



DECEPTIVELY BENIGN IMAGING OF MULTICENTRIC LOW-GRADE MYXOFIBROSARCOMA: A CASE OF ATYPICAL RECURRENCE

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ABSTRACT

Myxofibrosarcoma (MFS) is a common fibroblastic sarcoma in elderly patients, typically presenting in the sixth decade with no sex predilection, most frequently involving the extremities. We report a rare case of a 30-year-old male with a progressively enlarging left thigh mass, histologically diagnosed as low-grade MFS. Following surgical excision and adjuvant radiotherapy, the patient developed recurrent masses in the neck, chest, and abdomen within a year. Imaging findings on MRI and PET-CT suggested recurrence, which was histopathologically confirmed. This case highlights the deceptive radiologic appearance and unusual multicentric recurrence pattern of low-grade MFS, underscoring the importance of thorough follow-up and multidisciplinary management.

KEYWORDS : Myxofibrosarcoma, Soft Tissue Sarcoma, MRI, Recurrence

INTRODUCTION

Myxofibrosarcoma (MFS), previously considered a myxoid variant of malignant fibrous histiocytoma, is now recognized as a distinct soft tissue sarcoma entity [1,11]. The term "myxofibrosarcoma" was introduced by Scandinavian pathologists to emphasize its myxoid matrix and fibroblastic differentiation [1,11]. It typically affects elderly patients and arises in the subcutaneous tissues of the extremities; however, deep-seated and multicentric manifestations are rare [2,3]. Unlike other well-circumscribed soft tissue sarcomas, MFS demonstrates ill-defined, infiltrative margins and spreads centrifugally along fascial and vascular planes, contributing to incomplete resections and high recurrence rates [3,12,13]. Distant metastasis is uncommon in low-grade MFS, but transformation to higher-grade disease increases the risk [3,4,5,11].

Case Report

A 30-year-old male with no comorbidities presented with a gradually enlarging mass on the posteromedial aspect of the left thigh, present for two years. Clinical examination revealed a firm, non-tender 30×20 cm mass. MRI showed a large, encapsulated multilobulated soft tissue mass in the adductor compartment with heterogeneous peripheral contrast enhancement and no necrosis or cystic change. There was no evidence of neurovascular invasion. Histopathology confirmed low-grade MFS (FNCLCC Grade 1) with indeterminate margins. The patient received adjuvant radiotherapy (66 Gy in 33 fractions) using VMAT, IGRT, and SGRT techniques.

At 4-month follow-up, MRI revealed post-treatment muscle volume loss and fibrotic changes, with no evidence of local recurrence. However, one-year post-treatment, the patient presented with right supraclavicular swelling and abdominal discomfort. CT imaging identified soft tissue lesions in the neck, lungs, and abdomen. MRI of the neck showed a T1 isointense and T2 hyperintense supraclavicular lesion with heterogeneous post-contrast enhancement. PET-CT revealed metabolically active lesions in the neck, chest, and abdomen. Ultrasound-guided biopsy of the abdominal mass confirmed recurrent low-grade MFS.

Myxofibrosarcoma commonly arises in the extremities and presents as a slow-growing subcutaneous mass [3,12,13]. Grossly, tumors may appear multinodular with fibrous septa and show hemorrhagic or necrotic areas. Infiltrative low-

grade lesions can lack clear nodularity, extending along fascial planes [3,12,13]. Despite low histologic grade, local recurrence exceeds 50%, independent of depth or grade [3,11,13]. Recurrence may lead to dedifferentiation and increased metastatic potential [3,4,5].

MRI is the preferred imaging modality. MFS typically shows heterogeneous T1 and T2 signals, T2 hyperintensity, and variable post-contrast enhancement. T1 isointense or mildly hyperintense areas may be seen. The characteristic 'tail sign'—a T2 hyperintense, contrast-enhancing fascial extension—can help differentiate true infiltration from peritumoral edema [3,5].

Differential diagnosis includes myxoid liposarcoma, myxoinflammatory fibroblastic sarcoma, undifferentiated pleomorphic sarcoma, and nodular fasciitis. The 'tail sign' has moderate sensitivity and specificity and may also be seen in mimics [6,7].

Waters et al. documented atypical metastatic sites including pleura, adrenal glands, mesentery, bone, and soft tissue [7]. In a study of 158 patients, the 5-year survival rate was approximately 77%, higher than for undifferentiated pleomorphic sarcoma (50–60%) [9,10].

Given the atypical metastatic pattern, comprehensive imaging beyond the thorax is warranted. MRI is crucial for preoperative planning and surveillance [8].



Figure 1: Right supraclavicular swelling

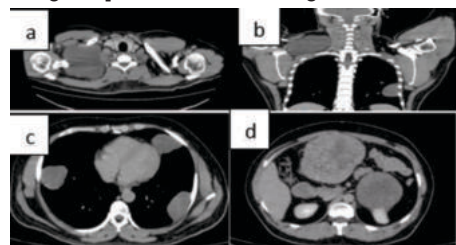
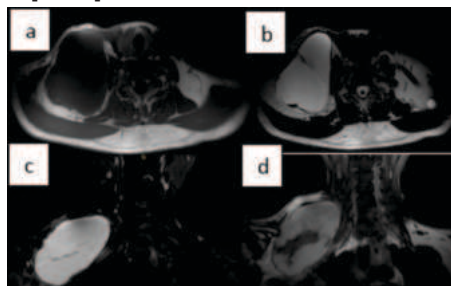


Figure 2: CT chest and abdomen

2a: Large well defined soft tissue density lesion in right supraclavicular region,

2b,2c: Multiple variable sized pleural based and parenchymal hypodense mildly enhancing well defined soft tissue lesion with no calcification are seen in both the lung fields

2d: Three Well defined, hypodense soft tissue lesion with mild heterogenous post contrast enhancement in right hypochondriac region, along second part of duodenum and left perinephric space.



3a: T1w NFS axial image shows well defined homogeneously isointense lesion in left supraclavicular region

3b, 3c: T2w NFS axial and STIR coronal image predominantly hyperintense lesion with few hypointense areas

3d: Post contrast T1 post contrast FS shows heterogenous post contrast enhancement with non-enhancing areas

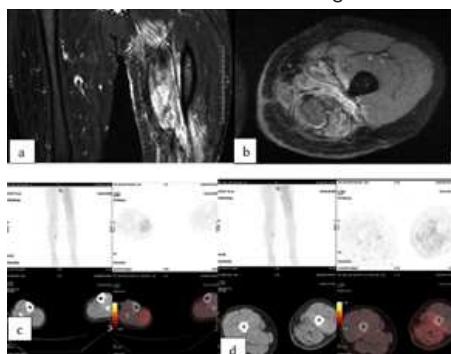


Figure 4: MRI left leg

4a. STIR coronal image shows heterogeneously hyperintense signal in posterior compartment

4b: T1 post contrast axial image shows heterogenous post contrast enhancement in posterior compartment

4c, 4d: PET-CT - Low grade metabolic activity in right gastrosoleus muscle and left gluteal muscle, Ill-defined low grade FDG avid collection seen in intermuscular plane of visualized left thigh along posterior compartment with diffuse fat stranding-likely post-operative changes.

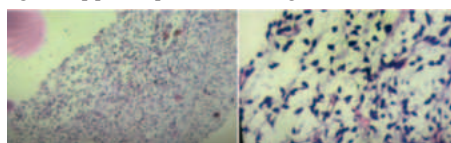
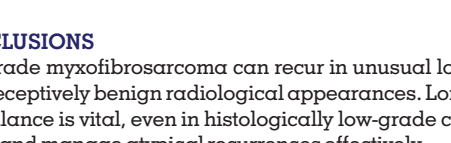


Figure 5: HPE of abdominal lump biopsy (HP No. 6937) demonstrates Low grade myxofibrosarcoma with fibrous part (collagenous rosette) and Myxoid parts with classical whirling spindle cells and curvilinear thin vessels.



CONCLUSIONS

Low-grade myxofibrosarcoma can recur in unusual locations with deceptively benign radiological appearances. Long-term surveillance is vital, even in histologically low-grade cases, to detect and manage atypical recurrences effectively.

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