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Case Report

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Dedifferentiated Liposarcoma with Meningothelial-Like Whorls: A Diagnostic Challenge on Biopsy

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ABSTRACT

Dedifferentiated liposarcoma (DDL) is a malignant adipocytic tumor characterized by the juxtaposition of a well-differentiated liposarcoma and a non-lipogenic sarcomatous component. The presence of meningothelial-like whorls is a rare histological pattern that may complicate diagnosis, especially on small biopsies.

Case Presentation: We report a case of a 60-year-old man presenting with a 5 cm inguinal mass. Histological examination of a core biopsy showed a spindle cell neoplasm arranged in meningothelial-like whorls. Immunohistochemistry revealed positivity for smooth muscle actin (SMA), p16, CD10, and CD99, with a Ki-67 proliferative index of approximately 25%. Molecular analysis confirmed MDM2 gene amplification. No well-differentiated liposarcoma component was identified. The diagnosis of dedifferentiated liposarcoma with meningothelial-like whorls was established based on the combined histologic, immunohistochemical, and molecular findings.

Conclusion: This case highlights the diagnostic challenge of identifying DDL on biopsy specimens lacking a differentiated lipogenic component. The meningothelial-like whorl pattern may mimic other spindle cell tumors. Demonstration of MDM2 amplification is essential for accurate diagnosis.

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Introduction

Dedifferentiated liposarcoma (DDL) represents a malignant progression from a well-differentiated liposarcoma (WDL) to a non-lipogenic sarcomatous component. It commonly arises in the retroperitoneum but may also occur in peripheral soft tissue sites, including the inguinal region.

The presence of a meningothelial-like whorl pattern is an uncommon histological feature that can lead to diagnostic confusion with other soft tissue sarcomas such as pleomorphic sarcoma, myxofibrosarcoma, or solitary fibrous tumor. Accurate diagnosis requires careful correlation between morphology, immunohistochemistry (IHC), and molecular studies-especially MDM2 amplification, which serves as a hallmark of DDL.

We report a challenging case of DDL diagnosed on biopsy, in which the absence of a well-differentiated lipogenic component made molecular confirmation indispensable.

Case Presentation

A 60-year-old man presented with a painless, slowly enlarging inguinal mass measuring 5 cm in greatest diameter. There were no systemic symptoms. Ultrasound and MRI revealed a well-circumscribed, heterogeneous lesion located in the inguinal soft tissue. An ultrasound-guided core biopsy was performed.

Histopathological Findings

Hematoxylin and Eosin (H&E) Microscopy

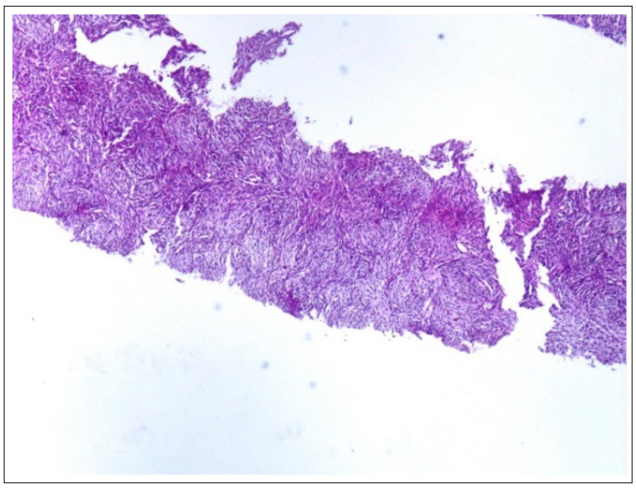

Figure 1 (H&E ×5): Low-Power Examination Revealed a Moderately Cellular Spindle cell Proliferation Arranged in Intersecting Fascicles and Forming Whorled, Meningothelial-like Structures

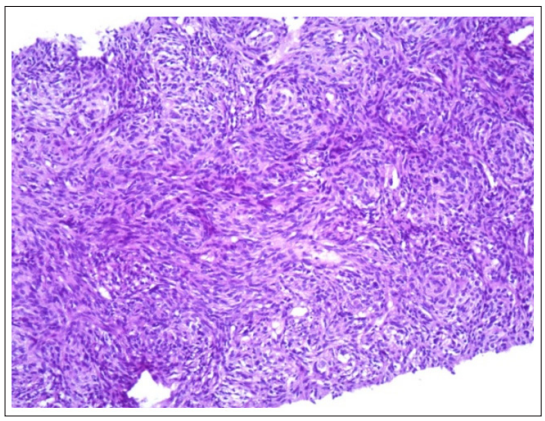

Figure 2 (H&E ×10): Tumor Cells Appeared Cohesive with Focal Concentric Arrangements Around Small Vascular Spaces. No Adipocytic Differentiation was Identified

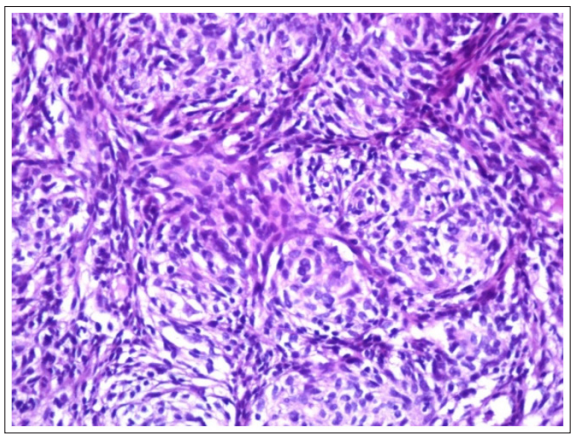

Figure 3 (H&E ×25): The Cells Displayed Oval to Spindle-Shaped Nuclei with Finely Dispersed Chromatin and Small Nucleoli. Cytoplasm was Eosinophilic, and Cell Borders were Indistinct

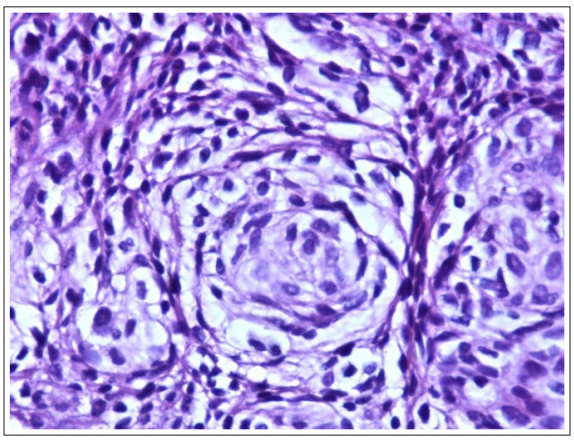

Figure 4 (H&E ×40): At high Magnification, Nuclear Atypia was Mild to Moderate, With Occasional Mitoses but no Necrosis. The Background Showed Delicate Collagen Bundles and Focal Myxoid Areas

No well-differentiated liposarcoma component was seen in any of the biopsy fragments, making the diagnosis particularly challenging.

Immunohistochemical Findings

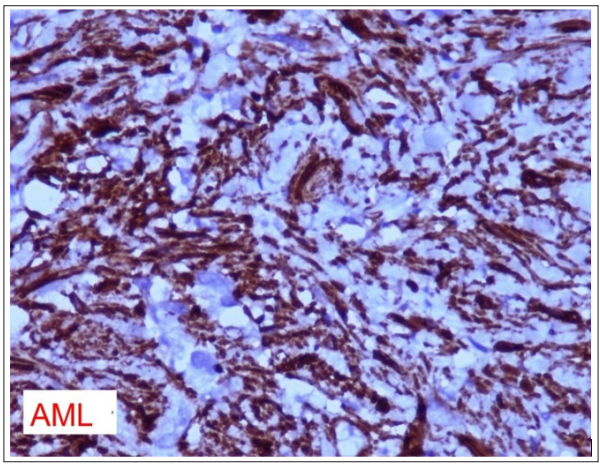

Figure 5 (SMA): Cytoplasmic Positivity Confirming partial Myogenic Differentiation

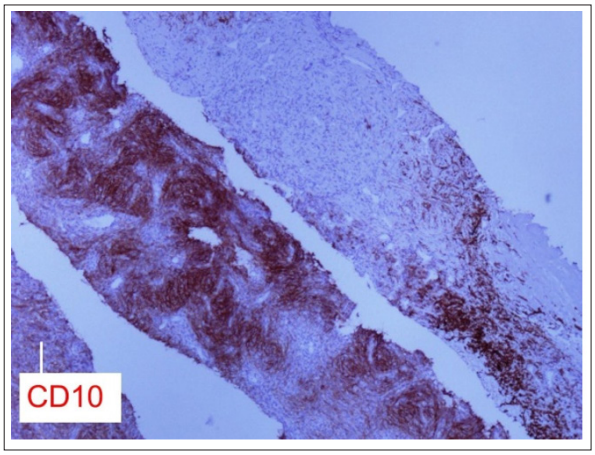

Figure 6 (CD10): Diffuse Membranous and Cytoplasmic Staining

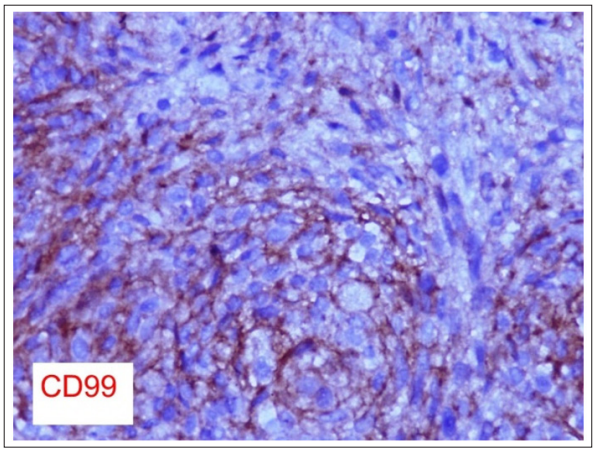

Figure 7 (CD99): Diffuse Positivity with Strong Membranous Pattern

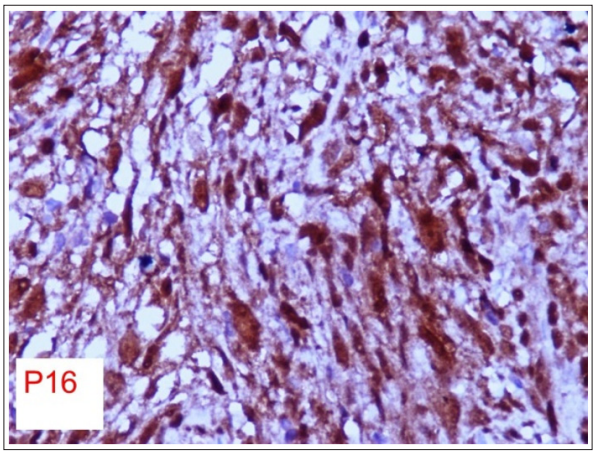

Figure 8 (p16): Diffuse and Intense Nuclear Positivity

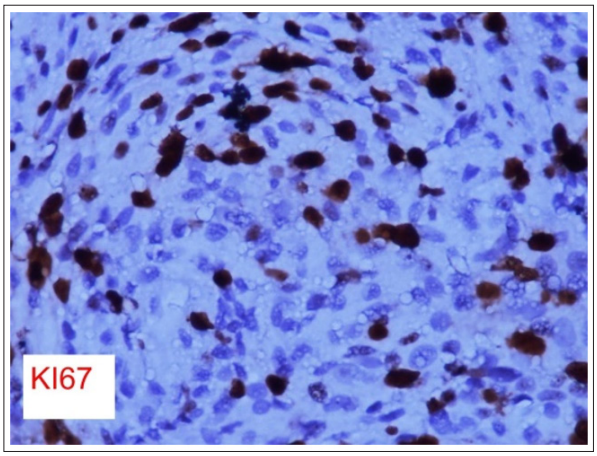

Figure 9 (Ki-67): Proliferative Index Estimated at 25%

Molecular fluorescence in situ hybridization (FISH) analysis demonstrated MDM2 gene amplification, confirming the diagnosis of DDL.

The IHC profile (SMA+, CD10+, CD99+, p16+, MDM2+) and morphologic pattern ruled out differential diagnoses such as myofibrosarcoma, meningioma-like sarcoma, and pleomorphic sarcoma.

Discussion

DDL with meningothelial-like whorls is an uncommon morphologic variant that can cause diagnostic pitfalls, particularly in limited biopsy material. In our case, the lack of a well-differentiated lipomatous component necessitated molecular confirmation via MDM2 amplification, which remains the gold standard for distinguishing DDL from other spindle cell sarcomas.

The p16 positivity and moderate Ki-67 index support an intermediate to high-grade neoplasm. Expression of CD10 and CD99 may reflect mesenchymal differentiation, while SMA positivity suggests myofibroblastic features.

According to previous studies, meningothelial-like whorls may represent an early phase of dedifferentiation or a peculiar stromal response [1,2]. Recognition of this feature and molecular testing for MDM2 are crucial for avoiding misclassification.

Complete surgical excision remains the treatment of choice. The prognosis depends on location, completeness of excision, and histologic grade. DDLs have a significant risk of local recurrence and metastasis, particularly in deep or retroperitoneal locations [3-5].

Conclusion

This case emphasizes the importance of recognizing meningothelial-like whorls as a possible pattern in dedifferentiated liposarcoma. When the well-differentiated lipomatous component is absent, molecular confirmation by MDM2 amplification is essential for accurate diagnosis. A multidisciplinary approach combining histopathology, immunohistochemistry, and molecular pathology ensures optimal diagnostic accuracy and patient management.

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