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Histopathological features of rare soft tissue sarcomas: A case series.

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Introduction

Soft Tissue Sarcomas (STS) represent a heterogeneous group of malignant tumors derived from mesenchymal tissues. While common subtypes such as liposarcomas, leiomyosarcomas, and undifferentiated pleomorphic sarcomas are relatively well characterized, rare variants present diagnostic challenges due to their atypical histopathological features and overlapping morphological characteristics. This case series highlights the histopathological spectrum of rare STS, underscoring the importance of detailed microscopic evaluation and ancillary studies for accurate diagnosis and management.. [1].

A 28-year-old female presented with a slowly enlarging mass in the thigh. Histopathology revealed nests of polygonal cells with abundant eosinophilic cytoplasm arranged in an alveolar pattern, separated by delicate fibrous septa. The tumor cells displayed prominent nucleoli and minimal pleomorphism. Periodic acid-Schiff (PAS) staining showed diastase-resistant crystalline inclusions, which are characteristic of ASPS. Immunohistochemistry (IHC) showed strong nuclear expression of TFE3, confirming the diagnosis.[2].

A 35-year-old male had a firm, painless mass near the ankle. Microscopic examination showed nests and fascicles of clear to eosinophilic spindle cells with centrally located nuclei and prominent nucleoli. Melanin pigment was noted in some cells. IHC was positive for S-100, HMB-45, and Melan-A, mimicking melanoma. However, molecular studies identified the EWSR1-ATF1 fusion gene, definitive for CCSTA. [3]

A 42-year-old male presented with a recurrent forearm lesion. Histopathology revealed nodules of epithelioid and spindle cells with central necrosis, resembling granulomas. The tumor exhibited prominent nucleoli and moderate cytoplasm. IHC showed loss of INI-1 (SMARCB1), a key diagnostic marker. The co-expression of epithelial markers (cytokeratin and EMA) confirmed the diagnosis of epithelioid sarcoma. [4].

A 19-year-old female exhibited a superficial soft tissue mass in the shoulder. Microscopic examination demonstrated pseudovascular spaces, fibrous pseudocapsule, and prominent lymphoplasmacytic infiltrate. Tumor cells were round to spindle-shaped with a syncytial appearance. AFH was confirmed by EWSR1-CREB1 gene fusion and focal desmin positivity. A 55-year-old male had a mass in the thigh. Histology revealed cords and clusters of small round cells embedded in abundant myxoid stroma. The cells had uniform nuclei and scant cytoplasm. Mitoses were rare. IHC was positive for vimentin and NSE but negative for S-100 and cytokeratin. Molecular analysis demonstrated NR4A3 gene rearrangement, consistent with EMC. [5].

Conclusion

These cases underscore the necessity of integrating morphology with immunohistochemical and molecular diagnostics. Rare soft tissue sarcomas often mimic benign or more common malignancies, leading to potential misdiagnosis. Recognizing key histopathological features—such as unique cell

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patterns, cytoplasmic characteristics, and stromal background—alongside genetic studies is critical for precise classification. As treatment decisions and prognostic outcomes are increasingly subtype-specific, accurate histopathological diagnosis plays a pivotal role in clinical manageme nt.

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