Necrotic orbital melanoma arising de novo

Jerry A Shields, Carol L Shields, Ralph C Eagle Jr, Patrick De Potter, Glen L Oliver

Abstract
A 76-year-old man with compressive optic neuropathy secondary to a retrobulbar mass was managed by orbitotomy and removal of the mass. The lesion proved histopathologically to be an unusual orbital melanoma with massive central necrosis. There was no histopathological evidence of congenital melanocytosis. Dermatological and systemic evaluation before and after orbital surgery revealed no evidence of primary melanoma elsewhere. The patient developed hepatic metastasis 2 years after excision of the orbital tumour. It appears that the melanoma was a primary orbital tumour and not a metastatic melanoma from an occult primary lesion.

Case report
A 76-year-old white male, who had no previous ocular problems except for mild amblyopia of the left eye, developed blurred vision in the right eye associated with epibulbar redness. An orbital computed tomography (CT) detected a retrobulbar mass. The initial clinical diagnosis was orbital inflammatory pseudotumour. After a 14-day course of oral corticosteroids failed to relieve his symptoms, the patient was referred to the ocular oncology service on 10 December 1990 for further evaluation and management.

The patient had a history of medically controlled hypertension, three previous myocardial infarctions, a prostatectomy for benign prostatic hypertrophy, and an inguinal herniorrhaphy. Two histopathologically confirmed seborrhoeic keratoses had been recently excised from his right scapular area. There was no history of ocular or cutaneous melanoma.

Our evaluation revealed best corrected visual acuities of 6/12 in the right eye and 6/21 in the amblyopic left eye. Intraocular pressures were normal. There was mild oedema of the right upper and lower eyelids and no proptosis. Ocular motility and colour plates were normal. Fundus examination of the right eye showed an elevated, hyperaemic optic disc and several juxtapapillary flame shaped haemorrhages. The left eye was normal except for decreased visual acuity due to amblyopia.

B-scan ultrasonography showed a rounded retrobulbar mass with acoustic hollowness and good sound transmission. CT revealed a 1.5 cm round, well circumscribed, intraconal mass abutting the globe and the optic nerve super temporally (Fig 1).

10.1136/bjo.77.3.187
Since the lesion was causing optic nerve compression, pain, and visual impairment, it was elected to excise the mass via a supertemporal conjunctival approach with temporary disinsertion of the lateral rectus muscle. Local anaesthesia was employed because of the patient's advanced cardiovascular disease. At the time of surgery, the round mass was identified but was tightly adherent to the sclera and optic nerve. Intraoperatively, a capsular rent developed and a small amount of yellow cheesy material extruded from the lesion.

**Pathology and follow up**

Microscopic examination revealed an extensively necrotic tumour (Fig 2) composed of spindle and pleomorphic epithelioid cells some of which contained intracytoplasmic melanin (Fig 3). Sheets of necrotic tumour cells occupied the centre of the lesion. The necrotic zone was rimmed peripherally by viable tumour cells which were immunoreactive for melanoma specific antigen (HMB-45) and S-100 protein. No evidence of congenital melanocytosis, blue naevus, or cellular blue naevus was found. The final histopathological diagnosis was orbital malignant melanoma.

The patient subsequently underwent another thorough systemic evaluation, including chest x ray, liver enzymes, dermatological evaluation, colonoscopy, and total body CT, all of which revealed no evidence of tumour. During the first few weeks after surgery, the patient's visual acuity in the right eye improved to 6/9 and the optic disc oedema completely resolved. The patient developed liver metastasis and died about 22 months after excision of the tumour.

**Comment**

Although ophthalmologists occasionally encounter cases of secondary orbital extension of uveal melanoma, primary malignant melanoma of the orbit is exceedingly rare. In the Wills Eye Hospital series of 645 biopsied orbital tumours, there was only one primary orbital melanoma. The Mayo Clinic series of 674 orbital tumours included only three cases of primary orbital melanoma. Primary orbital melanoma usually occurs in patients who have predisposing pigmentedary conditions such as congenital orbital melanocytosis, blue naevus, or cellular blue naevus. It has also developed in a patient who had received earlier orbital irradiation.

Perhaps the most puzzling aspect of the case reported here is whether the melanoma was a metastatic focus from an occult primary site or whether it was a primary orbital melanoma. Melanoma metastatic to the orbit almost always occurs in patients who have a known primary cutaneous melanoma, in whom it generally is a part of widespread melanomatosis. Affected patients are often in a near terminal state and their subsequent survival time is usually only a few weeks. The metastatic lesions usually contain epithelioid melanoma cells. Our patient had no history or findings of a primary melanoma and the necrotic portion of the tumour contained many spindle cells. Thus, we consider it unlikely

**Figure 1** Orbital computed tomogram showing well circumscribed retrobulbar mass compressing the anterior portion of the optic nerve.

**Figure 2** Low power photomicrograph showing rim of viable melanoma cells around a central area of necrotic tumour (haematoxylin and eosin ×15).

**Figure 3** Photomicrograph of viable tumour cells near peripheral portion of lesion showing spindle and epithelioid melanoma cells (haematoxylin and eosin ×63).
Late onset esotropia in monozygous twins

S Ahmed, J D H Young

Abstract
Monozygous twins who developed typical acute late onset esotropia are described. Neither had evidence of other neurological disease and both responded well to bimedial rectus muscle recessions. This twin presentation suggests a hereditary basis for the development of late onset esotropia in at least some cases. It provides further support for a policy of avoiding invasive CNS investigations in those patients who have binocular potential and are otherwise normal.

University of Dundee, Department of Ophthalmology, Ninewells Hospital and Medical School, Dundee DD1 9SY S Ahmed J D H Young

Correspondence to: Dr S Ahmed
Accepted for publication 9 December 1992

The sudden onset of strabismus in a previously normal child over the age of 4 is alarming to the parents. If there are no local aetiological factors it will also be of concern to the treating ophthalmologists aware that this might be the presenting sign of serious intracranial pathology. Typically strabismus secondary to neurological disease will be paralytic but concomitant strabismus is also a recognised presenting feature, especially of posterior fossa lesions.

Concomitant esotropia not associated with any detectable neurological disorder may also occur in this age group. This has been described as late onset or normosensorial esotropia and is characterised by:

1. An acute onset of esotropia with diplopia, in some cases initially intermittent for a short period.
2. A potential for normal binocular function, demonstrable on orthoptic testing.
3. No refractive error of relevance to the strabismus.

1.