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Unmasking The Impostor: Myxoid Liposarcoma Of The Arm Masquerading As A Spindle Cell Lesion – A Rare Case Report

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ABSTRACT:
BACKGROUND: Myxoid liposarcoma (MLPS) is a malignant tumour composed of uniform, round to ovoid cells with variable numbers of small lipoblasts, set in a myxoid stroma with arborizing vasculature. MLPS typically present within deep soft tissues of the extremities, often thigh and rarely from the upper extremities, subcutis or retroperitoneum. MLPS accounts 20-30% of liposarcoma and 5% of adult soft tissue sarcomas with a peak incidence in the fourth to fifth decades.
CASE REPORT: We present a case of 23 years old female with sudden onset painful right arm swelling. MRI right arm was suggestive of vascular lesion while MRI with contrast suggested it was a neurogenic tumor. Cytology findings suggested a mesenchymal lesion, with uncertain malignant potential. The histopathological and immunohistochemical findings were suggestive of Adipocytic tumor - Likely Myxoid Liposarcoma (FNCLCC Grade 2).
CONCLUSION: The case highlights the importance of including myxoid liposarcoma (MLPS) in the differential diagnosis of spindle cell lesions, especially when imaging and cytological findings are ambiguous. A multidisciplinary approach is essential for accurate diagnosis. Early detection of MLPS enables prompt treatment, ultimately improving patient prognosis and outcomes.

KEYWORDS: Myxoid liposarcoma, liposarcoma, lipoblasts, spindle cell neoplasm.

INTRODUCTION

Liposarcoma is the most common soft tissue sarcoma of adult life with an incidence of 20%.[1] WHO divides it into four subtypes: atypical lipomatous tumour/well-differentiated liposarcoma (ALT/WDL), dedifferentiated liposarcoma, myxoid/round cell liposarcoma (MRCLPS), and pleomorphic liposarcoma Myxoid liposarcoma (MLPS) is a rare and aggressive soft tissue sarcoma accounting for approximately 30-35% of all liposarcomas [1]. It typically presents in the deep soft tissues of the extremities, especially the thigh, though uncommon sites like the trunk, retroperitoneum, and pericardium have also been reported [2]. MLPS is characterized by a gelatinous, myxoid stroma containing pleomorphic lipoblasts and arborizing vasculature, often mimicking other spindle cell lesions. Such overlapping features frequently pose diagnostic challenges, particularly in cytology and imaging studies.
Early diagnosis significantly improves prognosis. MLPS demonstrates variable behaviour ranging from indolent tumors to aggressive malignancies with metastatic potential. This case report highlights an unusual and rare presentation of MLPS in the arm (with an incidence of <1%) [3] that was initially misdiagnosed as a spindle cell lesion, emphasizing the need for a comprehensive diagnostic approach.

CASE REPORT

A 23-year-old female presented with a gradually increasing painful swelling in the right arm measuring 5x5 centimetres. Physical examination revealed a firm, mobile, and tender mass measuring 5x5 cm.

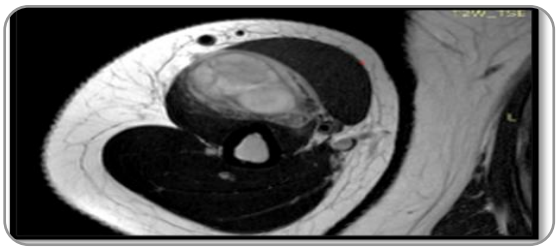

Figure1: Heterogeneously T2 hyper intense lesion on MRI

Imaging Findings: MRI of the right arm suggested a vascular lesion, while MRI with contrast indicated features consistent with a neurogenic tumor. Imaging showed a well-defined, heterogeneously T2 enhancing lesion (Figure 1) without surrounding tissue infiltration.

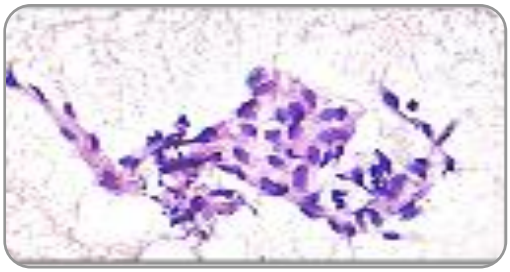

Figure 2: MGG stain 400X

Cytological Findings: FNAC revealed loosely cohesive clusters of spindle cells with plump, ovoid nuclei and occasional bizarre cells dispersed in a myxoid background raising a suspicion of a mesenchymal lesion with uncertain malignant potential. Figure 2.

Histological Findings:
Incisional Biopsy: Two linear cores revealed a well-differentiated adipocytic neoplasm with fibrous bands showing atypical spindle cells having hyperchromatic nuclei, nuclear pleomorphism, and bizarre forms admixed with adipocytes and lipoblasts of varying sizes. The provisional diagnosis was a malignant mesenchymal tumor – Atypical Lipomatous Tumour/Well Differentiated Liposarcoma (ALT/WDL). Figure 3.

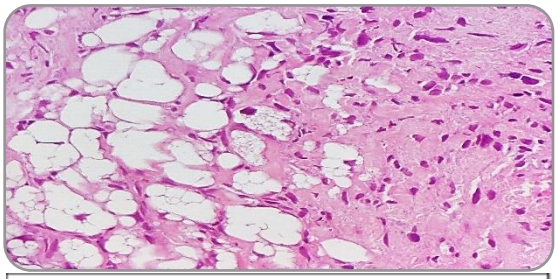

Figure3: H&E stain 100X; lipoblasts

Excision Biopsy: Sections from a well-encapsulated globular tissue mass showed predominantly spindled tumor cells with pleomorphic and hyperchromatic nuclei, bizarre multinucleated tumor cells, and abundant lipoblasts. The stroma displayed widespread myxoid degeneration and delicate arborizing vasculature. Mitosis was 2-3/hpf. Surgical margins were uninvolved by tumor cells. Figure 4

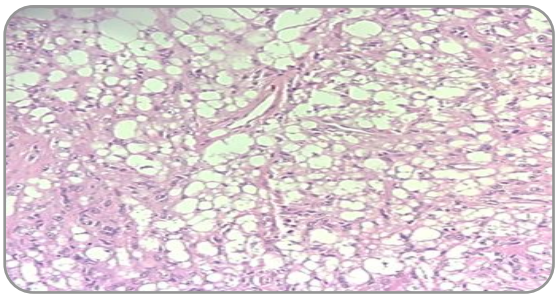

Figure 4: H&E 100X; tumour cells, arborizing vasculature

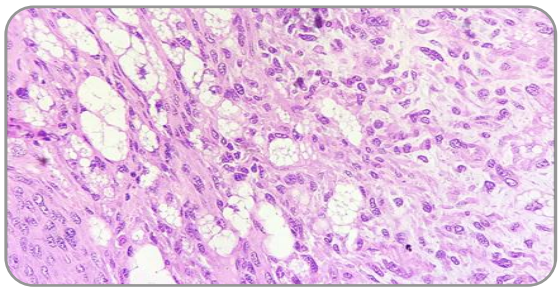

Figure 5: H&E 400X; Lipoblasts and spindle cells

Immunohistochemistry (IHC): Vimentin (Figure5) and p16 were positive, while S100, SMA, and Desmin were negative. These findings confirmed the diagnosis of Myxoid Liposarcoma, classified as FNCLCC Grade 1.

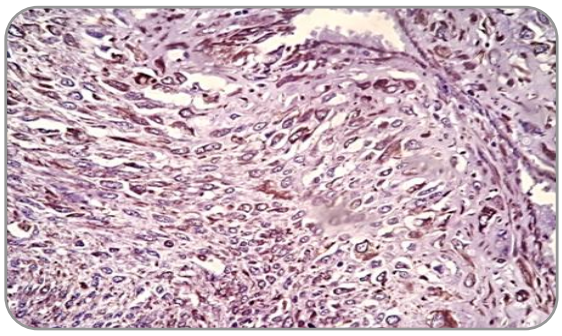

Figure 6: IHC 400X ; Vimentin positive, cytoplasmic staining

DISCUSSION

Myxoid liposarcoma is the second most common liposarcoma subtype in children and adolescents but peaks in incidence during the fourth to fifth decades with no sex predilection [1]. MLPS is known for its unique metastatic pattern, often targeting uncommon locations such as the trunk, extremities, bone, retroperitoneum, chest wall, pleura, and pericardium [1]. This unusual metastatic profile is associated with a poorer prognosis compared to other liposarcoma subtypes.

MLPS is characterized by the molecular hallmark recurrent translocation t (12; 16) or rarely t (12; 22) on the 12q13.3 locus resulting in FUS-DDIT3 and EWSR1-DDIT3 gene fusion transcripts, respectively[4].

MLPS must be distinguished from ALT/WDL which shares similar characteristics but is marked by the presence of more mature adipocytes, giant cells, and rare mitotic activity. Additionally, ALT/WDL lacks arborizing vasculature and typically exhibits giant/ring chromosomes with amplified regions at 12q, resulting in MDM2/CDK4 positivity on molecular testing.[5,6]

Histological grading of MLPS is crucial for predicting clinical behavior and guiding treatment decisions. FNCLCC Grade 1 MLPS is typically associated with a more favorable prognosis, while Grade 2 and 3 tumors demonstrate greater metastatic potential and worse outcomes [1].

Surgical excision with clear margins is the primary treatment modality for localized MLPS. In cases of high-grade MLPS or tumors with positive margins, adjuvant radiotherapy is recommended to reduce local recurrence rates. Chemotherapy, particularly anthracycline-based regimens, may be considered in metastatic or unresectable cases.

MLPS exhibits a variable prognosis, with five-year survival rates ranging from 70-90% in low-grade tumors but significantly lower in high-grade or metastatic cases.[7]

CONCLUSION

This case underscores the importance of considering myxoid liposarcoma in the differential diagnosis of spindle cell lesions, particularly when imaging and cytological findings are inconclusive. Accurate diagnosis is crucial for effective management and treatment. A multidisciplinary approach, integrating clinical, radiological, cytological, and histopathological data, is essential for timely and appropriate intervention in cases of soft tissue masses.

CONFLICT OF INTEREST: None

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ABBREVATIONS:

MLPS: Myxoid liposarcoma
MRI: Magnetic Resonance Imaging
MGG: May-Grünwald-Giemsa
ALT/WDL: Atypical lipomatous tumour/well-differentiated liposarcoma
MRCLPS: Myxoid/round cell liposarcoma
FNCLCC: Fédération Nationale des Centres de Lutte Contre le Cancer
WHO: World Health organization.
FUS: Fused in Sarcoma
DDIT3: DNA damage-inducible transcript 3
EWSR1: Ewing sarcoma breakpoint region 1 or EWS RNA-binding protein 1