Case Report

Epithelioid angiosarcoma of the scalp: An advanced and rare presentation

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Abstract

Epithelioid angiosarcoma (EA) is a rare, aggressive vascular malignancy that often presents with nonspecific cutaneous findings, leading to delayed diagnosis. We report the case of a 73-year-old man with a persistent, painful lesion on the vertex and parietal scalp that failed to respond to multiple courses of antibiotics and topical therapies for presumed infectious and inflammatory dermatoses. Examination revealed crusted erosions and plaques with a nodular component, initially mimicking chronic inflammatory skin disease or a potentially neoplastic entity. Histopathologic evaluation of a punch biopsy demonstrated sheets of atypical epithelioid cells with marked nuclear pleomorphism and frequent mitoses. Immunohistochemical staining confirmed endothelial origin with diffuse positivity for ERG and CD31, while excluding squamous, melanocytic, lymphoid, and epithelial neoplasms. These findings established the diagnosis of EA. The patient was referred for oncology evaluation and initiated on radiation therapy. Cutaneous EA most commonly affects elderly White men and arises on the scalp or face. Prognosis remains poor, with 5-year survival rates below 15% because of high recurrence and early metastatic potential. This case underscores the importance of maintaining a high index of suspicion for EA in nonhealing scalp lesions and highlights the central role of histopathology and immunohistochemistry in establishing the diagnosis.

Introduction

Epithelioid angiosarcoma (EA) is a rare, highly aggressive vascular malignancy characterized by malignant endothelial cells with epithelioid morphology. Although angiosarcomas comprise less than 2% of all soft tissue sarcomas,[1] the epithelioid variant is even more uncommon and presents significant diagnostic challenges because of its histologic mimicry of poorly differentiated carcinomas, melanomas, and other neoplasms.[2] EA most frequently arises in deep soft tissues, but cutaneous involvement, especially on the scalp, has been sporadically reported and is associated with a poor prognosis because of rapid progression and early metastasis.[3]

We present a rare case of EA of the scalp in a 73-year-old man with a history of nonmelanoma skin cancers who was initially misdiagnosed and treated for presumed infectious and inflammatory dermatoses. This case highlights the importance of maintaining a broad differential diagnosis when evaluating nonhealing scalp lesions in older adults and underscores the critical role of histopathologic evaluation and immunohistochemistry in establishing the correct diagnosis.

Case Synopsis

A 73-year-old man presented to the dermatology clinic for reevaluation of a persistent, painful lesion on the vertex and parietal scalp. The lesion had been present for several months and had not responded to multiple courses of oral antibiotics and topical therapies prescribed for presumed infectious or inflammatory dermatoses, including cellulitis, eczema, and erosive pustular dermatosis. His symptoms included tenderness and pruritus, but there were no systemic signs of infection.

On examination, the scalp demonstrated honey-colored, crusted erosions and plaques with a boggy nodular component on the vertex. Additional findings included hyperkeratotic papules with central crusting and excoriation on the left zygoma and preauricular region (**Figure 1**). No regional lymphadenopathy was appreciated.

A punch biopsy of the vertex lesion revealed a dermal proliferation of atypical epithelioid cells exhibiting marked nuclear pleomorphism and frequent mitotic figures. Immunohistochemical staining showed diffuse pos-

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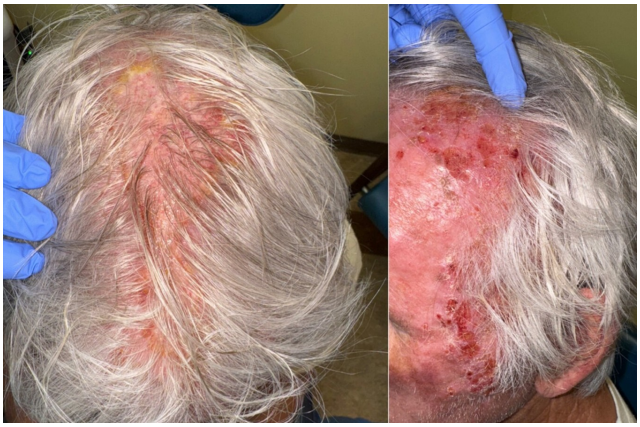

Figure 1. *Honey-colored crusted erosions and plaques with a boggy nodular component on the scalp and hyperkeratotic papules with central crusting and excoriation on the left zygoma and preauricular region.*

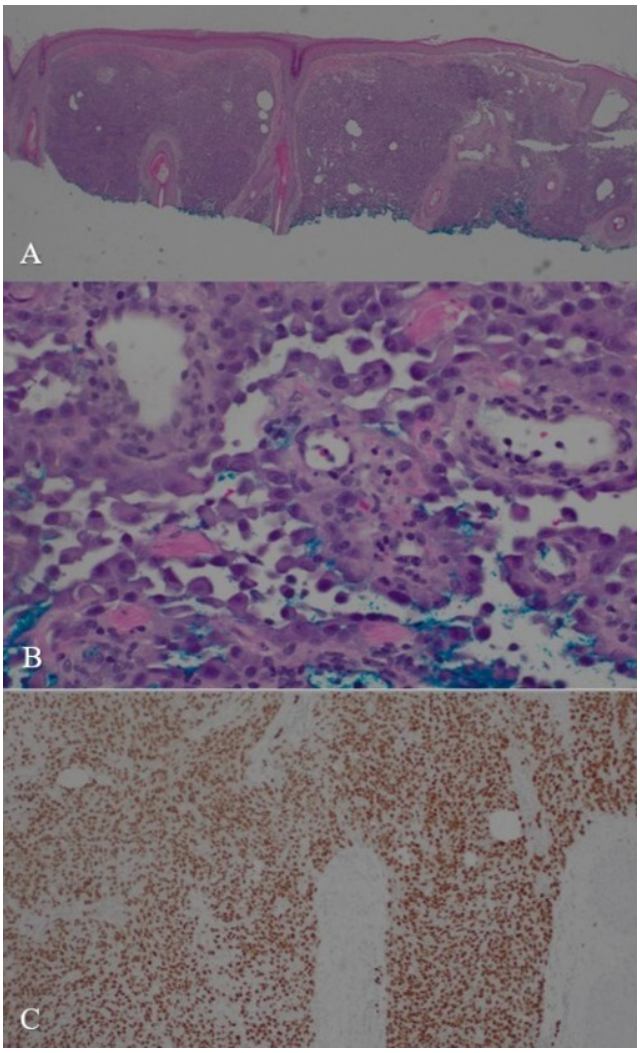

Figure 2. Histopathologic examination showing sheets of atypical epithelioid cells with marked pleomorphism and increased mitotic activity: **(A)** hematoxylin-eosin, low power; **(B)** hematoxylin-eosin, high power; **(C)** ERG immunohistochemical, intermediate power.

itivity for the endothelial markers ERG and CD31. The Ki-67 proliferation index exceeded 25% in focal areas, consistent with a high-grade vascular malignancy. Tumor cells were negative for markers of squamous, melanocytic, lymphoid, and epithelial origin, including p40, SOX10, CD20, CD3, CD10, CK20, and pancytokeratin. These findings were diagnostic of EA of the scalp. The patient was referred to oncology, where he underwent further staging and initiated radiation therapy.

Case Discussion

EA is a rare and highly aggressive vascular malignancy characterized by malignant endothelial cells with epithelioid morphology. Angiosarcomas account for less than 2% of all soft tissue sarcomas, and the epithelioid variant is even less common.[1] Unlike conventional angiosarcoma, which displays clear vascular channels, the epithelioid subtype consists of large, pleomorphic cells with abundant cytoplasm and vesicular nuclei, often arranged in sheets. These features can histologically mimic poorly differentiated carcinoma, amelanotic melanoma, and lymphoma, complicating diagnosis and delaying treatment.[2,3]

Cutaneous angiosarcomas typically present in elderly White men, frequently arising on the scalp or face.[2] Clinically, lesions may appear deceptively benign as ill-defined, bruised, crusted, or inflamed plaques, which can lead to misdiagnoses such as cellulitis, impetigo, or eczema.[4] In this case, the patient's chronic scalp lesion persisted despite multiple empiric treatments, resulting in delayed diagnosis.

Histopathologic examination revealed sheets of atypical epithelioid cells with marked pleomorphism and increased mitotic activity (Figure 2). Immunohistochemistry played a central role in diagnosis, confirming endothelial origin with ERG and CD31 positivity and ruling out carcinoma, melanoma, and lymphoma with negative staining for epithelial and melanocytic markers. This diagnostic approach highlights the importance of a broad immunohistochemical panel in evaluating atypical cutaneous lesions.

Prognosis in EA is generally poor, with 5-year survival rates often below 15%, largely owing to the tumor's aggressive behavior, high recurrence rates, and propensity for early metastasis.[4] Management typically includes wide local excision with or without adjuvant radiation or chemotherapy, although treatment strategies remain poorly defined owing to the condition's rarity.[5] This case emphasizes the need for a high index of suspicion when evaluating chronic, nonhealing scalp lesions in older adults and demonstrates the critical role of histopathology and immunohistochemistry in achieving an accurate and timely diagnosis of EA.

Conclusion

This case illustrates the diagnostic challenges of EA of the scalp, a rare and aggressive vascular malignancy that can

clinically resemble common inflammatory or infectious dermatoses. The patient's prolonged course of empiric treatments highlights how easily the diagnosis may be delayed without biopsy and immunohistochemical evaluation. Recognition of atypical, nonhealing scalp lesions in elderly patients, coupled with prompt histopathologic workup, is critical for timely diagnosis and initiation of appropriate therapy. Ultimately, this case underscores the importance of maintaining a broad differential diagnosis for refractory cutaneous lesions and the essential role of dermatologists in facilitating early detection of rare but life-threatening malignancies.

Potential conflicts of interest

The authors declare no conflicts of interest.

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