Angiosarcoma Imitating a Morpheaform Basal Cell Carcinoma

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PRACTICE **POINTS**

- Angiosarcoma is an aggressive vascular tumor with a poor prognosis.
- Angiosarcomas can arise in the setting of chronic lymphedema or prior radiation therapy or can arise spontaneously.
- Classically, angiosarcoma presents as a violaceous patch or plaque but occasionally can exhibit atypical clinical features. Angiosarcomas should be considered on the differential for any changing plaque on the head or neck.

To the Editor:

Basal cell carcinoma (BCC) is the most common of the nonmelanoma skin cancers and is a highly curable skin growth.[1,2] Conversely, angiosarcomas are aggressive vascular tumors of endothelial origin that classically appear as reddish purple patches or plaques that exhibit rapid growth and invasion.[3] Sporadic cutaneous angiosarcomas are the most common type of this soft tissue tumor, occurring most often in the head and neck regions in men older than 70 years.[4,5] Other types of angiosarcomas include those associated with radiation therapy and chronic lymphedema. Postradiation angiosarcomas have been most frequently reported after treatment of breast cancer and appear as infiltrative plaques over the irradiated area.[4,5] Patients with chronic lymphedema, which most commonly is related to axillary lymph node dissection for breast cancer (90% of cases), may develop angiosarcoma presenting as a violaceous indurated plaque.[5] Although angiosarcomas most often are seen with these distinct clinical characteristics, especially their violaceous color, they have been shown to mimic a few other skin disorders such as eczema and keratoacanthoma, but a limited number of cases of angiosarcoma mimicking BCC have been reported.[1,6,7] We present a case of an elderly man with a unique presentation of a lesion that clinically appeared as a morpheaform BCC but was confirmed to be an angiosarcoma on histopathology.

A 75-year-old man was referred to our dermatology clinic for evaluation of a flesh-colored plaque on the face that initially had developed 2 years prior on the right central malar cheek. Computed tomography of the head and neck 1 year prior, which the patient reported was for workup of the lesion, was found to be negative; however, these medical records were not obtained for confirmation. The lesion had been stable in size and remained flesh colored until 6 months prior to the current presentation when it exhibited a rapid increase in size. An initial biopsy was performed 1 month prior to presentation by an outside dermatology office and had been read as an angiosarcoma.

Physical examination revealed a 6-cm, flesh-colored, indurated, ill-defined plaque distributed on the right malar cheek below the eye and extending to the nasal bridge (Figure 1). There was no cervical or facial lymphadenopathy. The clinical features resembled a morpheaform BCC, and the lesion did not exhibit any reddish or purple color indicating it was of vascular origin. However, due to the prior histopathology report and recent rapid enlargement, a repeat sampling with a larger punch biopsy was performed, which confirmed the diagnosis of angiosarcoma. Histopathology demonstrated multiple atypical vascular channels lined by hyperchromatic cells extending from the upper dermis to the base of the biopsy site (Figure 2). Large, oval, atypical nuclei were present in multiple endothelial cells in the vascular channels, with some forming irregularly contoured and

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slitlike formations (Figure 3). Immunochemical staining was intensely and uniformly positive for CD31 and CD34, both endothelial markers. Diffuse positive staining with CD31 is considered to have high sensitivity and specificity for the diagnosis of angiosarcoma.[4] Other pertinent staining demonstrated 2+ positivity for factor VIII and 1+ positivity for D2-40; CD45, AE1/AE3, S-100, and human herpesvirus 8 were negative, consistent with angiosarcoma. The patient was referred to radiation oncology and otolaryngology at our Multidisciplinary Head and Neck Oncology Center for further investigation of the extent of the disease and discussion of treatment. Computed tomography of the head and neck region at this time showed extensive disease extending into the medial canthal area without metastasis. Due to the extent of disease and facial location, he was not deemed a candidate for surgery. He was treated with 6 weeks of targeted radiation therapy with concurrent chemotherapy. He tolerated this treatment with minimal side effects and was found to be free from clinical disease 1 year after diagnosis. He was followed for 20 months by our Multidisciplinary Oncology Clinic without recurrence of his disease but was then lost to follow-up.

This case illustrates a rare presentation of an angiosarcoma clinically mimicking a BCC, which has been described in a small number of case reports and retrospective reviews. One study of 656 patients diagnosed with BCC based on clinical features revealed that 48 of these lesions were proven to be a BCC-mimicking lesion and only 1 was an angiosarcoma.[1] Cutaneous lesions that appear on physical examination to be a highly curable BCC may not induce the same urgency for treatment as an angiosarcoma. Although the clinical presentation may mimic a morpheaform BCC, our case demonstrates that it is imperative to include angiosarcoma in the differential diagnosis and underscores the utility of tissue sampling. Angiosarcoma has a poor overall 5-year survival rate, and patients often are found to have multiple metastatic lesions at diagnosis. However, diagnosis prior to metastasis may improve prognosis.[8]

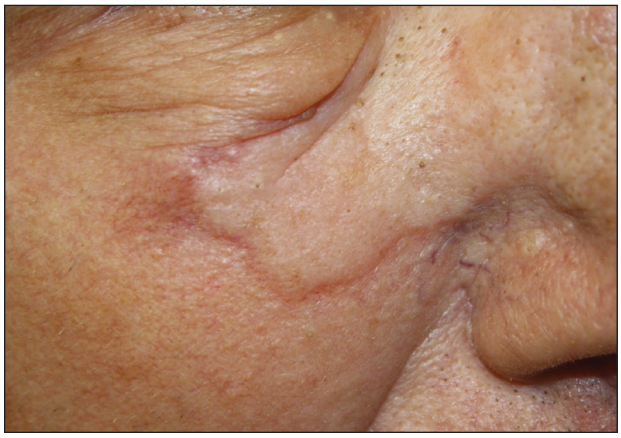

FIGURE 1. A 6-cm, indurated plaque distributed on the right malar cheek extending to the nasal bridge.

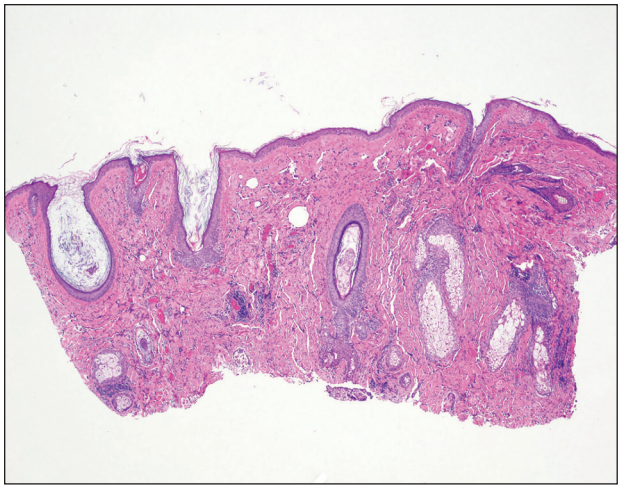

FIGURE 2. Punch biopsy demonstrated epidermal thinning with a perivascular lymphocytic infiltrate and background solar elastosis. There was a proliferation of atypical vascular channels from the upper dermis extending to the base of the biopsy site (H&E, original magnification ×40).

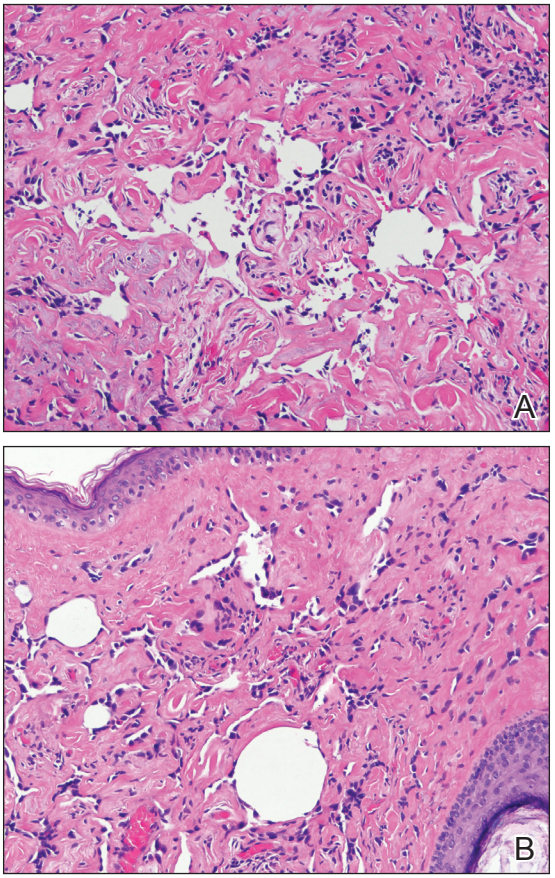

FIGURE 3. A and B, Atypical vascular channels and hyperchromatic cells. Many of the endothelial cells exhibited hyperchromatic nuclei, with some forming slitlike and irregularly contoured channels (H&E, original magnifications ×200 and ×200).

Our patient's angiosarcoma did not exhibit metastasis at the time of diagnosis, and he was able to achieve a favorable outcome. However, the 5-year survival rate is only 40%, and close clinical monitoring after diagnosis is required.[8] Including angiosarcoma in the differential diagnosis for our patient, particularly upon lesion appearance 2 years prior, may have resulted in diagnosis antecedent to local invasion, possibly providing more treatment options. Employing a higher index of clinical suspicion for angiosarcoma may lead to decreased mortality in other patients due to increased detection.

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