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Necrotic orbital melanoma arising de novo

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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.

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Orbital malignant melanoma most often occurs from direct orbital extension of uveal, conjunctival, or eyelid melanoma.¹⁻⁴ Less often, orbital melanoma can occur as a metastasis from a previously diagnosed non-ocular melanoma. Primary orbital melanoma tends to occur in patients with predisposing melanocytic lesions such as congenital orbital melanocytosis or cellular blue naevus.¹⁻¹⁸ Primary orbital melanoma arising de novo without such pre-existing conditions is exceedingly rare. We report an unusual case of orbital melanoma that apparently developed as a primary orbital lesion in a patient who had no clinical or histopathological evidence of congenital orbital melanocytosis or cellular blue naevus. The lesion presented as a circumscribed orbital mass with extensive central necrosis.

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 superotemporally (Fig 1).

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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 or viable melanoma cells around a central area of necrotic tumour (haematoxylin and eosin $\times 15$).

Figure 3 Photomicrograph of viable tumour cells near peripheral portion of lesion showing spindle and epithelioid melanoma cells (haematoxylin and eosin $\times 63$).

Since the lesion was causing optic nerve compression, pain, and visual impairment, it was elected to excise the mass via a superotemporal 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.¹⁹ The Mayo Clinic series of 674 orbital tumours included only three cases of primary orbital melanoma.²⁰ Primary orbital melanoma usually occurs in patients who have predisposing pigmentary conditions such as congenital orbital melanocytosis, blue naevus, or cellular blue naevus.¹ It has also developed in a patient who had received earlier orbital irradiation.²¹

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

that our patient had melanoma metastatic to the orbit. Although it is rare for a primary orbital melanoma to arise without pre-existing congenital orbital melanocytosis, we believe that the patient reported here does represent a rare example of a primary orbital melanoma arising de novo, without clinical or histopathological evidence of pre-existing orbital melanocytosis or blue naevus.

Extensive necrosis of the central portion of the tumour in our case caused the lesion to have a cystic appearance on CT. The yellow cheesy material encountered at surgery suggested the diagnosis of dermoid cyst. However, orbital dermoid cysts usually occur in children in a superotemporal anterior location and may be attached to bone.²² They can occasionally occur in the orbital soft tissues, in which cases they are usually located on the nasal side and are lined by conjunctival epithelium.²²⁻²⁴

Based on the clinical findings, histopathological features, and follow up information, we believe that the lesion in our patient represents a highly unusual primary orbital melanoma.

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Late onset esotropia in monozygous twins

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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.

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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.¹

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 esotropia² 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.

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