Case Report

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Pathology: Myxofibrosarcoma - An Unusual Presentation in a Young Patient

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Abstract

Myxofibrosarcoma, a low-grade malignant mesenchymal tumor is the myxoid variant of malignant fibrous histiocytoma. It is most commonly seen in the extremities. A primary myxofibrosarcoma is extremely uncommon in young adults. We report a rare case of a 35-year-old female patient with a progressively enlarging soft tissue mass in the gluteal region in whom pathological examination of the resected mass revealed the classic diagnostic features of a low-grade myxofibrosarcoma. The subsequent clinico-radiological examination did not reveal evidence of malignancy in any other location. This case is reported for its rarity of occurrence in the young patient.

Key words: Low-grade, Malignant fibrous histiocytoma, Mesenchymal tumor, Myxofibrosarcoma, Myxoid

INTRODUCTION

Myxofibrosarcoma is a myxoid variant of malignant fibrous histiocytoma (MFH) with characteristic morphological appearance inclusive of nodularity, prominent myxoid matrix, and elongated curvilinear capillaries in the extremities of the elderly.[1-3] The tumor has been categorized into low, intermediate, and high-grade categories based on the degree of cytological atypia and the presence or absence of pleomorphic MFH-like pattern within the tumor by Mentzel *et al.*[3] In their study, low-grade tumors showed mainly myxoid areas with mild cytological atypia in the cellular areas while the high-grade tumors showed a pleomorphic appearance with a high mitotic activity, multinucleated tumor giant cells, and areas of necrosis.[3] Several studies have shown that approximately 80% of these tumors occur in the extremities.[1-3] The peak incidence was observed in the 5th-7th decades with a slight female preponderance.[2,4] We report a soft tissue low-grade myxofibrosarcoma in the gluteal region of a 35-year-old female patient in view of its rarity of presentation in the young patient.

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CASE REPORT

The 35-year-old female patient presented with a painless, progressively enlarging mass in the left gluteal region. Clinical and radiological examination did not reveal any bony involvement. A provisional clinical diagnosis of a soft tissue sarcoma was made, and a wide excision of the mass was performed. The excised mass was submitted for histopathological examination.

Gross Examination Findings

Irregular, unencapsulated, poorly circumscribed, gray-white, nodular mass measuring 9 cm × 6 cm × 3 cm (Figure 1). Cut section - nodular, gray-white with multiple myxoid areas (Figure 2).

Microscopy

Showed the mesenchymal tumor composed of multiple nodules of the spindle to stellate tumor cells amidst prominent myxoid matrix (Figure 3). Individual tumor cells showed scant cytoplasm, irregular, large, and pleomorphic nuclei with prominent nucleoli. Some bi and multinucleated tumor giant cells were also seen. Occasional atypical mitoses were seen. Many curvilinear capillaries with tumor cells aligned around them were seen (Figure 4).

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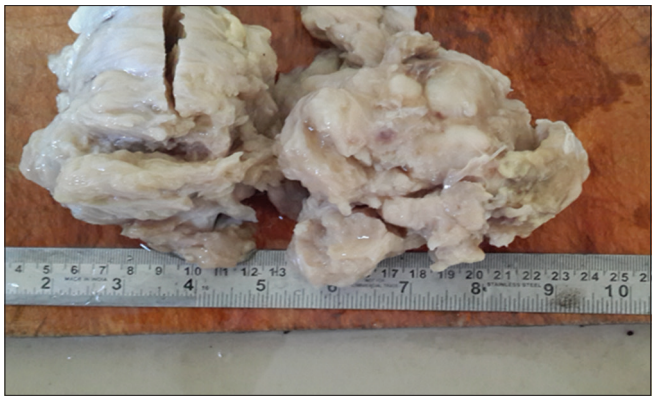

Figure 1: Irregular, unencapsulated, poorly circumscribed, gray-white, nodular mass measuring 9 cm × 6 cm × 3 cm

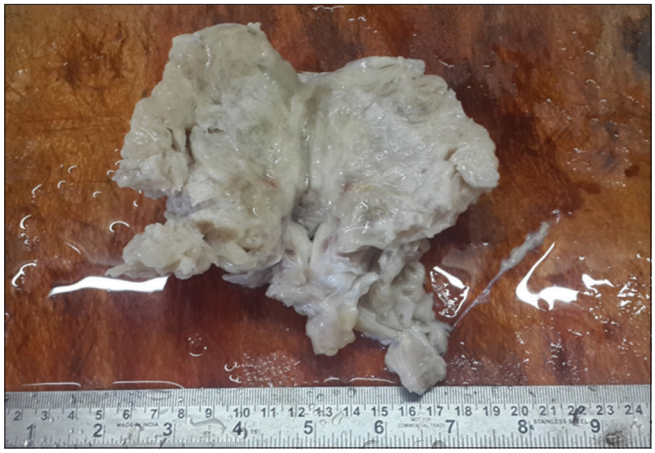

Figure 2: Cut section - nodular gray-white with multiple myxoid areas

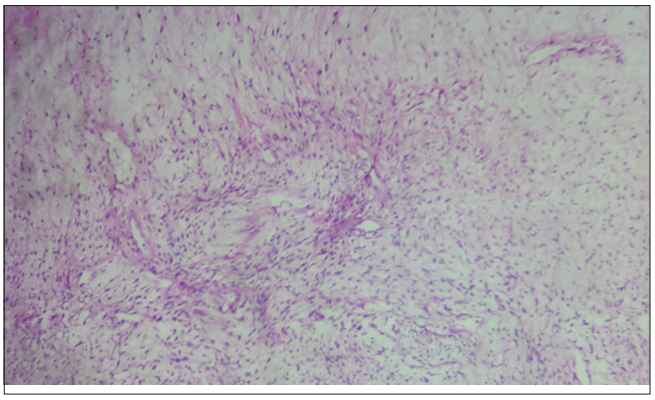

Figure 3: Mesenchymal tumor composed of multiple nodules of spindle to stellate tumor cell amidst prominent myxoid matrix (H and E - ×10)

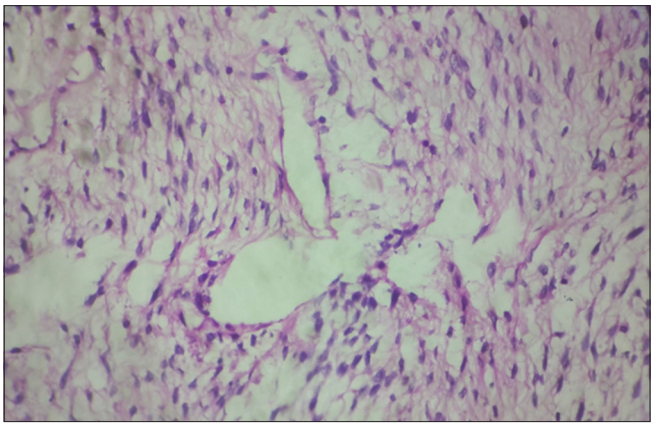

Figure 4: Many curvilinear capillaries with tumor cells aligned around them (H and E - ×45)

Final Diagnosis

Myxofibrosarcoma (low-grade myxoid MFH).

DISCUSSION

Myxofibrosarcoma is one of the frequently encountered soft tissue malignancies in elderly patients mainly seen in the extremities.[5] It belongs to the heterogeneous group of fibrohistiocytic tumors.[3,6-8] It is one of the most aggressive types of soft tissue malignancies with a high risk for local recurrence and a significantly high metastatic rate.[3] The clinical presentation is not pathognomonic thus delaying the diagnosis.

Histologically, myxofibrosarcomas show a spectrum ranging from hypocellular tumors consisting of widely spaced spindle cells amidst myxoid matrix to more cellular variants with pleomorphic nuclei and pseudo lipoblasts.[5]

The presence of curvilinear blood vessels is a consistent morphological feature. Enzinger and Weiss have classified this tumor as the myxoid variant of MFH.[2] Myxofibrosarcoma was first recognized as a distinct entity by Angervall *et al*.[1] It has been accepted as a separate entity and included in the 2002 WHO classification of tumors.[5] The prevalence of myxoid areas defines the lesion of the grade being more prominent in low-grade lesions. Grade II and III tumors have a metastatic potential and are hence recognized as truly malignant in contrast to the Grade I tumor which is just locally aggressive.[3] Metastasis usually occurs in patients with intermittent and high-grade myxofibrosarcoma.[3] However, myxoid fibrosarcoma, irrespective of the grade warrants closes surveillance.

The differential diagnoses of myxofibrosarcoma include all the other myxoid tumors. It is often difficult to distinguish them from each other since the differences are very subtle.[9] Wide resection is the treatment of choice.[3,10] Insufficient tumor-free margins make way for local recurrence and worse prognosis. Due to the high recurrence and metastatic rate in high-grade tumors, adjuvant radiotherapy is recommended. The size of the tumor (smaller than 5 cm versus >5 cm), anatomical location (distally versus proximally located tumors), and histological features (including the degree

of anaplasia and the number of mitoses) also correlate with prognosis.[3,10] A thorough follow-up is advocated to monitor eventual local recurrence.

CONCLUSION

Myxofibrosarcoma may present a diagnostic challenge to the pathologist since it is a close mimic of many other myxoid mesenchymal malignancies, and hence, caution needs to be exercised. Extensive surgical excision with adjuvant radiotherapy is the optimal therapeutic choice.

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