Posterior scleritis with retinal vasculitis and choroidal and retinal infarction

N Andrew Frost, John M Sparrow, A Ralph Rosenthal

Posterior scleritis is a rare but probably under-diagnosed disease process. The clinical features are variable and the lesions may cause diagnostic confusion. Visual impairment is common but visual loss due directly to vascular involvement is unusual. In the case presented vascular involvement was exceptionally severe and determined the long term visual outcome.
Case report
A 34-year-old white woman presented with a 3 day history of constant pain in the right eye and right temporal region followed by visual deterioration. She had no symptoms of systemic illness and had no significant medical or family history. On examination there was uncertain perception of light in the right eye, with a right afferent pupillary defect. The right anterior sclera was obscured by gross conjunctival chemosis. A generalised restriction of eye movement was found together with mild proptosis (2 mm). Mild anterior uveitis was present. There was no vitreous haze. Intraocular pressures were 8 mm Hg (right) and 14 mm Hg (left). The right retina was widely oedematous and ischaemic with intraretinal haemorrhages, segmentation of the blood column within retinal vessels, and swollen optic disc (Fig 1). The left eye was normal. General physical examination was normal apart from mild pyrexia (37.4°C).

Investigations were performed as follows: full blood count, urea and electrolytes, calcium, liver enzymes, and urate were normal; plasma viscosity 1·77 cp; C-reactive protein 1·1 mg/dl; C3, C4 normal; ANA (hep-2) negative; ANCA negative; A-ENA negative; VDRL and TPHA negative; creatinine clearance normal; chest x ray normal. B mode ultrasound scan of the right eye and orbit showed scleral thickening at the posterior pole and retrobulbar oedema (Fig 2). The retrobulbar optic nerve shadow appeared normal. There was no evidence of other orbital pathology. Fluorescein angiography demonstrated retinal vasculitis with multiple branch retinal arterial occlusions and vessel staining together with segmental choroidal ischaemia (Fig 3).

A diagnosis of posterior scleritis was made and she was treated with high dose oral prednisolone, together with topical corticosteroids and atropine. The inflammation responded rapidly but the retina was extensively infarcted and the choroid was segmentally infarcted in several areas. The visual acuity remaining was 'hand movements' only. The systemic corticosteroid therapy was uneventfully reduced over approximately 5 months. At 16 months' follow up there was no evidence of recurrent local inflammation or systemic disease. The anterior sclera appeared normal but funduscopy revealed gross right optic atrophy with wedge shaped areas of choroidoretinal atrophy and many attenuated and non-perfused retinal vessels. Her visual acuity remained unchanged.

Comment
In this case the diagnosis of posterior scleritis was made clinically and supported by the ultrasonographic findings. The diagnosis has been further supported by the rapid response to corticosteroid therapy and the absence of other systemic or local orbital disease over 16 months of follow up. Posterior scleritis is frequently accompanied by systemic vasculitis but, surprisingly, visual impairment directly caused by vascular involvement is poorly documented. Four cases of choroidal vasculitis (without choroidal infarction), four cases with retinal vascular cuffing (without retinal vasculitis), and one case with subclinical retinal vasculitis, detected by fluorescein angiography were documented by Calthorpe et al.1 Juxtapapillary scleral inflammation suggesting direct extension to the central retinal vessels has been described.1 A case of posterior scleritis with bullous exudative retinal detachment followed by 'segmentary retinal phlebitis' at the posterior pole was described by Bonin.1 Intraretinal haemorrhages in the area of a
Ocular albinism with unilateral sectorial pigmentation in the fundus

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A diagnosis of ocular albinism is usually made in individuals with nystagmus, diminished visual acuity, reduced iris and fundus pigment, and foveal hypoplasia. Hypopigmamentation of the hair and skin is not found in people with ocular albinism.

We describe a Japanese woman who had ocular albinism with good visual acuity in the left eye and no nystagmus. Her right eye showed low visual acuity. Interestingly, sectorial pigmentation, which reached in part to the macula, was seen only in her left fundus. No pigmentation was noted in the right fundus. To the best of our knowledge, no reports have been written previously about patients with this type of asymmetrical ocular albinism and good visual acuity.

Case report
A 60-year-old Japanese woman was referred to our clinic on 15 March 1991, for an ophthalmic examination after retinal changes were discovered during a routine examination of her fundus. She had mild diabetes mellitus, no ocular complaints, and no hearing loss. She had been aware of her hypopigmented irides since childhood. Her parents had brown irides. Their marriage was not consanguineous. The patient's three siblings and two daughters had brown irides. No one in her family was reported to have had nystagmus or other eye diseases including hypopigmentation.

Examination showed the patient to have dark brown hair, brown eyebrows, and brown eyelashes. Her skin was not albinotic. No spontaneous nystagmus or strabismus were detected. Her visual acuity was 20/40 in her right eye, with a refractive correction of -7.75 D, and 20/20 in her left eye, without correction; however, no stereopsis was found with use of a Randot stereotest. Slit-lamp examination of the anterior segment showed bluish-grey irides with a brown
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