Sir,

Massive infarction of ocular tissues complicating a necrotic uveal melanoma

Necrosis of uveal malignant melanoma is not unusual and may be accompanied by symptoms and signs of intraocular inflammation. It is more unusual for melanoma to be associated with orbital inflammation without evidence of extra-scleral spread. When orbital inflammation occurs, it is invariably associated with tumour necrosis and intraocular inflammation. This case further illustrates that uveal melanoma may present with a clinical picture mimicking orbital cellulitis. The novel observation is that the florid orbital inflammation found in this case was due to massive infarction not only of the tumour, but of most of the eye. The mechanism for this previously unreported scenario is discussed, with an interplay of vasculitis and raised intraocular pressure seeming likely.

Case report

A 64-year-old man presented with a 4 day history of right eye pain and swelling of the upper and lower lids. The affected eye was blind following an injury as a child. On examination, there was marked periorbital oedema and erythema. Severe conjunctival chemosis hid the cornea. Corneal oedema and a hyphaema obscured intraocular detail. Intraocular pressure was normal when assessed by palpation. B-scan ultrasonography revealed a mass in the posterior segment with the characteristics of choroidal melanoma. An enucleation was performed and the globe submitted for ophthalmic pathology.

Macroscopically, the exterior of the globe was covered by a thick sheath of matted fibrous connective tissue. On opening the eye, there was a large mushroom-shaped tumour arising in the mid-choroid and almost filling the globe (Fig. 1). On microscopic examination, the tumour was found to be almost completely necrotic with the few viable areas composed of malignant spindle cells. Immunohistochemical staining with HMB45 and NK1-C3 confirmed their melanocytic nature. There was no evidence of extra-scleral spread. Examination of the remainder of the globe revealed angle closure due to anterior displacement of the lens and iris by the tumour mass in the posterior segment. There was severe conjunctivitis and episcleritis and anterior ciliary arteritis was present. Extensive necrosis of the iris, ciliary body and lens epithelium was noted. There was also necrosis of the limbal tissues and peripheral cornea, although the axial cornea remained viable. The retina, which had completely detached, had infarcted. An unusual feature present was infarction of the retinal pigment epithelium (Figs. 2, 3). Adjacent to this there was choriocapillaritis and occlusion of choroidal vessels by thrombosis (Fig. 4). The central retinal artery was arteritic and was thrombosed in the prelaminar portion (Fig. 5). The optic nerve was devoid of myelinated axons and was atrophic, presumably related to the childhood injury further details of which are not available.

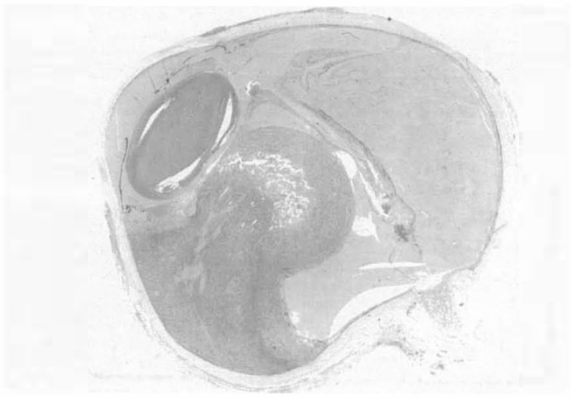

Fig. 1. *Low-power whole-mount section of the eye. There is a general pallor due to massive tissue infarction. A large mushroom-shaped choroidal tumour fills almost half the globe. (Haematoxylin & eosin, ×3.)*

Discussion

Focal necrosis within a choroidal melanoma is not uncommon[1] and, indeed, some melanomas undergo massive necrosis.[1] Extensive tumour necrosis may result in an inflammatory reaction in the intraocular structures and limited necrosis of intraocular stuctures can occur. As a result, signs of intraocular inflammation are not uncommon in cases of choroidal malignant melanoma,[2–5] particularly when the tumour is necrotic.[1,3] Orbital inflammation has also been described, more rarely, in association with uveal melanoma, even in the absence of extra-scleral tumour spread.[1–4,6,7] Once again, in such cases the tumour is at least partially necrotic.

This case further illustrates the presentation of a uveal malignant melanoma with the clinical symptoms and signs of predominantly orbital inflammation mimicking orbital cellulitis. Unusually, however, both the tumour and the other ocular tissues were almost totally infarcted. The extent of the infarction is very unusual and has never, to our knowledge, been described. The mechanism to account for this is probably multifactorial. One possible mechanism is that infarction is secondary to the mass effect of the tumour leading to angle closure, acute glaucoma and compromise of vascular supply by the raising of intraocular pressure above systemic blood pressure. However, intraocular pressure appeared normal, although this may be a misleading impression once ciliary body infarction was established. Alternatively, since there was massive necrosis of the tumour, cytotoxic products of tumour cell lysis via the associated inflammatory response may have diffused within the globe resulting in direct damage to other cells and, perhaps more importantly, vasculitis and thrombosis of intraocular vessels. Evidence of the latter mechanism is seen in this eye where vasculitis was observed in the anterior ciliary arteries, the central retinal artery and the choriocapillaris remote from the tumour. This may explain the unusual observation of infarction of both anterior and posterior segments including tissues such as the choroid, which has multiple vascular supplies via an anastomosing network.

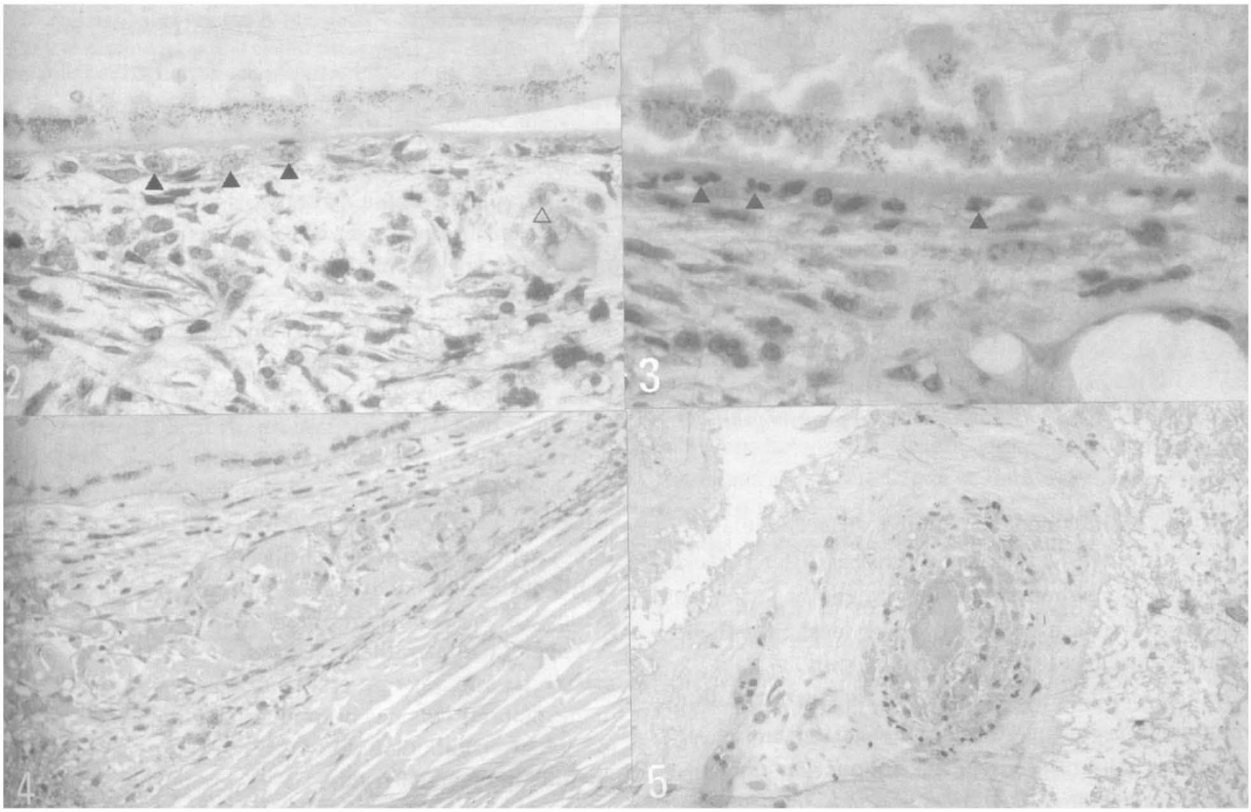

Fig. 2. *High-power micrograph of the inner choroid. The retinal pigment epithelium has infarcted and a proteinaceous exudate is present in the subretinal space. The patency of the choriocapillaris is compromised by endothelial cell swelling (black arrowheads). Similar endothelial swelling affects the other vessels of the choroid (open arrowheads) and there is interstitial oedema and mild chronic inflammation. (Haematoxylin & eosin, ×365.)* **Fig. 3.** *High-power micrograph of the inner choroid showing similar features to Fig. 2. In addition, there is active choriocapillaritis indicated by the presence of inflammatory cells, mostly neutrophil polymorphs (arrowheads). (Haematoxylin & eosin, ×550.)* **Fig. 4.** *Photomicrographs of the choroid showing dilatation and thrombosis of a large venule. (Haematoxylin & eosin, ×90.)* **Fig. 5.** *Photomicrograph of the prelaminar segment of the central retinal artery. There is active arteritis and thrombosis. (Haematoxylin & eosin, ×230.)*

References

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Sir,

An unusual presentation of primary sebaceous carcinoma of the conjunctiva

Sebaceous carcinoma of the conjunctiva is a rare tumour. It usually presents as a chronic inflammation of the conjunctiva and adjacent lid and, because of diagnostic delay, carries a poor prognosis. We would like to report an unusually localised presentation of conjunctival sebaceous carcinoma that has responded well to simple excision.

Case report

A 40-year-old man presented with a 3-week history of swelling of the right lower lid. There was a hyperaemic lesion in the right lower fornix and a provisional diagnosis of pyogenic granuloma was made. Hot spooning was recommended.

After 2 weeks he reported that the eye was sticky. The lesion was described as a 1 cm lump in the right lower