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LETTER TO THE JOURNAL

Sir,

Excisional surgery of periocular angiosarcoma

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Angiosarcoma is a rare and malignant vascular tumour of endothelial cell origin that may arise in the skin and subcutaneous tissues. Cutaneous angiosarcoma of the face and scalp forms a distinct subgroup that generally occurs in elderly, Caucasian patients.[1–3] This tumour carries a poor prognosis despite treatment.[3–5] We present a patient with periocular angiosarcoma treated with wide, but eye-sparing, surgical excision and eyelid reconstruction. During 3 years of follow-up, he has remained disease-free.

Case report

A 71-year-old Caucasian male was referred with a 6-week history of a red, painless swelling of his left upper eyelid and medial canthus, not responding to systemic antibiotics.

Histopathologic examination of an incisional biopsy, performed elsewhere, showed evidence of a chronic, fibrosing inflammation with a remarkable proliferation of capillary vessels, without signs of malignancy.

The patient complained of visual field loss owing to the upper lid swelling, a foreign body feeling and epiphora. On examination, we found a red, nontender pretarsal swelling of the medial part of the upper eyelid, the left medial canthus, and the medial part of the lower eyelid (Figure 1). There was no proptosis, and the ocular motility was not impaired. There was also a diffuse redness of the skin of the nose, suggestive of rhinophyma. The visual acuity of the right and left eye was 0.8 and 0.7 (Logmar acuity), respectively. An orbital CT showed a soft-tissue swelling in the left upper lid and medial canthal area.

Revision of the histopathology confirmed the earlier findings of nonspecific chronic inflammation. Internal examination showed no evidence for a systemic vasculitis. A biopsy from the lid lesion was repeated. Histopathologic examination now showed features suggestive of angiosarcoma. For confirmation of the diagnosis, the material was sent elsewhere, where the tumour was classified as an epitheloid angiosarcoma.

The patient was subsequently treated with wide surgical resection of the tumour with frozen section controls, which resulted in a defect consisting of half of the upper eyelid, the medial canthus, and one-third of the lower eyelid of his left eye.

For reconstruction of the defect, we used a glabella-flap, lateral cantholysis, tarsal sliding flaps of the upper and lower eyelid, and a free skin-graft. The histopathology showed a radically excised angiosarcoma (Figure 2).

Several months later, we biopsied the left nasal ala of his rhinophyma. The histopathology showed features suggestive of rosacea, but no angiosarcoma. After 3 years of follow-up, the patient is well and without evidence of local recurrence or distant metastasis of the tumour.

Comment

This case illustrates that in localized, periocular angiosarcoma, radical excision can be achieved with preservation of the eye. After adequate reconstruction of the lid and medial canthal defects, the patient has been able to normally use his left eye, despite mild complaints of tearing.

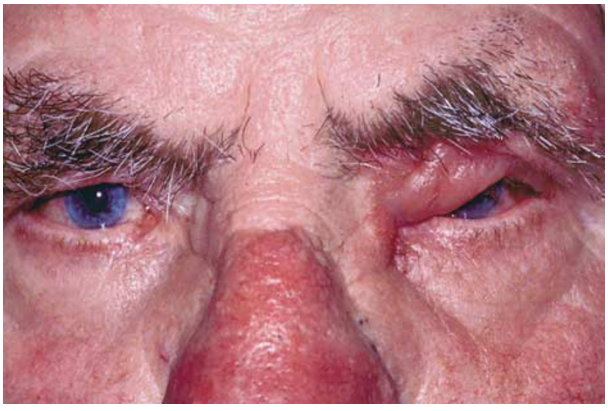

Figure 1 Initial presentation of the patient with swelling and purple-red discoloration of his left upper eyelid and medial canthal area. Note the rhinophyma.

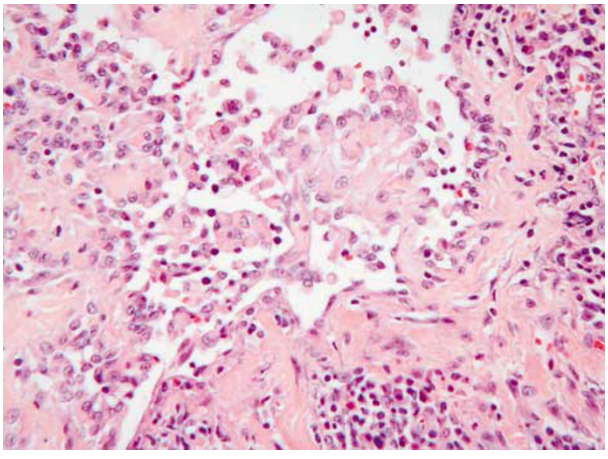

Figure 2 Large atypical cells proliferating as papillairy projections within a large vascular sinus (haematoxylin and eosin, $\times$ 400 magnification).

Wide surgical excision is the first choice of therapy in small angiosarcomas. This may be followed by wide-field radiotherapy in cases where insufficient tumour-free margin can be obtained. If surgical excision is not possible in the case of diffuse or multifocal tumour, radiotherapy is the first choice in therapy. Chemotherapy has been proven of very limited benefit.[3–5]

The most common features are bruise-like macules, plaques and nodules or ulceration. It remains unclear whether these tumours arise *de novo*, although some cases are reported in which angiosarcoma developed in postradiation tissue or after chronic lymphedema.[1–5] Furthermore, angiosarcoma can present as rosacea clinically[6,7] and Gallardo *et al*[8] have described a patient in whom an angiosarcoma arose on a rhinophyma. In our patient with periocular angiosarcoma, a rhinophyma was present, but histopathology of the involved nasal skin showed no features suggestive of angiosarcoma. It remains unclear if there is a correlation between rosacea and angiosarcoma.

The prognosis is very poor. Local recurrence and distant metastasis is frequent and the 5-year survival ranges from 12 to 40%. The best prognostic factors are a small tumour size and the ability to obtain wide surgical margins.[3–5] Angiosarcoma of the eyelid has been reported before, and indeed the patients who could be treated with radical surgical excision seemed to have a very good prognosis.[9–12]

In our patient, there has been no sign of local recurrence or distant metastasis during 3 years of follow-up, albeit that the latter may still develop.

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