Ocular melanocytosis associated with intracranial melanoma

Editor.—Ocular melanocytosis (melanosoma oculi) is a rare congenital abnormality characterized by hyperpigmentation of the episclera and uveal tract, and caused by increased numbers of melanocytes. Unlike the naevus of Ota, in which the oral and nasal mucosa, palate, tympanic membranes, orbit, cranial bones, and leptomeninges can also be involved, ocular melanocytosis is confined to the eye. 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 periorificial 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.1 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.1 There is evidence that malignant melanoma can develop in areas where abnormal melanocytes have been deposited during migration from the neural crest to the dermis.2 Leptomeningeal melanosis can be associated with these cutaneous lesions and is considered to predispose to intracranial melanomas.3 There is an established association between the naevus of Ota and melanomas of the iris, choroid, optic tract, or the orbit.4,5 Ocular melanocytosis on the other hand, classically known to affect the ocular structures only, has a 25% chance of developing intraocular melanoma.6 In these patients, melanoma may arise from the choroid or ciliary body but extremely rarely from the iris.7 Melanocytoma of the optic nerve head can also be encountered.8 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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Figure 1 Right eye of the patient showing marked patchy episcleral pigmentation in all quadrants.

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

REFERENCES


Vitreous photocoagotomy

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 photocoagotomy.1 They have advantages and disadvantages. A case of symptomatic 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, the right eye (Fig 1A). B scan ultrasound demonstrated a right sided, solitary, round 5.4 mm cyst situated near the retina within the syneretic 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 photocoagotomy. 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 was 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) photocoagotomy 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 x 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
which the cyst was mobile within a small cavity in the posterior vitreous overlying the optic nerve and macula, highlighted the controversy over their pathogenesis. Orellana and colleagues reported on the microscopic appearance of a free floating vitreous cyst with its wall made up of a layer of heavily pigmented cuboidal cells, intermingled with non-pigmented cells, forming papillae. Electron microscopy showed the lining cells to contain mature and immature melanosomes, polarised basement membrane, and apical microvilli. These findings support the hypothesis that the cysts originate from the pigmented ciliary epithelium and that trauma may play a role in their development. Awan, however, reported a history of trauma in only 2.7% of cases.

The likelihood is that vitreous cysts originate from different intraocular structures, the vascularised, attached cysts from hyaloid vascular remnants and pigmented, free floating cysts from the ciliary body epithelium. Although the majority are asymptomatic, troublesome symptoms can arise when they float across the visual axis or come within its vicinity. In the case reported, the onset of symptoms may have been associated with increased mobility of the cyst due to liquefaction of the surrounding vitreous gel or partial posterior vitreous detachment.

The severity of symptoms occasionally warrants treatment. Surgical excision through the pars plana has been reported, but there is potential for serious complications from this approach. Argon laser photocoagotomy offers an alternative to surgical treatment, but its effectiveness depends on the presence of extensive pigment in the cyst wall and there is a risk of inadvertent retinal photocoagulation. Neodymium:YAG laser has previously been used for the treatment of persistent subclinical limiting membrane and posterior hyaloid face haemorrhages, vitreous floaters, vitreous adhesions, and for the lysis of vitreous bands. In the case described, Nd:YAG laser was effective in disrupting the wall of a posterior vitreous cyst. Although the cyst did not disappear completely, disruption of the cyst wall caused a reduction in its size. In addition, the cyst wall, being denser than the surrounding liquefied vitreous, gravitated out of the visual axis with relief of symptoms.

In conclusion, vitreous cysts, though rare, can give rise to intractable visual symptoms. Surgical treatment is hazardous and argon laser photocoagotomy may not be effective. We report the successful treatment of a posterior vitreous cyst by Nd:YAG laser photocoagotomy.

The authors thank Mr M Restori for carrying out the ultrasound examination.

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Sudden unilateral visual loss and brain infarction after autologous fat injection into nasolabial groove

Error—Central retinal artery occlusion (CRAO) following cosmetic surgery seems to be a very rare and devastating disease inducing sudden visual loss. Even if vigorous and massive treatment is advocated initially, the prognosis of visual recovery is very disappointing.

In this paper, we report one case of CRAO combined with brain infarction resulting from an autologous fat injection for cosmetic problems.

We confirmed CRAO by fluorescein angiography and brain infarction by magnetic resonance imaging (MRI) and four vessel angiography.

To our knowledge, there have been no reports of CRAO combined with brain infarction in autologous fat injection procedures.

This case gives a warning to cosmetic plastic surgeons and ophthalmologists of the importance of careful manipulation and immediate awareness and treatment of iatrogenically induced ocular complications.

CASE REPORT

A 42-year-old woman came to the emergency room in an irradiated state. Two hours earlier, she noticed a sudden loss of vision in the left eye.

The fundus examination revealed a complete absence of light perception in the left eye. The remaining globe was normal.

On examination, the patient was found to have a left CRAO with complete opacification of the left vitreous cavity. The left optic nerve head was pale, with no evidence of retinal or choroidal vessels.

An ophthalmic examination was performed, and the patient was referred to a neurologist for evaluation.

The diagnosis was confirmed by magnetic resonance imaging (MRI) of the brain, which showed a left-sided brain infarction.

The patient was treated conservatively with antithrombotic therapy and close monitoring of her neurological status.

The follow-up examination showed a gradual improvement in visual function over the next few weeks, with partial restoration of vision in the left eye.

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