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Case study

Metastatic angiomatoid fibrous histiocytoma of the scalp, with *EWSR1-CREB1* gene fusions in primary tumor and nodal metastasis☆

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Summary Angiomatoid fibrous histiocytoma is a rare soft tissue tumor of intermediate biologic potential, predominantly arising in superficial extremities of children and young adults. Less than 5% metastasize. We describe a case of angiomatoid fibrous histiocytoma in the scalp of an 8-year-old boy, which metastasized to a postauricular lymph node 3 years after primary tumor excision. Histologically, primary and metastasis comprised nodules of bland ovoid cells, with primary additionally showing hemorrhagic cavities, fibrous capsule, and lymphoplasmacytic inflammation. Both strongly expressed desmin, with focal epithelial membrane antigen. Reverse transcription–polymerase chain reaction showed *EWSR1-CREB1* fusion transcripts in both primary and metastasis. This is, to our knowledge, the first description of genetically proven metastatic angiomatoid fibrous histiocytoma. Angiomatoid fibrous histiocytoma can mimic both benign and malignant lesions, and although most behave indolently, it is important to recognize their metastatic potential.

1. Introduction

Angiomatoid fibrous histiocytoma (AFH) is a rare tumor of intermediate biologic potential and uncertain differentiation. It typically arises in extremity soft tissue of children and young adults. Most behave indolently, and less than 5% have been shown to metastasize. We describe an AFH of the posterior scalp that metastasized 3 years later to a contralateral regional lymph node, with *EWSR1-CREB1* fusion transcripts detected in both primary and metastatic tumors. This is, to our knowledge, the first description of genetically confirmed metastatic AFH and adds further to the literature regarding the oncogenic potential of the *EWSR1-CREB1* gene fusion, which is present in a group of rare and often aggressive mesenchymal neoplasms [1]. AFH are often morphologically bland and underrecognized neoplasms, and this case highlights the importance of diagnostic familiarity as well as close clinical follow-up for their rare but documented potential for metastasis.

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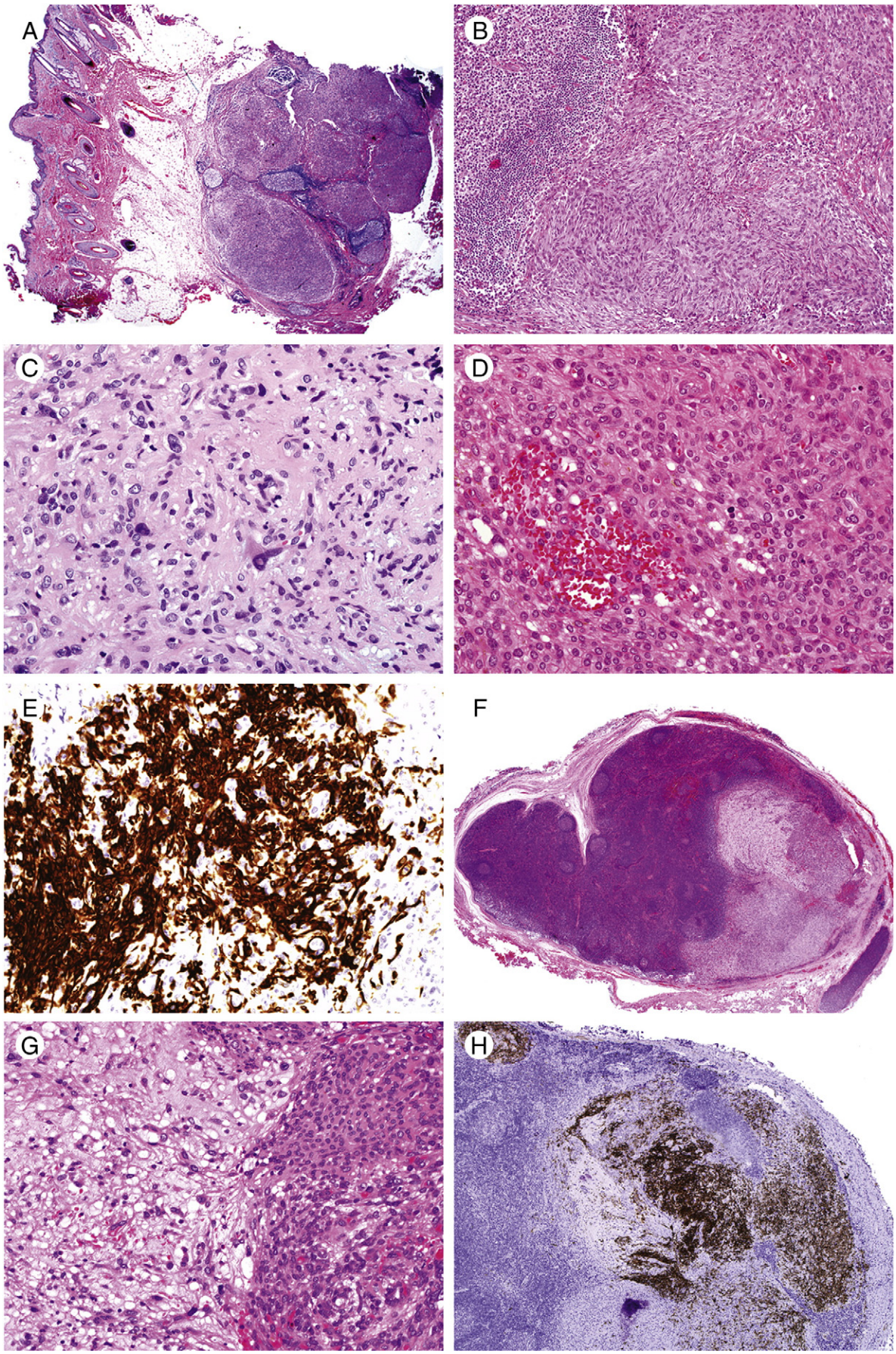
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