].

Myxofibrosarcomas are encountered most frequently on the extremities and limb girdles, but may also arise within the head and neck region, the trunk, mediastinum and retroperitoneum [9]. In our case also tumor was located over the extremity.

A gradually enlarging painless dermal or subcutaneous mass is the most usual clinical presentation [7]. Sub optimal resection often leads to local recurrence, and recurrent low-grade lesions may undergo progression to more malignant variant, a transition accompanied by increasing metastatic potential [7].

The chances of recurrence does not appear to be related to tumor depth, but the risk of metastasis and tumor-related death is much greater in deep-seated lesions and those with high-grade varieties [1,10]. Mentzel et al in their study of 75 cases of myxofibrosarcoma, reported metastasis in significant number of high-grade tumors [1]. Lungs and bones were the most common metastatic sites, but spread to regional lymph nodes may also occur in some individuals [1,11]. Fortunately our case did not have any evidence of recurrence nor distant metastasis.

The overall 5-year survival rate is about 60–70% [1]. Incidentally, previous studies show that superficial portions of myxofibrosarcomas are morphologically low grade, regardless of the

grade seen within the remainder of the tumor [4]. Furthermore, they may be deceptively infiltrative, making clinical assessment of margins difficult [1]. This further leads to enigma in the diagnosis of this condition in these individuals.

CONCLUSION

Myxofibrosarcoma is a malignant lesion and can behave deceptively due to their varied clinical presentation. The radiographic evidence may not correlate with the tumor. A high index of suspicion is needed in elderly individuals with superficial dermal infiltrative lesion. Complete treatment with excision and standard radiotherapy protocol is absolutely essential, as local recurrence and metastasis are high.

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