CASE REPORT

Post Viral Acute Multifocal Posterior Placoid Pigment Epithiopathy in a Teenage Child

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SUMMARY

We report a rare case of a young boy presenting with bilateral blurring of vision following a viral like illness. Fundus examination revealed multiple pale cream-coloured lesions scattered across the posterior pole of both eyes. Fundus fluorescein angiography showed characteristic features of early hypofluorescence and late hyperfluorescence, further confirming the diagnosis of acute posterior placoid pigment epitheliopathy (AMPPPE). He was treated with topical steroids for the accompanying mild anterior uveitis. He had a prompt visual recovery with no adverse sequelae.

KEY WORDS:
Acute multifocal posterior placoid pigment epitheliopathy, AMPPPE, White dot syndromes

INTRODUCTION

Acute multifocal posterior placoid pigment epitheliopathy is classified under the white dot syndromes which are a group of disorders characterized by multiple whitish-yellow inflammatory lesions located at the level of the outer retina, retinal pigment epithelium and choroid. It has been noted to have diverse manifestations ranging from mild transient visual loss to life-threatening systemic complications. This case illustrates a milder but more typical form of the disease.

CASE REPORT

A 14 year-old Sikh boy presented with a week's history of bilateral simultaneous gradual and painless blurring of vision. However, this affected only his distant vision. His ocular symptoms were preceded by conjunctivitis and a flu-like illness, comprising of cough and rhinitis. He denied any bowel symptoms or oral ulcers. There was no other significant history of note.

His best corrected visual acuity was 6/9 in both eyes. The intraocular pressure was 12mmHg bilaterally. There was mild anterior chamber activity bilaterally with no evidence of vitritis. However, multiple pale cream-coloured lesions were noted at the level of the retinal pigment epithelium (RPE) bilaterally with a lesion adjacent to the left fovea (Figure 1). The right macula was fortunately spared. The optic discs were normal and there was no vasculitis. Amsler tests were normal bilaterally. FBC was normal and ESR was not raised. A provisional diagnosis of acute multifocal posterior placoid pigment epitheliopathy was made.

A fundus fluorescein angiography revealed early hypofluorescence and late hyperfluorescence (Figures 2a-d), further confirming the diagnosis.

He was prescribed topical steroids for the anterior uveitis. In view of good vision and sparing of the macula, systemic corticosteroid was not commenced. A week later, his vision improved to 6/6 bilaterally and the anterior uveitis resolved. The RPE lesions appeared to have reduced in number, coalesced and more hyperpigmented. However, his vision was maintained at 6/6 throughout his follow-up visits up until a year later. He is still under follow-up in view of subsequent potential complications.

DISCUSSION

In 1968, Gass first described the clinical and angiographic findings in three young women who presented with the typical findings of what we know today as acute multifocal posterior placoid pigment epitheliopathy (AMPPPE)[1].

AMPPPE has a striking preponderance for young healthy adults between the ages of 20 and 50 years[2]. This patient presented with an antecedent viral illness which is present in one-third of affected individuals as reported by Gass[1]. He did not have any other systemic manifestations associated with AMPPPE such as mumps, sarcoidosis, Wegener's granulomatosis, polyarteritis nodosa, ulcerative colitis, Group A streptococcal infection, tuberculosis and Lyme disease[2]. Death has been reported but rare and is usually associated with central nervous system involvement[3].

AMPPPE presents with blurring of vision, scotomas or metamorphopsia, if the fovea is involved. It is usually bilateral, occurring simultaneously as in this case or sequentially. Patients typically present with bilateral, multifocal yellowish-white placoid lesions at the level of RPE. The lesions fade gradually over one to two weeks to reveal varying degrees of RPE atrophy and hyperpigmentation. Visual loss is usually due to foveal involvement. In this patient, his vision was preserved as the lesions had missed both his fovea. Other diverse ocular manifestations have been observed such as papillitis, episcleritis, anterior and posterior uveitis. Anterior uveitis was present in this case.

The aetiology is not well understood but has often been attributed to postviral infection as illustrated in this boy. Park and associates proposed delayed hypersensitivity as a possible cause of AMPPPE[4]. Diagnosis is largely clinical based

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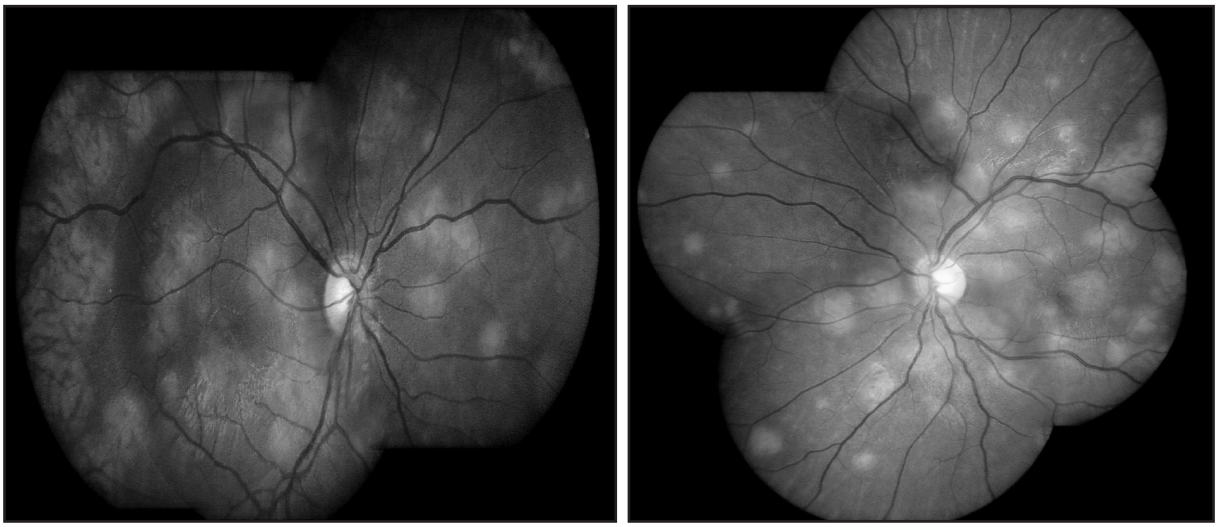
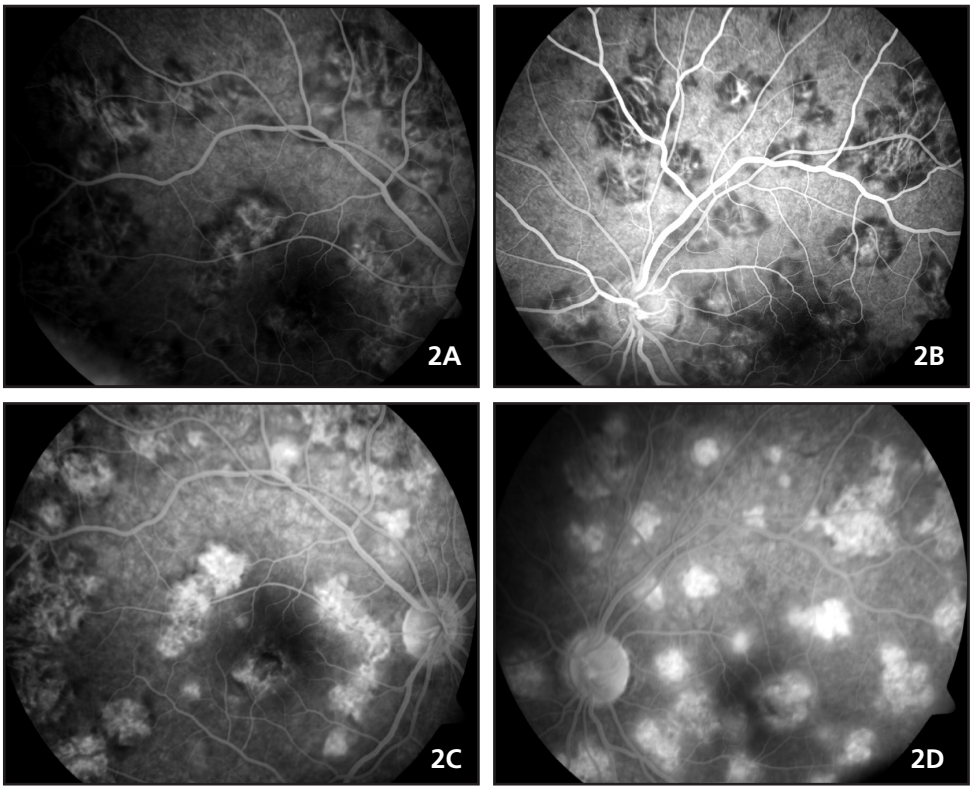


Fig. 1: Multiple pale cream-coloured lesions affecting both retina with a lesion just adjacent to the left fovea.



Figures 2a,b,c and d: Fundus fluorescein angiography of both the retina revealing early hypofluorescence (Fig 2a and b) and late hyperfluorescence (Fig 2c and d).

on typical clinical presentation and fundus fluorescein angiographic findings. The characteristic fundus fluorescein angiographic findings of early hypofluorescence and late hyperfluorescence were seen in this case. Indocyanine green angiography reveals hypofluorescence of both the acute and healed lesions highlighting the role of choroidal vascular occlusion in AMPPPE[4]. This further supports the notion that AMPPPE is a primary choroidal vasculitis with secondary involvement of the pigment epithelium.

Most patients have good visual prognosis with spontaneous recovery within 3 to 6 weeks, but in this case visual recovery was seen as early as one week.

In general, no treatment is required. However, corticosteroid has been suggested in cases with foveal involvement[5] and central nervous system manifestations. Immunosuppressive therapy has also been proposed in the latter group. This patient had excellent visual recovery despite the residual hyperpigmented retinal scars. However, he still needs to be

followed up as recurrence and choroidal neovascularization, a dreaded complication has been seen but rare.

CONCLUSION

AMPPPE although rare can be the cause for a post viral blurring of vision in the paediatric age group. Fortunately, the disease is self-limiting with good visual prognosis provided that the macula is not involved.

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