Indurated purple plaques on the scalp

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Abstract

Cutaneous epithelioid angiosarcoma is a rare neoplasm of vascular endothelial cell origin that can mimic a cutaneous lymphoma, metastatic carcinoma, or Kaposi sarcoma. It is one of the most malignant cutaneous tumors and early diagnosis is essential, as the tumor metastasizes quickly. We describe a 75-year-old man who presented with three tender, indurated violaceous plaques on his scalp. Biopsy revealed a poorly circumscribed infiltrate extending into the subcutaneous fat, composed of atypical epithelioid cells lining vascular spaces. We provide a brief review of the clinical presentation, histopathologic features, differential diagnosis, and management of this rare tumor.

Keywords: epithelioid angiosarcoma, cutaneous lymphoma, carcinoma, Kaposi sarcoma

Introduction

Epithelioid angiosarcoma is a rare morphologic subtype of angiosarcoma in which the malignant endothelial cells demonstrate epithelioid morphology. The majority of epithelioid angiosarcomas arise in the deep soft tissues of the extremities, although cutaneous presentations have also been reported. We describe a 75-year-old man with a remote history of radiation exposure who presented with three enlarging tender plaques on his scalp. On clinical exam, the plaques were worrisome for cutaneous lymphoma. However, routine histology and immunohistochemical staining confirmed the tumor's vascular origin, supporting a diagnosis of cutaneous epithelioid angiosarcoma.

Case Synopsis

A 75-year-old man presented to dermatology clinic with three purple plaques on his scalp present for two months. The lesions were tender and increasing in number and size. His past medical history was significant for hypercholesterolemia and a remote episode of radiation therapy to his scalp for presumed tinea capitis when he was ten years old. He had no personal or family history of skin cancer. Physical examination revealed three 2-3cm pink-to-violaceous indurated plaques — two on the right frontal scalp (**Figure 1**) and one on the left parietal scalp (not shown). There was no palpable cervical lymphadenopathy. The remainder of the skin examination was unremarkable. A 4.0mm punch biopsy specimen was obtained from the right frontal scalp for histologic analysis with hematoxylin-eosin (**Figures 2, 3**) and immunohistochemical staining (**Figure 4**).

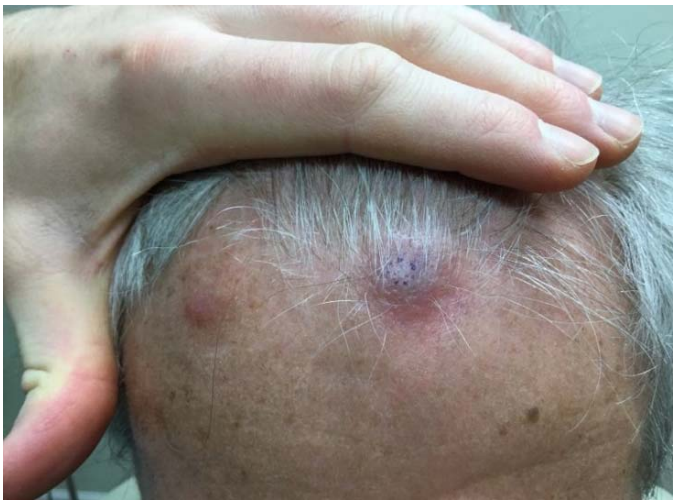

Figure 1. *Clinical photograph shows two 2-3cm violaceous indurated plaques on the frontal scalp.*

Biopsy of the right frontal scalp revealed a poorly circumscribed dense blue infiltrate involving the full thickness of the dermis extending into the subcutaneous fat (**Figure 2**). On higher power, the tumor was composed of atypical epithelioid cells with vesicular nuclei, some of which lined irregular slit-like fissures (**Figure 3**). The epithelioid cells lining the vascular spaces were diffusely positive for CD31 (**Figure 4**). The tumor was negative for CD20, SOX-10, and HHV-8. Taken together, these findings supported a diagnosis of cutaneous epithelioid angiosarcoma.

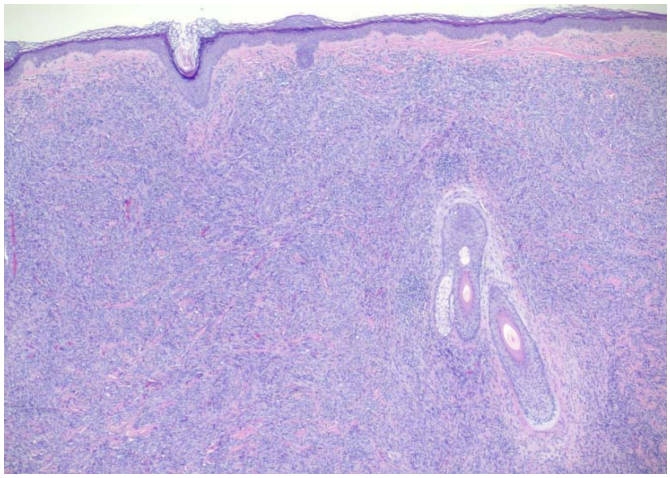

Figure 2. *Punch biopsy specimen from the right frontal scalp shows a dense blue infiltrate involving the full thickness of the dermis extending into the subcutaneous fat. H&E, 10×.*

Case Discussion

Cutaneous angiosarcoma is a rare neoplasm of vascular endothelial cell origin. It is one of the most malignant tumors involving the skin, with five-year survival rates between 12% and 20% [1]. Three common variants include idiopathic angiosarcoma on the head and neck of elderly individuals, radiation-associated angiosarcoma, and angiosarcoma in the setting of chronic lymphedema. The tumor often presents as a hematoma-like plaque or nodule, but lesions resembling rosacea, hemangioma, cellulitis, and angioedema have also been reported [2]. Common histopathologic features include poor circumscription with irregularly dilated matrices of vessels lined by atypical endothelial cells that dissect through the dermis [3].

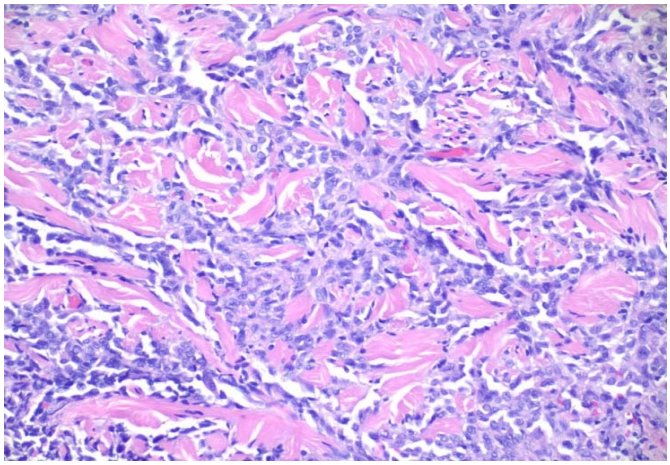

Figure 3. *Large epithelioid cells with abundant eosinophilic cytoplasm, vesicular nuclei, and prominent nucleoli can be seen, some of which line vascular spaces. H&E, 40×.*

Though focal epithelioid changes may be seen in many angiosarcomas, the term epithelioid angiosarcoma is typically reserved for tumors in which greater than 80% of malignant cells are epithelioid [4]. Microscopically, these cells appear large with abundant eosinophilic cytoplasm, central vesicular nuclei, and prominent nucleoli. The tumors most frequently arise in deep soft tissues, but cutaneous and visceral lesions have also been reported [5]. Cutaneous epithelioid angiosarcoma is more common in males and generally occurs later in life, with the highest incidence occurring in a patient's seventh decade of life, as presented in this case [6].

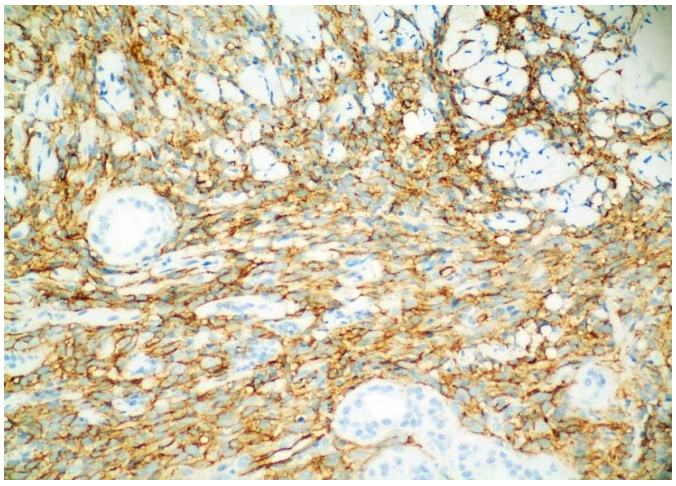

Figure 4. *The epithelioid cells seen here are diffusely positive for CD31 staining. Immunohistochemical, 40×.*

The morphologic and histologic appearance of epithelioid angiosarcoma may resemble primary or metastatic carcinoma. However, areas of focal vasoformation or intracellular lumina reveal the endothelial nature of epithelioid angiosarcoma. Moreover, carcinomas are negative for endothelial markers such as CD31 or CD34, whereas positivity for CD31 is seen in almost all cases of epithelioid angiosarcoma [7]. A negative SOX-10 in tumor cells helps to exclude a melanocytic neoplasm such as melanoma.

Other diagnoses to consider based on clinical and histopathological characteristics include Kaposi sarcoma and cutaneous lymphoma. Kaposi sarcoma — associated with HHV-8 — may also appear as violaceous plaques. Histopathology would reveal spindle cells and vascular structures in a network of reticular fibers; nuclear atypia is usually less prominent [8]. Immunohistochemical staining for B and T cell neoplasms with routine histology can help exclude cutaneous lymphomas. Tumor cells, positive for CD31 and CD34 but negative for B and T cell markers and HHV-8, favor a vascular etiology and can aid in the diagnosis of angiosarcoma.

Epithelioid angiosarcoma often metastasizes early, particularly to the lungs, bone, soft tissue, and skin. Adverse prognostic factors include advanced age, tumor size greater than 5cm, bleeding, and an increased proliferative index [5]. Treatment options typically consist of surgical resection of the primary tumor, with any need for adjuvant radiotherapy or chemotherapy determined on an individual basis. Our patient chose to receive wide surgical excision followed by radiation therapy.

Conclusion

Cutaneous epithelioid angiosarcoma is a morphologic subtype of angiosarcoma that may mimic metastatic carcinoma, Kaposi sarcoma, or cutaneous lymphoma. Indeed, the initial low power impression in this case may raise suspicion for a B-cell lymphoma. However, the patient's remote history of scalp radiation for presumed tinea capitis, advanced age, and tumor location should serve as a reminder to consider angiosarcoma in the differential diagnosis, even when the initial clinical presentation may not appear vascular. This patient demonstrates a presentation of both head and neck location and possible radiation-associated angiosarcoma. A thorough clinical evaluation with appropriate understanding of clinical and histopathologic characteristics is essential to arrive at the correct diagnosis.

Potential conflicts of interest

The authors declare no conflicts of interests.

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