Acute Posterior Multifocal Placoid Pigment Epitheliopathy- A Case Report

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An 18 yr old male presented at the outpatient department with decreased vision in the right eye of seven days duration and blurring of vision in the left eye of three days duration. Visual loss in the right eye was rapidly progressing and was not associated with pain, redness or photophobia.

There was no history of floaters, flashes of light, scotomas, or coloured halos around the light or any associated micropsia, macropsia or metamorphopsia.

There was no relevant systemic or ocular illness in the past or any relevant ocular illness in the family. General examination and systemic examinations were normal. Best Corrected Visual acuity was 5/60 in the right eye and 6/18 in the left eye.

Slit lamp examination in the right eye showed few cells and strands in the anterior vitreous while the left eye showed only few cells, and no strands. Intraocular pressure was 18 and 16 in the right and left eye respectively.

Fundus flourescein angiography showed hypofluorescence in the choroidal and arterial phase due to blocked fluorescence. In the early venous phase there was leakage, and in the late phases there was staining corresponding to the lesions (Fig 3).

Ophthalmoscopic examination revealed multiple small subretinal yellowish-white, round, discrete, flat lesions <1/4th disc diameter in size scattered all over the posterior pole. More confluent lesions were present near the macula. Lesions were scattered temporal to the macula and nasal to the disc up to the midperiphery. The peripheral lesions were more oval and more ill defined. Associated retinal edema was present and the disc was normal.

Left eye also showed similar lesions in lesser numbers (Fig 1 & 2).

Field examination with Humphrey field analyzer revealed a paracentral scotoma corresponding to the lesions in the right eye while the field charting of the left eye was normal.

Laboratory evaluation revealed a normal blood and urine routine examination except ESR which was 40 mm/hr. ANA showed a positive result. Mantoux test
and Rheumatoid factor were negative. Serology for syphilis was negative. Chest X ray was normal. He was diagnosed as having Acute Multifocal Posterior Placoid Pigment Epitheliopathy [AMPPE] and treated with tapering dose of systemic steroids. Clinical improvement was noted in the right eye after 3 days of treatment, but the LE developed more lesions with worsening of vision initially and started showing improvement after one week (Fig 4). At the end of 2 weeks BCVA in the right eye was 6/18 and left eye was 6/12 and by the 4th week it improved to 6/9 in both eyes.

Fig. 4. Fundus picture after 1 week showing resolving lesions

Two months later the patient had vision of 6/6 both eyes and the only residual finding was a mild granularity of the right fovea (Fig 5).

**Discussion**

Originally described by Gass in 1968, acute posterior multifocal placoid pigment epitheliopathy (APMPPE), is a disorder characterized by the sudden appearance of multiple, yellow white, flat inflammatory lesions at the level of the retinal pigment epithelium and chorio capillaris. A flu-like prodrome consisting of fever, malaise and headache precedes most cases of APMPPE. This is followed by a sudden, usually bilateral, painless loss of vision. In patients with a monocular onset of symptoms, involvement of the fellow eye may occur within the following days to weeks. Central or paracentral scotomas may occur in patients with retinal lesions involving the foveal or parafoveal areas. Fundus examination reveals the characteristic multiple round, circumscribed, flat, yellow-white subretinal lesions involving the retinal pigment epithelium. As these lesions resolve over several weeks, vision improves in most cases to slightly less than initial acuity, and in some patients acuity may return to pre-onset levels. With time, fundus lesions are replaced by areas of depigmentation and pigment epithelial clumping. Additional ocular findings may include episcleritis, anterior uveitis, vitritis, retinal vasculitis, and papillitis. Associations with cerebral vasculitis, and erythema nodosum along with a host of immune-mediated disorders have been reported. For this reason, it is suggested that all patients with APMPPE should undergo a systemic and neurologic evaluation. Fluorescein angiography reveals characteristic changes during the evolution of the disease. During the acute, active stage of the disease, early films disclose areas of hypofluorescence in inflamed areas, secondary to RPE cell edema, leukocyte infiltration, and capillary nonperfusion. However, hyperfluorescence occurs in late films, as leakage occurs from the choriocapillaris through damaged RPE cells. During the inactive stage, as APMPPE lesions resolve, areas of hyperfluorescence occur at these sites secondary to RPE atrophy.
Indocyanine green angiography reveals areas of choroidal hypofluorescence during the acute stage of the disease, resulting from capillary non-perfusion, and these persist during the later stages of the disease.

Gass and colleagues suggest that inflammation begins at the level of the retinal pigment epithelium. Others, however, propose that the disorder primarily involves the choriocapillaris, and acute inflammation at this level occurs secondary to a hypersensitivity reaction to an external antigen, leading to occlusion of choroidal arterioles, ischemia, and secondary RPE changes.

Although the ocular disease has a self-limiting course, with approximately 80% of untreated patients having a visual acuity of 20/40 or better, 20% are left with impaired vision. Therefore all patients with APMPPE with macular involvement should be treated with systemic steroids. Use of systemic steroids rapidly resolves inflammation, and may result in a better final visual outcome.

References