

# Acute Posterior Multifocal Placoid Pigmented Epitheliopathy

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## Introduction

White dot syndrome (WDS) is a group of ocular disorders characterized by the presence of discrete or converging white lesions located at various levels of the retina depending on the class of WDS condition. Acute Posterior Multifocal Placoid Pigment Epitheliopathy (APMPPE), described for the first time by Gass in 1968, is a rare inflammatory eye disease characterized by the emergence of discrete but occasionally converging placoid injuries, creamy white-yellowish in color, which involve the choriocapillaris, outer retina, retinal pigment epithelium (RPE) and is preferably located at the posterior pole.<sup>1</sup> The incidence and prevalence of APMPPE is unknown. However, since the landmark description of Gass, APMPPE has been reported frequently and widely from ophthalmic centers, primarily in the United States and Western Europe. According to a study in Minnesota, the incidence over 20 years from 1998 to 2008 was found to be 0.15 per 100,000.<sup>2</sup> Men and women are equally affected with a range from 8 to 66 years, most commonly between the ages of 20 to 30 years in otherwise healthy adults, and is thought to occur more commonly in Caucasians.<sup>3</sup> It is associated with a viral prodrome in over one third of the cases.<sup>1</sup> The condition is bilateral in 75% of the cases.<sup>4</sup> If one eye is affected, the other eye will follow only days later or sometimes delayed by several weeks.<sup>5</sup> APMPPE is considered to have a favorable prognosis compared to other WDS. In general, visual symptoms improve in 2-4 weeks. Approximately 50% of patients completely recover their vision; 25% have 20/40 vision or worse; and 60% have residual visual symptoms.<sup>6</sup>

## Introduction

A 31-year-old male presented to the eye clinic after complaining of seeing white floaters and metamorphopsia in his left eye developing 2 months after flu like symptoms caused by a viral upper respiratory tract infection. No significant clinical findings were found at the time of presentation. Optical Coherence Tomography (OCT) showed very mild sub-retinal fluid (SRF), and retinal examination revealed multiple white yellowish lesions limited to the posterior pole. Anterior segment examination revealed no evidence of abnormalities. His lens were clear, cornea and anterior chamber were within normal limits. 9 days later, the amount of SRF increased significantly that resulted in his VA to drop to CF. (Fig. 1) Dilated exam showed multiple para-central white lesions slightly elevated with disk hyperemia in the left eye. (Fig. 2) The right eye was unremarkable. There was no evidence of vitritis in both eyes. FA showed early hypofluorescence corresponding to blockage by the lesions and late hyperfluorescence staining. (Fig. 3) The clinical findings together with the patient's symptoms were suggestive of WDS. Initial investigations were done to rule out sarcoidosis and infectious causes like syphilis and tuberculosis. A PPD test was positive; however, his chest X-ray was negative. With the help of OCT and FA, diagnosis of APMPPE was established.

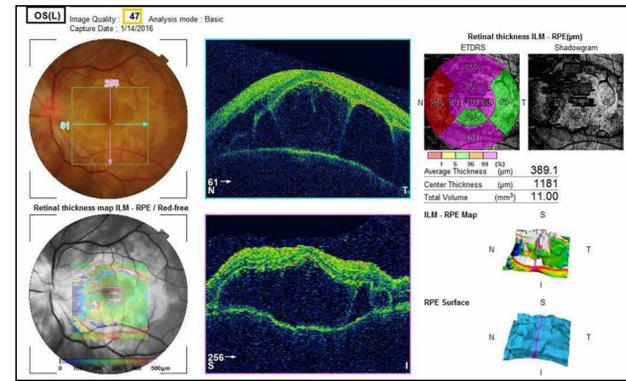


Figure. 1 Optical Coherence Tomography shows large sub-retinal fluid and the visual acuity has dropped to counting fingers.

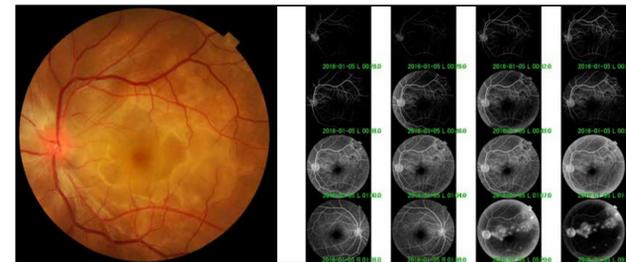


Figure. 2 (LEFT) Fundus photograph shows multifocal confluent creamy lesions with indistinct borders involving the macular of the left eye.

Figure. 3 (RIGHT) Fluorescein angiography shows early blockage of the sub-retinal lesion with late staining in the left eye. The right eye is normal. This change from early hypofluorescence to late hyperfluorescence of the lesions is characteristic of APMPPE.

## Discussion

Both infectious and noninfectious inflammatory diseases can present with multifocal chorioretinal lesions. The entities included in the WDS share some features, which make them a particular group of ocular disorders that include the following diseases:

- Acute posterior multifocal placoid pigment epitheliopathy (APMPPE)
- Serpiginous choroidopathy
- Multiple evanescent white dot syndrome (MEWDS)
- Diffuse unilateral subacute neuroretinitis
- Birdshot retinochoroidopathy
- Multifocal choroiditis and panuveitis syndrome (MCP)
- Punctuate inner choroidopathy (PIC)
- Acute zonal occult outer retinopathy (AZOOR)
- Acute Idiopathic Exudative Polymorphous Vitelliform Maculopathy

It is still an area of controversy whether white dot syndrome is a spectrum of the same disease process or has separate disease entities.<sup>7</sup> According to a study in Minnesota, the incidence of APMPPE over 20 years, 1998 to 2008, was found to be 0.15 per 100,000.<sup>2</sup> Due to the rarity of the disease and insufficient collected data, accurate

estimates of the incidence and prevalence of the disease are still unavailable. It is associated with a viral prodrome in over one third of the cases.<sup>1</sup> The condition is bilateral in 75% of the cases.<sup>4</sup> If one eye is affected, the other eye will follow only days later or sometimes delayed by several weeks.<sup>5</sup> There is no consensus on treatment; however, steroids have been attempted to hasten visual recovery especially in cases with macular involvement. The disease is self-limiting in most cases but has the risk if left untreated of scarring and macular folding resulting in permanent vision loss. Because of the nature of the vision loss in this patient, he was prescribed with prednisolone starting with 60 mgs orally and tapered over 6 weeks after which the VA resolved to 20/20. (Fig. 4) Unfortunately, after that, the patient had a small attack away from the macula with fluid collection. Re-treatment was started with a smaller dose aiming to go gradual with more prolongation in steroid cessation. The amount of fluid in the OCT decreased until the patient fully recovered. His VA has resolved to 20/20.

Diagnosing APMPPE can be a challenge due to the rarity of the disease and its non-specific presenting symptoms. Common presenting symptoms include flashes, blurred vision, floaters, and visual field loss. The etiology is unknown and typically affects young adults. The temporary symptoms and the frequent associations with previous prodromal viral infections (especially of the upper respiratory tract) suggest a potential inflammatory etiology. Fluorescein angiogram show early hypofluorescence with late staining of the sub-retinal lesions. This suggests that choroidal ischemia caused by choroidal vasculitis as the disease's main mechanism. This is supported by APMPPE association with various types of systemic vasculitis such as Wegener's granulomatosis, sarcoidosis, cerebral vasculitis,<sup>8</sup> and other conditions that cause stimulation of the immune system due to infections including tuberculosis<sup>9</sup> or due reactions to vaccination such as varicella.<sup>10</sup> These conditions are strong evidence supporting the hypothesis that APMPPE is caused by a delayed type IV hypersensitivity reaction. The response to steroid treatment can vary widely from complete resolution to multiple relapses. This patient demonstrated excellent response to the steroid treatment with no residual symptoms.

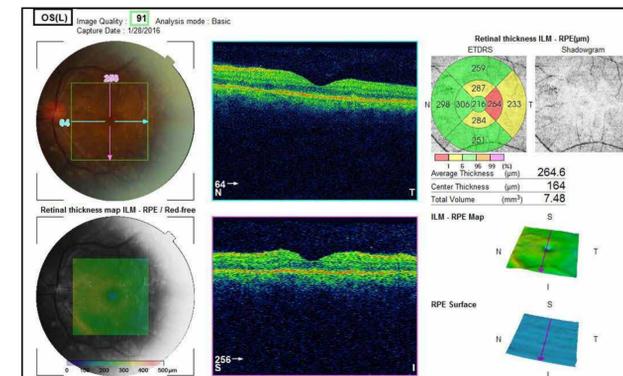


Figure. 4 Optical Coherence Tomography shows complete resolution of the sub-retinal lesions and the visual acuity of the left eye is back to 20/20 within 6 weeks of steroid treatment.

## Conclusion

Further studies are needed to assess the prevalence and etiology of this disease as it has been linked with prodromal viral infection and tuberculosis as previously described in which our patient was exposed to both. Early presentation and successful diagnosis are the key factors toward successful management of APMPPE.

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