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LETTERS TO THE JOURNAL

Sir,

Retinitis pigmentosa associated with Fuchs' heterochromic uveitis

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The term retinitis pigmentosa (RP) describes a group of hereditary retinal dystrophies, characterized by the early onset of night blindness and progressive loss of the visual field. Associated findings include myopia, optic disc drusen, posterior subcapsular cataracts, and cystoid macular oedema. In addition to these common findings, RP can on occasions be found in conjunction with other ophthalmic syndromes. One such syndrome very rarely linked to RP is Fuchs' heterochromic uveitis (FHU), which has so far been described in a total of 12 patients.[1–4] Of these 12 patients, only one had bilateral FHU (and an appearance of 'salt and pepper' in his fundi), the remainder being unilateral. More than 40 years following the first and only report of RP associated with bilateral FHU, we describe a second case.

Case report

A 38-year-old male was referred to the RP clinic by his optometrist who had noticed bone spicule pigmentation in both fundi. Over the previous 5 years, he admitted to having increasing difficulty with night vision, although he was not aware of any recent deterioration. Otherwise he was asymptomatic. He had a past ocular history of strabismic amblyopia affecting his left eye, for which he had received orthoptic treatment as a child. There was no family history of ocular disease and his past medical history was unremarkable.

On examination, with a correction for hypermetropia, his visual acuities were 6/12 and 6/36 in his right and left eyes, respectively. Slit-lamp examination demonstrated a low-grade anterior uveitis bilaterally with white 'stellate'-type cellular opacities on the corneal endothelium of both eyes (Figure 1a). There was no associated ciliary injection. Both irides had a greyish atrophic appearance with no evidence of posterior synechiae, and his intraocular pressures were normal. He had minimal posterior subcapsular lens opacities in his right eye but his left lens was clear. A low-grade inflammatory reaction in the anterior vitreous of both eyes was present, although there was no associated chorioretinitis, vasculitis, or pars planitis. However, fundal examination did show perivascular bone spicule pigmentation, bilateral optic disc pallor, and attenuated retinal vessels (Figure 1b). His Goldmann visual fields were markedly constricted in both eyes. The electroretinogram showed a reduction and delay in all rod and cone responses bilaterally. The multifocal electroretinogram confirmed a diffuse loss of retinal function in both eyes. These findings along with the fundal appearance confirmed the diagnosis of RP. The anterior segment signs were thought to be typical of FHU.

Comment

It is difficult to be certain of the mode of inheritance in our case, but this patient may well be a case of simplex, which in this country accounts for about 50% of patients.[5]

It is unclear whether the association of RP with FHU, whether unilateral or bilateral, is coincidental. A chronic low-grade inflammatory activity similar to that seen in

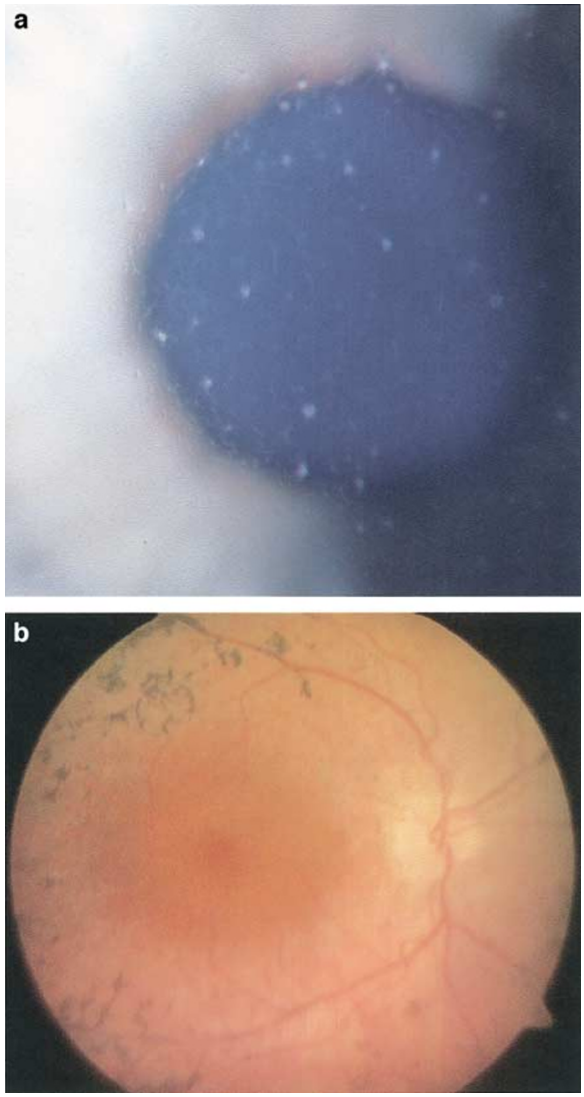

Figure 1 (a) Anterior segment photograph showing the 'stellate' corneal endothelial opacities. (b) Fundus photograph showing bone spicule pigmentation, pale disc, and attenuated retinal vessels—changes characteristic of RP.

FHU has been described in patients with RP. Recently, Chowers *et al*[1] showed that patients with RP can develop autoimmune reactions to anterior chamber antigens which bear some resemblance to retinal antigens, leading to the clinical manifestation of FHU.

It is well recognised that autoimmune reactions to retinal S antigen, a rod outer segment protein, have been implicated in the pathogenesis of various forms of uveitis and hereditary retinal degeneration. In addition, another report has demonstrated that some patients with RP have circulating B cells reactive with the retinal antigens, especially S antigen.[6] These suggest that the tendency for immune reactions in patients with RP could increase their susceptibility to develop FHU. It is important for clinicians to recognise that although it is extremely rare, bilateral FHU can be found in association with RP. This in turn may influence the management of complications such as low-grade uveitis in this select group of patients.

References

1 Chowers I, Zamir E, Banin E, Merin S. Retinitis pigmentosa associated with Fuchs' heterochromic uveitis. *Arch Ophthalmol* 2000; **118**: 800–802.
2 François J, Mastilovic B. L'hétérochromie de Fuchs associée aux hérédo-dégénérescences chorioétiniennes. *Ann Oculist* 1961; **194**: 385–396.
3 van den Born LI, van Schooneveld MJ, de Jong PT, Bleeker-Wagemakers EM *et al*. Fuchs' heterochromic uveitis associated with retinitis pigmentosa in a father and son. *Br J Ophthalmol* 1994; **78**: 504–505.
4 Vuorre I, Saari M, Tiilikainen A, Rasanen O *et al*. Fuchs' heterochromic cyclitis associated with retinitis pigmentosa: a family study. *Can J Ophthalmol* 1979; **14**: 10–16.
5 Jay M. On the heredity of retinitis pigmentosa. *Br J Ophthalmol* 1982; **66**: 405–416.
6 Reid DM, Campbell AM, Forrester JV. EB-virus transformed human lymphocytes from uveitis and retinitis pigmentosa patients secrete antibodies to retinal antigens. *J Clin Lab Immunol* 1988; **26**: 107–111.

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Sir,

Delayed acute angle closure after macular-hole surgery

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Intraocular pressure (IOP) elevation is a common complication of pars plana vitreous surgery.[1] Acute glaucoma may occur in the postoperative period secondary to angle closure caused by surgical adjuvants such as long-acting expansile gases, silicone oil, and perfluorocarbon liquids; by postoperative inflammation, and by oedema of the ciliary body and anterior rotation of ciliary processes.[1–4] Macular-hole surgery predisposes to angle closure because of prolonged prone positioning and the routine use of cycloplegic agents postoperatively. Since surgery to close a macular hole is a relatively recent innovation, to our knowledge only two cases of angle-closure glaucoma following this surgery have been recorded.[5] We report two further cases of acute angle-closure glaucoma following macular-hole surgery in patients with axial hypermetropia, in whom neither symptoms nor signs suggestive of angle closure existed preoperatively.

Case reports

Two hypermetropic women in their seventh and eighth decades of life were referred with a diagnosis of a unilateral macular hole (detailed clinical characteristics are summarized in Table 1). They had no past ocular or medical history of note and gave no history suggestive of intermittent angle closure. After dilatation with guttae tropicamide 1% and guttae phenylephrine 2.5%, the IOP was found to be within the normal range, and they did not experience any subsequent symptoms of angle closure. Preoperative gonioscopic examination was not undertaken in these patients as there was no obvious abnormality of the anterior chamber depth. They underwent a routine vitrectomy with epiretinal membrane and internal limiting membrane peel (under indocyanine green visualisation), followed by 15% perfluoropropane gas injection.

Postoperatively, guttae dexamethasone 0.1% four times daily, guttae cyclopentolate 1% twice daily, and guttae chloramphenicol four times daily were prescribed, and they were instructed to posture face-down for 50 min every hour for 2 weeks. On the first postoperative day, the IOP was normal in both patients, with no evidence of pupillary block. The anterior chamber was formed and there was a 70–90% gas fill. At 1 week postoperatively, both patients developed an acutely painful red eye and reduced vision. Examination revealed an inflamed eye with corneal oedema and an IOP of 45 mmHg in the first