Cutaneous angiosarcoma of the head and neck: A case presentation and review of the literature

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Abstract
Cutaneous angiosarcoma of the head and neck is a rare vascular neoplasm. When it does occur, it is most common in elderly white men. Early diagnosis and treatment are essential for local control of this aggressive tumor, but recognition can be delayed because of its rarity or because of difficulty in making a pathologic diagnosis. A combined-modality treatment approach is most often advocated. We report the case of a 77-year-old black man who presented with a 1-month history of two painless, violaceous, subcentimeter nodules of the upper lip. After a diagnosis of low-grade angiosarcoma was definitively established, the lesions were locally excised with good cosmetic and functional results. The patient subsequently was found to have probable metastatic disease, but he declined further intervention. We review the literature on cutaneous angiosarcoma, and we discuss its epidemiology, presentation, tissue diagnosis, treatment, and prognosis in an effort to increase awareness of this rare malignancy.

Introduction
Angiosarcoma is a rare malignancy of the head and neck. It is usually seen in the elderly, and the prognosis is dismal. Diagnosis can be delayed as a result of the entity’s rarity or confounding tissue histology. Early detection and treatment are essential to control this highly malignant soft-tissue cancer.

Case report
A 77-year-old black man presented to another subspecialty clinic with a 1-month history of two painless, violaceous, subcentimeter nodules of the upper lip. Initial histologic findings suggested that these lesions were pyogenic granulomas. After a few months, similar nodules developed on the skin of the midface; they were biopsied and diagnosed as low-grade angiosarcomas. The initial pathologic specimens were reviewed and considered to be epithelioid hemangioendotheliomas. The patient was referred to our clinic for evaluation and management.

On clinical examination, the patient was generally fit and in no distress. Several nontender, violaceous, smooth, fleshy, nonblanching, pedunculated lesions measuring less than 1 cm each were scattered around the upper lip and nose (figure 1, A). No cervical lymphadenopathy was found. Findings on the remainder of his head and neck examination were unremarkable.

A metastatic workup was initiated. Results of the basic metabolic profile and liver function tests were within normal ranges. The complete blood count revealed normocytic anemia. A finding on chest plain film was suspicious for an ascending aortic aneurysm. Computed tomography of the chest and abdomen demonstrated a large, low-density left retroperitoneal mass with enhancing peripheral nodules and two similar masses in the right hepatic lobe. These lesions were radiologically consistent with metastasis of the angiosarcoma.

The patient refused further diagnostic procedures for the abdominal masses, but he did undergo local excision of the facial lesions. The postoperative cosmetic and functional results were good (figure 1, B).

Pathologic examination revealed an angiosarcoma of varied differentiation. The specimen was made up of pleomorphic spindle cells with an indistinct luminal formation (figure 2). These findings were in contrast with those of the patient’s previous biopsy specimens, which lacked nuclear pleomorphism.

Discussion
Epidemiology/presentation. In the United States, soft-tissue sarcomas account for less than 1% of all malignancies; 5 to 15% of these tumors occur in the head and neck. The most common soft-tissue sarcomas encountered in otolar-
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Pathology. Angiosarcoma is a difficult diagnosis to make, even for experienced pathologists, because of its rarity and complex histology. Cutaneous angiosarcomas may be grouped on the basis of gross and histologic features. Grossly, a lesion may be nodular, ulcerative, or macular. Histologically, tumors contain a proliferation of ramifying and anastomosing vascular channels that “dissect” through surrounding structures. Increased numbers of plump endothelial cells line the vascular channels; these cells may be spindled, epithelioid, or polygonal. Tumors are classified as low-grade or high-grade, depending on their cellularity, pleomorphism, and mitotic activity. Necrosis is a common finding. Although no immunohistochemical stain is pathognomonic, the diagnosis may be aided by a finding of positivity for factor-VIII-related antigen, Ulex europaeus, laminin, CD31, and CD34, although a finding of factor-VIII-related antigen positivity may not be reliable. Electron microscopy will occasionally reveal rod-shaped, microtubulated Weibel-Palade bodies, but again, this finding may not be particularly reliable, as only 1 of 13 specimens demonstrated this structure in one series.

Treatment/prognosis. The optimal treatment has not been completely established. For advanced disease, radical excision has historically been advocated. Surgery alone or irradiation with or without chemotherapy may yield similarly dismal results. Mark et al reported that only 2 of 19 patients (10.5%) who received a single-modality treatment were alive at the time of their review, and all 19 had a component of local failure.

Surgical control is quite often poor because of the multifocality of angiosarcoma and the lack of clinical correlation with pathologic margins. In a small series, Pullen et al achieved good local control with Mohs’ surgery and punch-biopsy tumor mapping followed by electron-beam radiation.

The failure of irradiation therapy can often be attributed to an underestimation of the peripheral extension of disease. Therefore, some authors advocate total scalp irradiation. Surgeons at the M.D. Anderson Cancer Center in Houston reported using limited surgical resection followed by wide-field irradiation for focal disease, and they were able to treat patients with advanced disease who would have otherwise required radical excision plus radiation with or without chemotherapy. Although their overall results were poor, they did see a statistically significant difference in 5-year locoregional control rates above the clavicles when gross tumor resection was possible prior to radiation therapy (40 vs. 24%). Therefore, many authors advocate postoperative irradiation therapy despite negative surgical margins.

Definite advances in systemic therapy have been slow to develop. Because of the low incidence of cervical metastasis, elective treatment of the cervical lymph nodes is not recommended. Paclitaxel as a single agent appears to be useful for scalp and facial angiosarcomas. Sasaki et al recommended curative radiation therapy combined with recombinant interleukin-2 (rIL-2) immunotherapy for nodular lesions smaller than 5 cm, in part because high-dose rIL-2 administration may suppress the development of distant metastasis. The current consensus is to use a combined-modality approach, including excision of disease with negative margins if possible with adjuvant radiotherapy with or without chemotherapy.

A clear correlation exists between the prognosis and the size or initial stage of an angiosarcoma. The importance of early capture cannot be understated. Bleeding, pain, and lesion size greater than 5 cm are predictors of a poor outcome. Lydiatt et al reported that all patients in their series whose initial tumor was surgically unresectable and/or larger than 10 cm died of disease. The results of most series have indicated that neither the tumor’s clinical pattern nor its grade is predictive of local recurrence or survival. However, one study indicated that long-term survival might be positively correlated with gross nodular morphology, while an endophytic presentation was uniformly fatal within 2 years. Recurrent disease also confers a dismal prognosis.

References