

A rare case of unilateral acute posterior multifocal placoid pigment epitheliopathy with features of Vogt-Koyanagi-Harada disease



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ABSTRACT

The purpose of this study is to report a rare case of unilateral acute posterior multifocal placoid pigment epitheliopathy (APMPPE) with features of Vogt-Koyanagi-Harada (VKH) disease. Both the diseases have their own unique presentations and can lead to vision loss in the affected eye. Overlapping features of both diseases in the same eye is rare. A 28-year-old healthy female presented with loss of vision in her right eye for 10 days duration. Visual acuity in the affected eye was 20/200, N36. Fundus examination revealed numerous creamy yellow lesions overlying the posterior pole with subretinal fluid. Further investigations including fundus fluorescein angiography and optical coherence tomography were suggestive of overlapping features of both APMPPE and VKH disease. Baseline laboratory investigations and markers for Sarcoidosis, Tuberculosis, and Syphilis were normal. The patient was treated with tapering doses of oral steroids. One month following initiation of treatment, the vision had improved to 20/20, N6 with complete resolution of subretinal fluid in the right eye. The overlapping clinical and imaging features suggest that both diseases may be a part of a common inflammatory process that secondarily damages the outer retinal structures.

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1. Introduction

Acute posterior multifocal placoid pigment epitheliopathy (APMPPE) was first described by Gass (1968). It is a self-limiting chorioretinal inflammatory disorder typically affecting healthy young adults. It is characterized by rapid loss of central vision and multiple yellowish-white deep lesions at the level of the retinal pigment epithelium. Vogt-Koyanagi-Harada (VKH) disease is a bilateral granulomatous panuveitis presenting with disc hyperemia/edema, exudative retinal detachment, sunset glow fundus, and potential involvement of auditory, neurological, and integumentary systems (Moorthy et al., 1995). We report a very rare case of unilateral APMPPE with features of VKH disease.

2. Case report

A 28-year-old healthy female presented with a sudden loss of vision in her right eye (RE) of 10 days

duration. It was associated with throbbing pain in the right periorbital area. The patient complained of flu-like symptoms of fever, malaise, and fatigue 1 week prior to the onset of ocular symptoms. At the initial visit, the best-corrected visual acuity (BCVA) in the RE was 20/200, N36 and 20/20, N6 in the left eye (LE). Anterior segment examination and intraocular pressure were within normal limits in both eyes. Funduscopy with slit-lamp biomicroscopy with 78D lens and indirect ophthalmoscopy showed numerous placoid-shaped creamy yellow lesions in the posterior pole and around the vascular arcades superiorly and inferiorly. Fig. 1A represents a color fundus photograph of the right eye showing numerous yellow placoid lesions in the posterior pole and subretinal fluid in the macular area. Few choroidal striae were noted around the macular area. Funduscopy of the LE was normal.

Fundus fluorescein angiography (FFA) was performed. Fig. 1B is the early phase FFA image of RE showing multiple areas of hypofluorescence corresponding to the placoid lesions seen on funduscopy. Fig. 1C is the late phase FFA image of RE showing increasing hyperfluorescence (staining) of the lesions as the angiography proceeded. The FFA study of LE was normal. Spectral-Domain Optical coherence tomography (SDOCT) of the RE showed a

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large area of neurosensory detachment in the RE. Fig. 1D is an SDOCT scan of RE depicting neurosensory retinal detachment with septa like structure within the detachment. There was gross disorganization of the external limiting membrane, ellipsoid zone, and photoreceptor zone. SDOCT of the LE was normal. The clinical and FFA findings were suggestive of APMPE but SDOCT was more in favor of VKH disease.

Baseline laboratory investigations and markers for Sarcoidosis, Tuberculosis, and Syphilis were normal. The patient was treated with oral Prednisolone 60 mg (1mg/kg body weight), oral ranitidine (150mg/day), and oral calcium (500mg/day). One week after initiation of treatment, there was no improvement in vision in RE. The throbbing periorbital pain had reduced. The placoid

lesions were slightly smaller in size but there was no reduction in the subretinal fluid in the macular area. At 2 weeks, the vision had improved to 20/32, N8 and there was a significant reduction in the subretinal fluid at the macular area. Oral Prednisolone was slowly tapered at 10 mg/week after the 2nd week. At 1 month follow up, vision in RE had improved to 20/20, N6. Fig. 1E and Fig. 1F is a color fundus photograph of RE 1 month following presentation showing complete resolution of all placoid lesions. Fig. 1F is an SDOCT scan of RE depicting complete resolution of subretinal fluid, 1 month following the presentation. The structural integrity of the outer retina was well maintained. Oral steroids were tapered. The vision was maintained at 20/20, N6 at 6 months follow up.

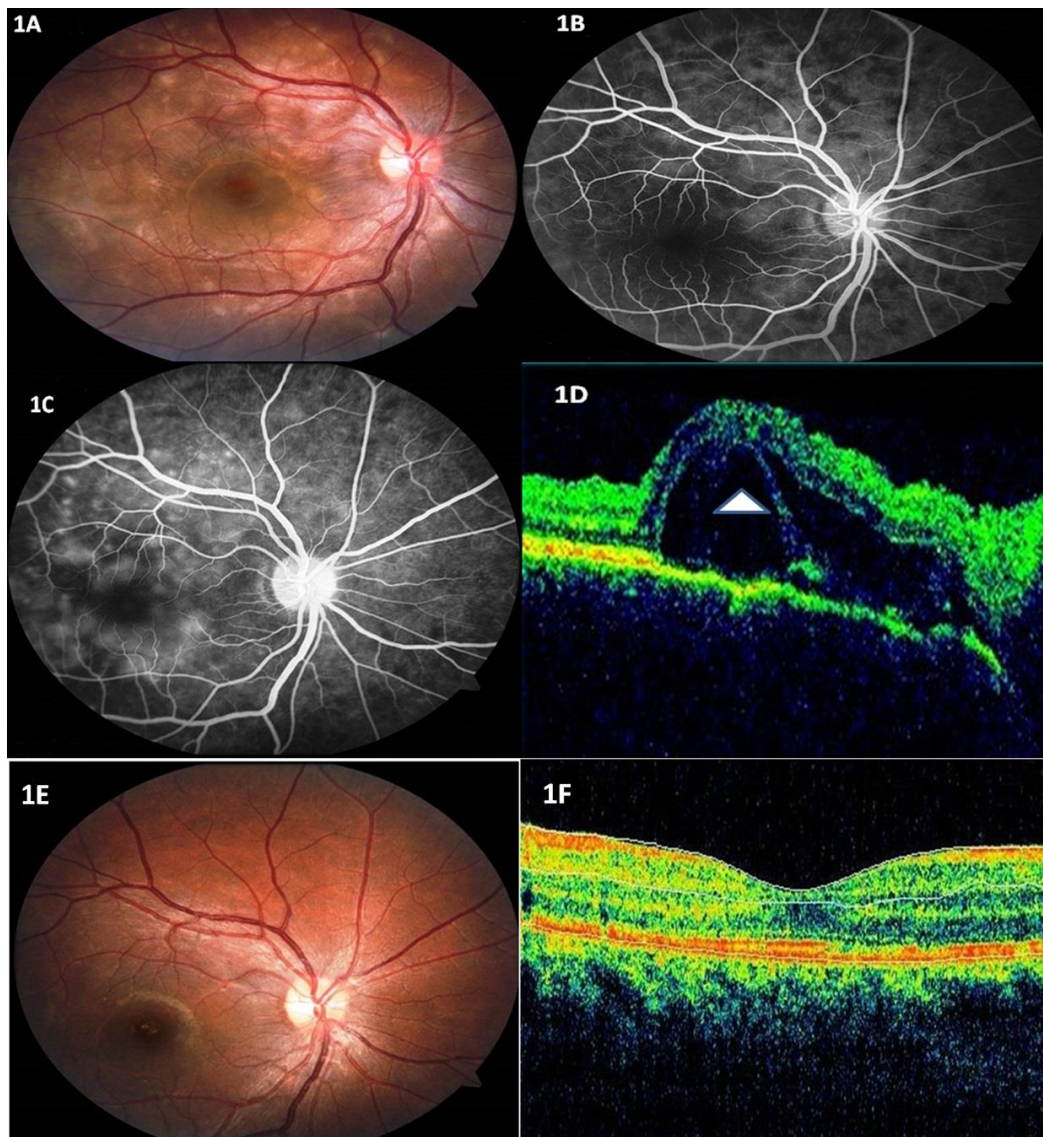


Fig. 1: Fig. 1A: Colour fundus photograph of the right eye showing numerous yellow placoid lesions in the posterior pole and subretinal fluid in the macular area; Fig. 1B: Early phase of Fundus fluorescein angiography showing multiple hypofluorescent patches corresponding to the placoid lesions; Fig. 1C: Late phase of Fundus fluorescein angiography showing increasing hyperfluorescence (staining) of the lesions; Fig. 1D: Spectral-domain optical coherence tomography showing a large neurosensory detachment of right eye. A septum (white arrowhead) can be seen within the detachment-Gross distortion of outer retinal layers noted; Fig. 1E: Colour fundus photograph of the right eye showing complete resolution of all placoid lesions and macular subretinal fluid 1 month after initial presentation; Fig. 1F: Spectral-domain optical coherence tomography of the right eye showing well preserved outer retina and complete resolution of neurosensory detachment

3. Discussion and conclusion

APMPPE is usually bilateral disease. If unilateral, the other eye can get involved within a few weeks. FFA findings include early because of blockage of underlying choroidal hyperfluorescence. Late phases of the angiogram show progressive irregular staining of the lesions (Gass, 1968). VKH disease is also bilateral disease. Unilateral VKH is a very rare entity. Angiographic features of VKH disease include early pinpoint hyperfluorescence with increasing leakage and pooling of the dye in the subretinal space in the late phase. OCT shows large pockets of neurosensory detachment in VKH disease.

This case has typical and atypical features of both APMPPE and VKH disease. The disease was unilateral. There was no involvement of the second eye even at 6 month follow up. The rapid onset of central vision loss, the placoid lesions are seen on funduscopy and angiographic features of early hypofluorescence and late hyperfluorescence are characteristic of APMPPE. The pockets of subretinal fluid seen on funduscopy and the OCT findings are more commonly seen in VKH disease.

Li et al have described a similar case of unilateral and spontaneously resolving posterior uveitis with overlapping features of APMPPE and VKH disease (Li et al., 2016). Vedantham and Ramasamy (2006) have reported 4 cases of unilateral APMPPE with atypical features of neurosensory retinal detachment, retinal vasculitis, and papillitis.

This is a report of a very rare case of unilateral posterior uveitis with features of both APMPPE and

VKH disease. This case adds to the body of literature available on atypical APMPPE and VKH disease. The overlapping clinical and imaging features suggest that both diseases may be a part of a common inflammatory process that secondarily damages the outer retinal structures.

Compliance with ethical standards

Conflict of interest

The author(s) declared no potential conflicts of interest with respect to the research, authorship, and/or publication of this article.

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