CASE REPORTS

Exudative retinal detachment and posterior scleritis associated with massive scleral thickening and calcification treated by scleral decompression

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Abstract

A 66-year-old man presented with massive bilateral scleral thickening and calcification associated with a unilateral exudative retinal

detachment which did not respond to systemic anti-inflammatory agents including steroids and cyclophosphamide but improved with scleral resection. This patient shows the features of both posterior scleritis and the uveal effusion syndrome, providing further evidence for the role of a thickened sclera and interference with the trans-scleral flow of fluid in the formation of such an exudative retinal detachment.

We report a case of bilateral massive scleral thickening and calcification associated with a unilateral exudative retinal detachment which responded to scleral resection. This resulted in an improved visual acuity which has been maintained for 4 years. This case shared many of the features of both posterior scleritis and the uveal effusion syndrome, and provides support for the hypothesis that the thickened abnormal sclera has a role in the formation of the subretinal fluid.

Figure 1A B-scan ultrasound of the right eye. The grossly thickened sclera measures 5.3 mm (between callipers).

Figure 1B B-scan ultrasound of the left eye. The sclera measures 4.7 mm (normal 1.1-1.8 mm).

Figure 2 Plain anteroposterior skull x ray. Dense shadowing of the calcified sclera in the orbits (arrowed) is seen.

Figure 3 CT scan shows gross scleral thickening of calcific density (large arrow). There is a shadow representing the exudative retinal detachment in the right eye (small arrow).

Case report

A 66-year-old man was referred to the scleritis clinic 3 months after noticing decreased visual acuity in both eyes, worse in the right. He had been seen 12 years previously with red sore eyes but no diagnosis had been made and no treatment given. His medical history was of confirmed osteoarthritis and previous surgery for peptic ulceration.

On ocular examination at presentation the visual acuity of the right eye was counting fingers and the left 6/18. Both eyes were red with episcleral vascular dilatation. The anterior sclera was visibly thickened with a cream coloured lumpy appearance most noticeable at the equator and tapering to the limbus. There was a slight aqueous flare in the right eye and a few cells in the anterior vitreous. The choroidal vascular pattern appeared normal. However the choroid was thickened by yellow nodular infiltrates and the retina showed marked clumping of pigment in both eyes. In the right eye there was an

exudative retinal detachment with shifting subretinal fluid.

Ultrasound showed a grossly thickened posterior sclera (Fig 1A and 1B). A ring of calcification was found on the plain skull x ray (Fig 2). These features were demonstrated both on the computed tomography (CT) scan (Fig 3) and the magnetic resonance image (MRI) (Fig 4).

Haematological investigation revealed a mild iron deficiency anaemia and a paraproteinaemia which was quantified showing IgG λ 17 g/l and IgA λ 12 g/l. Urinary Bence Jones proteins showed λ light chains. Bone marrow aspirates did not reveal evidence of myeloma.

An inflammatory aetiology was suspected and, after initial unsuccessful treatment with flurbiprofen, he was treated with pulsed methyl prednisolone 500 mg and cyclophosphamide 500 mg. Following this, treatment was continued with oral prednisolone 20 mg daily and cyclophosphamide 50 mg twice daily. Additional treatment with oral iron and cimetidine was given. One month later however the clinical findings were unchanged and the iron deficiency had resolved.

Surgical exploration of the right eye was undertaken (PGW). The vortex veins were identified as thread like and non-perfused. Two wedges of sclera were excised in the inferonasal and inferotemporal quadrants and the choroid was exposed.

The retina has remained flat since the operation except for a small area inferiorly which has not altered in 4 years. The visual acuity has been stable at 6/36 and 6/18, right and left respectively.

Histology of the resected material showed dense fibrous tissue with small zones of hyalinisation and calcification. There was some irregular inflammatory cell infiltrate consisting largely of lymphocytes and plasma cells (Fig 5A and B). There was no evidence of myeloma.

Discussion

This patient shared many of the features of posterior scleritis and the uveal effusion syndrome.

Posterior scleritis usually presents with severe unilateral pain and decreased vision. Thickening of the coats of the eye are seen in the active stage¹⁻⁵ on ultrasound. Other features are choroidal folds, optic disc swelling, retinal pigment epithelial changes, and exudative retinal detachments.³⁻⁶ It most frequently occurs in middle aged women¹⁷ when it is often associated with connective tissue disease or in young patients who are rarely found to have systemic disease. It may go unrecognised if it is pain free.^{8,9} The condition often responds to non-steroidal anti-inflammatory drugs (NSAIDs) or to systemic treatment with steroids and immunosuppressives.^{10,11} Management with surgery is not described.

The uveal effusion syndrome presents with a painless decrease in vision. An annular choroidal detachment with exudative retinal detachments are found on examination.^{2,12-14} This condition is bilateral but may be asymmetrical. The sclera is

Figure 4 Axial orbital MRI scan taken with a sagittal scan. The thickened sclera produces a low image (black arrow).

Figure 5A Low power histopathological section (H and E) showing dense fibrous tissue with small zones of hyalinisation and calcification.

frequently thickened^{15 16} and may be abnormal¹⁷ but inflammatory cells have not been found histologically.¹⁸ Anatomical variation or occlusion of the vortex veins is described. The uveal effusion syndrome typically occurs in healthy males. Resolution of the symptoms with systemic treatment including steroids is rare; however these patients respond to scleral surgery⁴ such as vortex vein decompression with scleral incisions and partial thickness sclerectomies.^{12 13 17 19 20}

An exudative retinal detachment may occur when normal movement of fluid from the sub-

retinal space to the suprachoroidal space and out of the eye via the vortex veins is disturbed.^{21 22}

The retinal pigment epithelium has a mechanism for pumping water but not colloid²³ resulting in a protein rich subretinal fluid. In the uveal effusion syndrome there is a high protein concentration in the subretinal fluid.²⁴

Brubaker and Pederson²⁵ emphasise the importance of scleral permeability to water and colloid in maintaining hydrostatic and osmotic gradients for the apposition of choroid to sclera. When ocular inflammation is present there is excess transudation of colloid from the choroidal

Figure 5B Higher powered view showing a few scattered inflammatory cells (arrow).

vasculature into the extracellular compartment which can only leave via bulk flow through the sclera.

Our patient has a 12 year history of red uncomfortable eyes. Chronic inflammation was found on histological sections with calcification which is likely to have occurred as a dystrophic event.²⁶

It appears that the physical signs observed in our patient were the result of several factors. There was inflammation leading to scleral thickening, calcification, and vortex vein occlusion with consequent inability to transfer fluid and colloid through the sclera. The abnormal pigment epithelium allowed colloid and fluid to be retained in the subretinal space. Immunosuppressive treatment was insufficient to reverse this. However scleral resection to allow trans-scleral flow of suprachoroidal and subretinal fluid was successful

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