What Is Your Diagnosis?

An 83-year-old man presented with left upper eyelid ptosis and painless left periorbital swelling of 9 months' duration following facial trauma. He initially had been treated with oral antibiotics and steroid-antibiotic ointment without improvement. The affected eyelids and medial canthus displayed a purple to yellow discoloration.

PLEASE TURN TO PAGE 159 FOR DISCUSSION

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The Diagnosis: Angiosarcoma Arising in the Eyelid

An 83-year-old Hispanic man presented with swollen but painless left upper and lower eyelids of 9 months' duration. He attributed the swelling to a fall sustained at home 9 months prior when he tripped and landed on the left side of his face. Following the trauma, he initially was treated with oral antibiotics and steroid-antibiotic ointment without any improvement. The patient's medical history was notable for congestive heart failure and a cardiac ejection fraction of 20%. There was no history of radiation therapy or chronic lymphedema. Physical examination revealed a thickened left upper eyelid with 2 mm of blepharoptosis. Purple to yellow discoloration was present in the medial aspect of the upper and lower eyelids, extending into the medial canthus (Figure 1). Motility was full and there was no proptosis or displacement of the globe of the eye. A computed tomography scan revealed eyelid soft tissue thickening. Both orbits were unremarkable, but there was slight ethmoid and maxillary sinus disease.

Angiosarcomas are rare malignant neoplasms of vascular origin. These mesenchymal tumors can arise in numerous sites of the body but most commonly present in the skin of the head and neck. They may occur de novo or in association with a history of chronic lymphedema or prior radiotherapy. Older light-skinned individuals tend to be affected most often, leading some to implicate actinic exposure as a causative effect.[1,2] The clinical presentation of angiosarcoma is variable. It typically has a bruiselike appearance and can resemble cellulitis or edema but may alternatively lack a vascular appearance altogether.[1] Unusual presentations such as yellow plaques or fleshy nodules have been described.[1,3] Not surprisingly, accurate diagnosis of angiosarcoma often is delayed because the clinical presentation is not suspicious enough to warrant a biopsy.[2]

Figure not available online

Figure 1. A ptotic left upper eyelid and purple to yellow discolored left periorbital skin.

Biopsy results of the left upper eyelid of our patient (Figure 1) revealed angiosarcoma. Histology showed an infiltrative irregular network of anastomosing slitlike vascular channels lined by large, atypical, hobnailed endothelial cells with hyperchromatic round to oval nuclei (Figures 2A and 2B). Immunohistochemical stains for factor VIII, CD31 (Figure 2C), and CD34 confirmed the vascular differentiation of the tumor.

The predominant histopathologic appearance of angiosarcoma, as seen in our case, includes a well to moderately differentiated lesion showing an invasive pattern of slitlike or cystically dilated vascular channels lined by large, atypical, hobnailed endothelial cells. Some tumors display a less common, moderately to poorly differentiated pattern of spindled cells with eosinophilic cytoplasm in sheets or islands.[1,2] Mixtures of these 2 patterns also are seen. Immunohistochemical stains for factor VIII, CD31, and CD34 confirm the diagnosis.

The clinical differential diagnosis of angiosarcoma is broad because of its protean presentation and includes inflammatory skin lesions and trauma, as well as benign, malignant, or metastatic lesions.[2] Histologically, the diagnosis typically is not difficult if an adequate biopsy is performed. The histologic differential diagnosis includes Kaposi sarcoma, other poorly differentiated sarcomas, and benign lymphatic and vascular proliferations.

Angiosarcoma is aggressive and often has a poor prognosis. Reports of a 5-year survival rate range from 12% to 34%.[1,2,4] Local recurrence and metastatic spread occurring months and years following resection are common, with 5-year local recurrence rates reported as high as 84%.[1,2] Tumors often spread hematogenously to the lungs or liver but also can metastasize via the lymphatic system to lymph nodes. A study of 23 cases of angiosarcoma demonstrated that the only remarkable prognostic factor is the histologic grade of the tumor, with high-grade lesions (poorly differentiated, high mitotic rate, necrosis) having the worst prognosis.[4]

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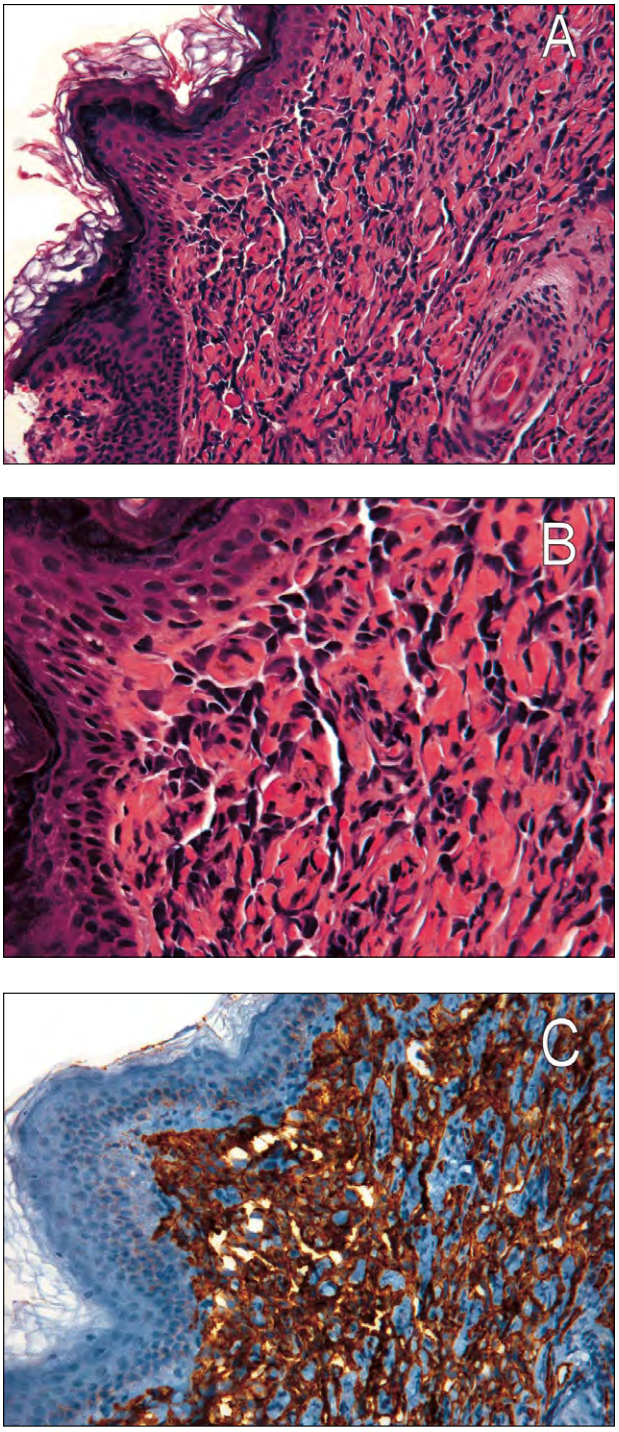

Figure 2. Anastomosing vascular spaces filled with erythrocytes and lined by atypical endothelial cells. Numerous extravasated erythrocytes were seen (A and B)(H&E; original magnifications ×20 and ×40, respectively). Immunostain for CD31 showed strong immunoreactivity of the neoplastic cells (C)(original magnification ×20).

Wide excision with negative margins, when possible, is still the preferred treatment, but delineation of surgical margins can be difficult because angiosarcomas tend to be more microscopically extensive than clinically perceived. Additionally, location of the tumor, such as the periorbital region in our case, can make wide excision challenging.[5] Chemotherapy and radiation as primary or adjuvant treatment have demonstrated mixed results.[2,4] Doxorubicin and paclitaxel have shown some usefulness as neoadjuvant and postoperative chemotherapies[4]; however, early detection and removal remain the best chance for cure.

Angiosarcoma involving the eyelid is rare and most reported cases represent secondary local spread to the eyelid from an adjacent site on the face.[3,5-8] In our case, the tumor initially presented as upper eyelid swelling and ptosis that spread to the adjacent lower face. Our patient declined surgical and medical therapy because of his cardiac disease. However, his presentation of eyelid swelling reminds the clinician of the potentially confounding clinical appearance of this rare but highly aggressive vascular neoplasm.

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