CLINICAL AND EXPERIMENTAL
OPTOMETRY

CASE REPORT

Acute posterior multifocal placoid pigment epitheliopathy (APMPPE)

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A 20-year-old female presented with distorted vision after a viral illness and was found to have acute posterior multifocal placoid pigment epitheliopathy (APMPPE). This case is described with presenting signs and symptoms and the final outcome. The general features and aetiology of APMPPE are discussed.
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Key words: acute posterior multifocal placoid pigment epitheliopathy, retina

LC, a 20-year-old, presented with distorted vision in both eyes after a viral illness, the left eye being more affected. Visual acuities were right 6/6- and left 6/6=, the letters in the left eye appearing smaller and tilted. Pupil reactions were normal. Amsler charts showed macular distortion which was greater in the left eye than the right. There were no corneal or anterior chamber lesions visible with the slit lamp. LC also exhibited a 1.5 prism dioptre right hyperphoria, which gave her intermittent vertical diplopia, which she described as 'ghosting'. Mydriatic examination showed multiple deep placoid, cream-coloured areas involving the posterior pole. The left eye also showed pigment clumping (Figures 1 and 2).

The patient was referred for an ophthalmological opinion. Fluorescein angiography of the right eye showed areas of hypofluorescence suggesting choroidal non-perfusion (Figure 3). The diagnosis of acute posterior multifocal placoid pigment epitheliopathy was confirmed. No treatment was offered. The ophthalmologist found no evidence of fourth nerve

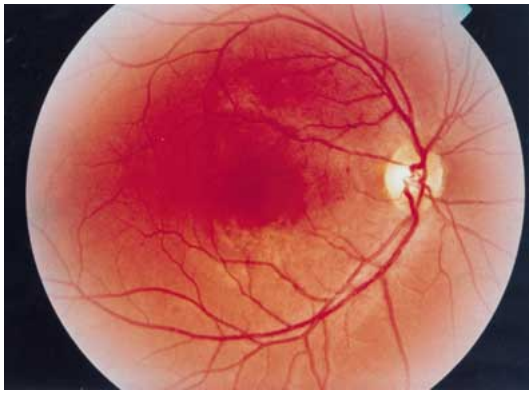

Figure 1. Right eye showing active areas of sub-retinal yellow-white placoid lesions at the posterior pole

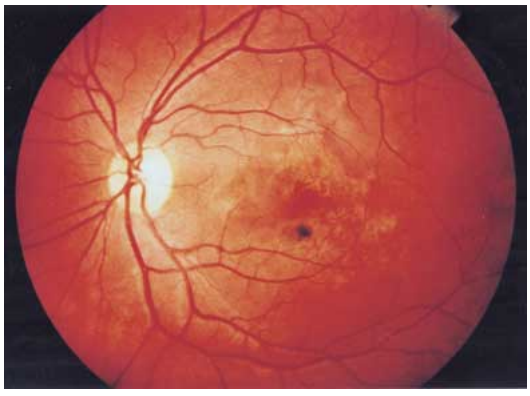

Figure 2. Left eye showing areas of pigment clumping and depigmentation as the placoid lesions resolve

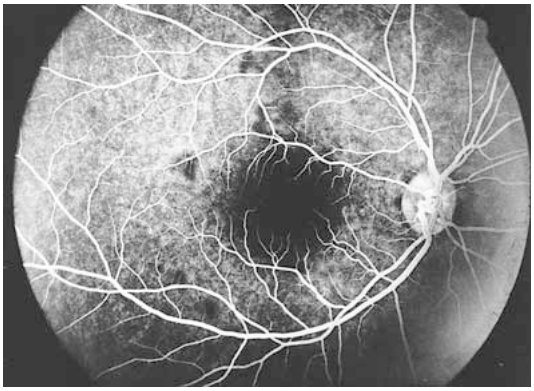

Figure 3. Right eye venous phase of fluorescein angiography showing areas of hypofluorescence suggesting choroidal non-perfusion

palsy. The phoria was thought to be a manifestation of a latent phoria.

Two years after onset visual acuities were still right 6/6- and left 6/6= and the phoria was still manifest. Prismatic correction was not offered as the ghosting caused by the phoria was not a significant problem. The distortion to the central field had not resolved completely. LC is now able to work full-time as a woodwork teacher and continues to drive a motor vehicle.

DISCUSSION

APMPPE is a rare idiopathic disease, which typically affects both eyes of a young adult of either sex. Approximately 50 per cent of patients have a prodromal influenza-like illness. The retinal pigment epithelium has been implicated as the primary site of involvement, although it has also been suggested that the disease might represent a 'vasculopathy' of the choriocapillaris.[1]

Symptoms

Patients may complain of blurred vision, metamorphopsia or scotoma.[2] While the decrease in visual acuity is usually dramatic, it can vary from 6/6 to count fingers. The disease is most often bilateral.

Signs

APMPPE was first described by Gass[3] in 1968 but the pathogenesis remains a mystery. An acute decrease in central vision in association with sub-retinal yellow-white placoid lesions of the posterior pole, followed by a spontaneous recovery is the typical presentation and course. Generally, the lesions are from 1/8 to 1/4 disc diameters, occasionally the lesions are confluent.[4] As the disease progresses, fresh patches arise more peripherally and may eventually involve the equator.[5] Hence, the lesions are often at different stages of healing. Sub-retinal neovascular membranes are an extremely rare complication of APMPPE.[6] Anterior segment manifestations include episcleritis, iritis and marginal thinning of the cornea.[7,8] The sub-retinal lesions spontaneously resolve in two to five weeks. Central healing is often followed by pigment clumping and depigmentation of the retinal pigment epithelium. Improvement in visual acuity often lags weeks to months behind the resolution of the lesions. Recurrences have been recognised but are not common.[9]

Fluorescein angiography

Fluorescein angiography of the placoid lesions is distinctive. Hypofluorescence is observed early in the angiogram, followed by hyperfluorescence in the late venous phase.[10] Early hypofluorescence is attributed to irregular filling of the choriocapillaris. The late hyperfluorescence is caused by leaking through the compromised cellular membrane of the blood-brain barrier.[4]

Aetiology

Despite extensive investigation, no aetiological factor has been identified.[10] However, using indocyanine green angiography, choroidal blood flow anomalies are present in APMPPE and this suggests that the clinical findings of this disease reflect a primary choroidal vascular disease.[11] Thus, the pigment epitheliopathy may be secondary to a multifocal choroidal vasculitis, which may have an immunological basis.[12] Each placoid area seen clinically may represent an area of focal swelling of the RPE overlying a non-perfused lobule of choriocapillaris.[10]

Therapy

No treatment has been identified as useful. However, corticosteroids have been used in numerous cases. Their efficacy has not been demonstrated[8,10] because evaluation of possible treatments is difficult due to the rare nature of the condition.

Prognosis

The visual prognosis is generally good.[2] Eighty per cent of affected eyes may have a final visual acuity of 6/12 or better.[13] However, a more recent study of 15 patients by Roberts and Mitchell[10] found visual acuities of 6/6 or better in 57 per cent of eyes, 6/9 to 6/18 in 14 per cent, 6/24 to 6/60 in 18 per cent and worse that 6/60 in 11 per cent of eyes. Most patients have residual symptoms and paracentral scotomata[2] and those with persistent central and paracentral scotomata and recurrent cases tend to have poorer visual outcomes.[8]

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