Ocular melanocytosis associated with intracranial melanoma

EDITOR,—Ocular melanocytosis (melanosis oculi) is a rare congenital abnormality characterised by hyperpigmentation of the episclera and uveal tract, and caused by increased numbers of melanocytes.^{1,2} Unlike the naevus of Ota, in which the oral and nasal mucosae, palate, tympanic membranes, orbit, cranial bones, and leptomeninges can also be involved, ocular melanocytosis is confined to the eye.³ In this report, a patient with ocular melanocytosis who developed ipsilateral intracranial melanoma is described.

CASE REPORT

A 33-year-old right handed, fair skinned male patient was hospitalised because of left hemiparesis and right central facial palsy that progressed gradually in 3 months. Ophthalmic examination revealed his best corrected visual acuity to be 20/60 in the right and 20/20 in the left eye. The left eye was unremarkable. There were prominent patchy areas of episcleral pigmentation of the right eye (Fig 1). The iris and choroid were hyperchromic compared with the left eye. He also had mild exposure keratitis. There was no evidence of any degree of pigmentation in the right periorcular skin and in the visible areas of distribution of ophthalmic and maxillary divisions of the trigeminal nerve. Magnetic resonance imaging studies demonstrated a well defined, right sided, large intracranial mass close to the mesencephalon (Fig 2). The tumour was hyperintense compared with the adjacent tissues on T1 weighted images. The mass was removed surgically with the presumptive diagnosis of meningioma. However, histopathological examination of the tumour showed malignant melanoma with positive reactions to HMB-45 and S-100 antibodies. Careful systemic evaluation for a possible primary site was negative and the case was classified as primary intracranial melanoma. The patient then received whole brain radiotherapy.

COMMENT

Intracranial non-metastatic melanocytic tumours may sometimes be associated with cutaneous pigmented lesions. These lesions include neurocutaneous melanosis, cellular blue naevus, and the naevus of Ota.⁴ It is currently believed that the neural crest from which the melanocytes are derived, also contributes to the formation of meninges and the pia mater.^{4,5} There is evidence that malignant melanoma can develop in areas where abnormal melanocytes have been deposited during migration from the neural crest to the

Figure 2 Axial T1 weighted cranial magnetic resonance image demonstrating a well circumscribed, right sided intracranial tumour with increased signal intensity.

dermis.³ Leptomeningeal melanosis can be associated with these cutaneous lesions and is considered to predispose to intracranial melanomas.⁴

There is an established association between the naevus of Ota and melanomas of the iris, choroid, optic tract, or the orbit.^{1-4,6} Ocular melanocytosis on the other hand, classically known to affect the ocular structures only, has a 25% chance of developing intraocular melanoma.^{7,8} In these patients, melanoma may arise from the choroid or ciliary body but extremely rarely from the iris.¹ Melanocytoma of the optic nerve head can also be encountered.¹ Recently, cavernous haemangioma of the optic disc in a patient with ocular melanocytosis has been described.⁹ However, to the best of our knowledge, intracranial melanoma along with ocular melanocytosis has not been reported in the English language journals.

Our patient demonstrates that neurological signs and symptoms in the presence of ocular melanocytosis should alert the clinician to the possibility of a central nervous system melanoma. Moreover, ocular melanocytosis should be added to the list of pigmented skin and ocular disorders that may be associated with intracranial melanoma.

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Accepted for publication 12 July 1996

Figure 1 Right eye of the patient showing marked patchy episcleral pigmentation in all quadrants.

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Vitreous photocystotomy

EDITOR,—Pigmented free floating vitreous cysts are rare ophthalmic findings whose origins remain uncertain. They were first reported by Tansley in 1899. The cysts are often asymptomatic and found only during ophthalmic examination for other reasons. Occasionally, they can give rise to troublesome, intermittent blurring of vision, floaters, and shifting field defects. A number of treatment modalities have been proposed, including pars plana vitrectomy and argon laser photostotomomy.^{1,2} Each of these techniques has disadvantages. A case of symptomatic pigmented posterior vitreous cyst treated by Nd-YAG laser photodisruptive cystotomy is reported.

CASE REPORT

A 50-year-old man, a professional archer, presented to the vitreoretinal department with a 1 year history of a large floater, causing intermittent blurring of vision in the right eye. This was particularly disabling when he tilted his head to the right in order to aim to shoot. His corrected visual acuity was 6/6 in each eye and ophthalmoscopy revealed a large, round, free floating posterior vitreous cyst with glistening wall, flecked with surface pigment, in the right eye (Fig 1A). B scan ultrasound demonstrated a right sided, solitary, round 5.4 mm cyst situated near the retina within the synergetic posterior vitreous. A partial posterior vitreous detachment was present in the region corresponding to the cyst, although the posterior hyaloid face was mainly attached (Fig 1B). No other cysts were identified. There was no history of previous ocular trauma or inflammation.

In view of the symptoms, the patient underwent argon laser photostotomomy. A 200 mm spot at a setting of 100 ms and 400-600 mW was aimed at the anterior wall of the cyst through the central and peripheral mirrors of a coated three way mirror fundus lens. The laser caused blanching in the anterior cyst wall without cyst rupture and a photocoagulation reaction in the retina behind.

The patient subsequently underwent Q switched Nd-YAG laser (Coherent) photostotomomy at a setting of 0.6 mJ single pulse using a vitreous contact lens (LASAG CGV1). The beam was focused on the anterior cyst wall and three pulses were delivered, each puncturing the wall, causing immediate collapse, shrinkage, and gravitation of the cyst (Fig 2A). The symptoms were relieved immediately. At 2 months' follow up, the patient remained asymptomatic with a visual acuity of 6/6. Funduscopy showed immobile remnants of the cyst wall overlying the inferior retina and further B scan ultrasound documented a shrunken cyst measuring 1.6 × 2.2 mm (Fig 2B).

COMMENT

Primary vitreous cysts are rare and their origin is uncertain. They were previously presumed to be congenital remnants of the hyaloid

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