

# Acute Posterior Multifocal Placoid Pigment Epitheliopathy: Association with Granulomatous Anterior Uveitis: Case Report

## Akut Posterior Multifokal Pigment Epitelyopatisinin Granülomatöz Anterior Üveit ile Olan İlişkisi

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Geliş Tarihi/Received: 30.12.2009  
Kabul Tarihi/Accepted: 10.02.2011

*This case report was presented as a poster in Turkish Ophthalmology Society 42<sup>nd</sup> National Ophthalmology Congress, 19-23 November 2008, Antalya, Turkey.*

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**ABSTRACT** In this paper, we report a 43-year-old male patient presented with sudden decreased vision of both eyes. His visual acuity was 20/125 in the right and 20/400 in the left eye. A complete ophthalmologic examination was performed. Examination of fundus revealed multiple placoid, yellow-white lesions located deep in the retina of both eyes. The fluorescein angiogram showed early hypofluorescence and late hyperfluorescence of the lesions, typical of acute posterior multifocal placoid pigment epitheliopathy (APMPPE). Four days later, anterior chamber showed 2+ cells, mild flare, mutton-fat keratic precipitates on the corneal endothelium in both eyes and posterior synechia in the left eye. Radiographic examinations and laboratory tests were normal. Treatment with topical corticosteroids, cyclopentolate drop and oral prednisolone were administered. After six weeks, the patient's symptoms regressed and the visual acuities returned to 20/20. Treatment is not necessary for most cases of typical APMPPE; however, systemic and topical corticosteroids and/or cycloplegics may be useful for cases with atypical clinical features.

**Key Words:** Granulomatous disease, chronic; panuveitis; hydrocorticosteroids; choroid diseases

**ÖZET** Bu yazıda her iki gözde ani görme azalması ile gelen 43 yaşındaki bir erkek hastayı sunmaktayız. Görme keskinliği sağ gözde 20/125 ve sol gözde 20/400 idi. Tam bir göz muayenesi yapıldı. Fundus muayenesinde her iki göz retinasının derininde çok sayıda plakoid, sarı-beyaz lezyonlar görüldü. Flöresein anjiyografide lezyonlarda akut posterior multifokal plakoid pigment epitelyopati (APMPPE) açısından tipik erken hipofloresans ve geç hiperfloresans görüldü. Dört gün sonra, her iki gözde ön kamarada +2 hücre, protein sızıntısına bağlı hafif bulanıklık, kornea endotelinde koyun yağı keratik presipitatlar, sol gözde posterior sineşi görüldü. Radyolojik muayene ve laboratuvar testleri normaldi. Topikal kortikosteroid, siklopentolat damla ve oral prednizolon tedavisine başlandı. Altı hafta sonra, hastanın semptomları geriledi ve görme keskinlikleri 20/20'ye geri döndü. Çoğu vakada, tipik APMPPE için tedavi gerekli değildir. Sistemik ve topikal kortikosteroidler ve/veya sikloplejiler atipik klinik özelliklere sahip seçilmiş APMPPE vakalarında yararlı olabilir.

**Anahtar Kelimeler:** Granülomatöz hastalık, kronik; panüveit; hidrokortikosteroidler; koroid hastalıkları

Türkiye Klinikleri J Med Sci 2012;32(2):532-6

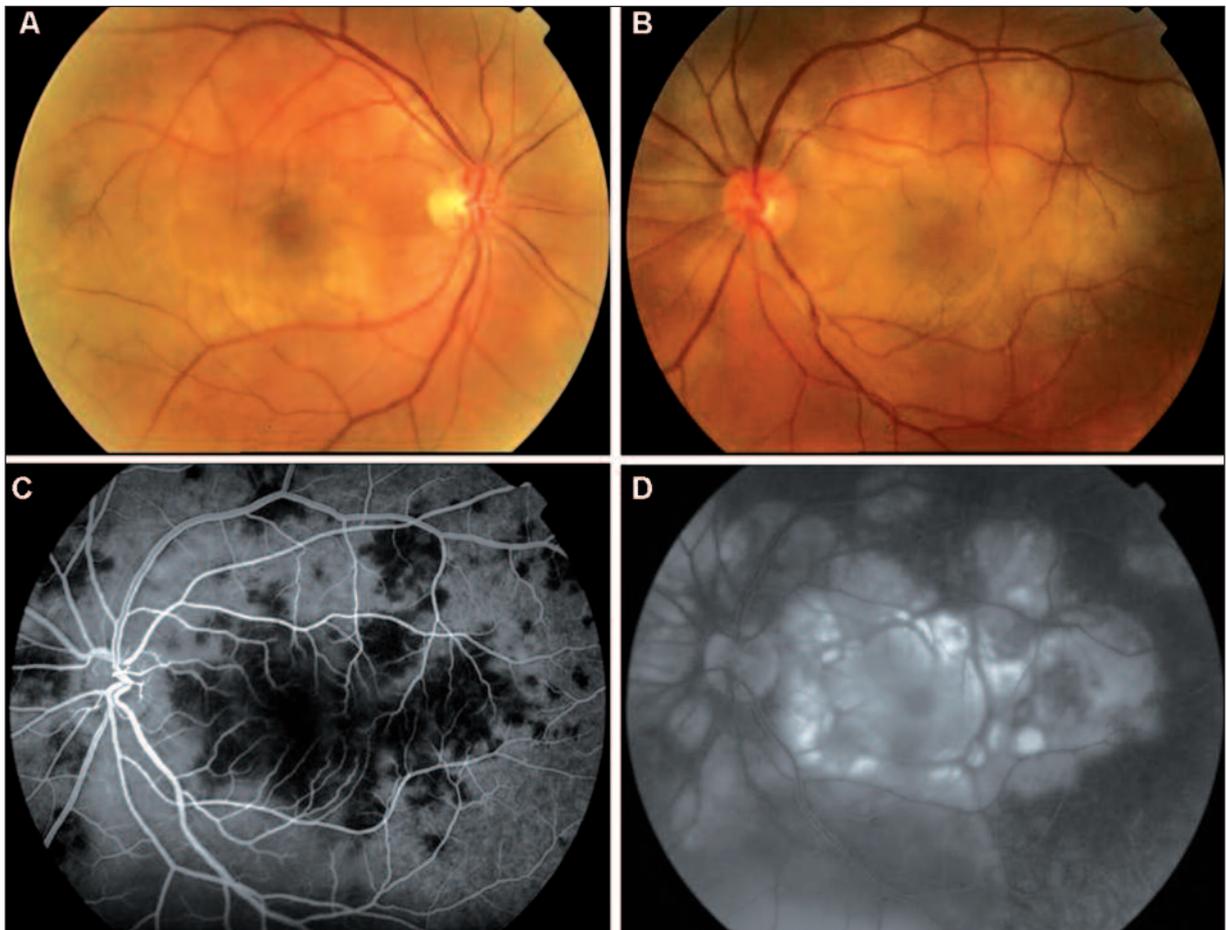
**A**cute posterior multifocal placoid pigment epitheliopathy (APMPPE) is a posterior uveitis that typically presents as an acute loss of vision in one or both eyes. It usually appears in young adults and affects both genders equally. APMPPE is characterized by multiple, yellow-white, wide plaque-shaped inflammatory lesions at the level of the retinal pigment epithelium in the macula and posterior pole, and is diag-

nosed using clinical findings and fundus fluorescein angiography (FFA).<sup>1</sup> The lesions are usually bilateral and resolve spontaneously within 2-3 weeks, leaving a discrete pigment epithelial scar.<sup>2</sup> Despite permanent alteration of the retinal pigment epithelium (RPE), the prognosis is usually good, and spontaneous visual recovery often occurs without treatment. Recently, it has been suggested that the presence of atypical features including age over 60 years, recurrent disease and unilateral involvement indicate a poor visual prognosis.<sup>2-4</sup> Corticosteroids have been used to treat cases of severe bilateral vision loss and cases with central nervous system involvement, but their efficacy has not been proven.<sup>2,5</sup> Other clinical findings associated with APMPE include vitreous cells in 50% of eyes, serous retinal detachment, episcleritis, iridocyclitis, periphlebitis, papillitis, disk edema and slight

dilation and tortuosity of the retinal veins.<sup>6</sup> Infrequent observations of granulomatous anterior uveitis associated with APMPE have been reported.<sup>7</sup> In the present paper, we report a patient with APMPE presenting with granulomatous anterior uveitis and posterior synechia.

## CASE REPORT

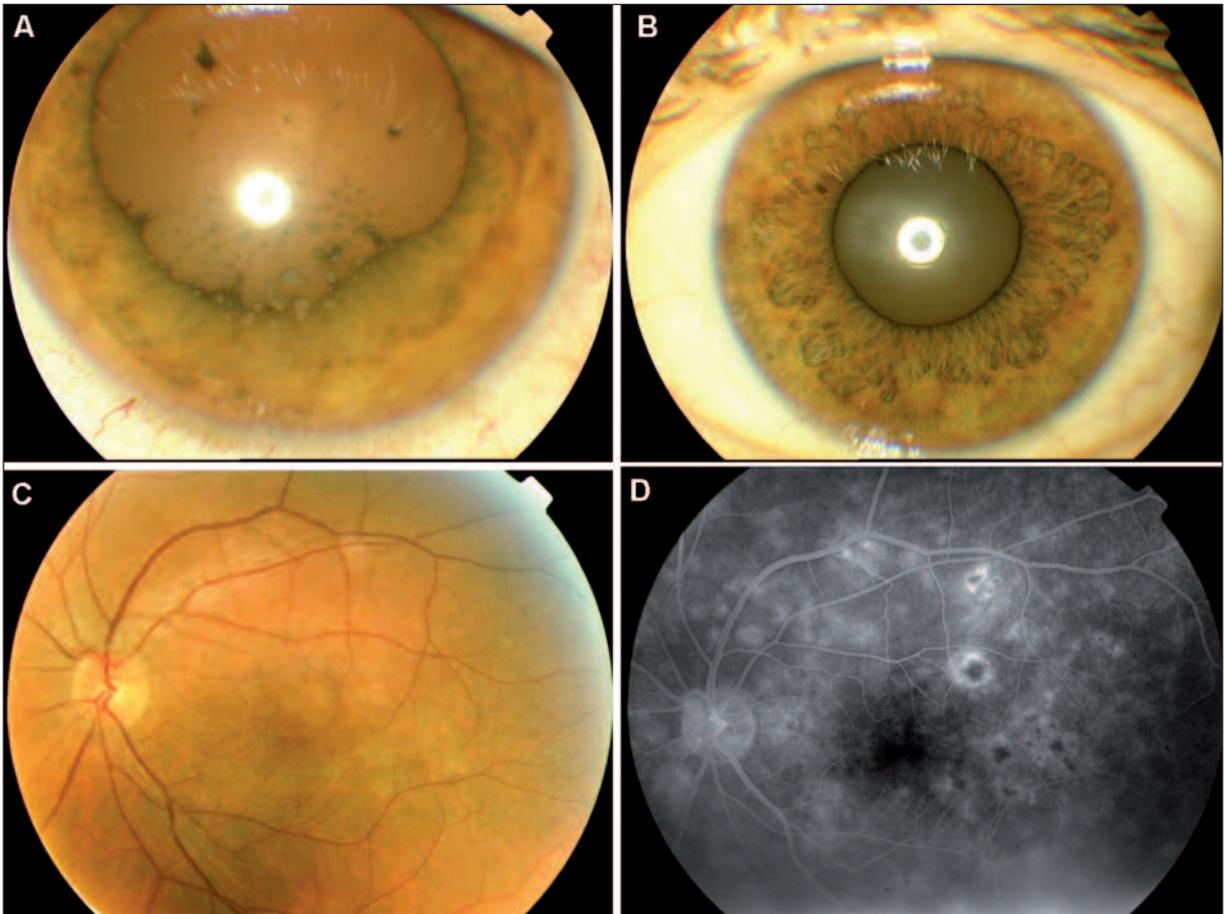
A previously healthy 43-year-old male presented with sudden bilateral vision loss. His visual acuity was 20/125 in the right eye and 20/400 in the left eye. The intraocular pressure and results of the slit lamp examination were unremarkable. Examination of the fundus revealed multiple placoid, yellow-white lesions located deep in the retina of both eyes. FFA revealed early hypofluorescence and late hyperfluorescence, typical of APMPE (Figure 1).



**FIGURE 1:** Color fundus photographs of both eyes (A, right eye; B, left eye) showing multiple yellow-white placoid lesions. (C) The early-phase fluorescein angiogram of left eye shows hypofluorescence of placoid lesions and (D) the late-phase fluorescein angiogram of left eye shows diffuse staining of the placoid lesions. (See for colored form <http://tipbilimleri.turkiyeklinikleri.com/>)

The patient was not treated at that time. Four days later, he complained of bilateral floaters, red eyes and photophobia. His visual acuity was 20/200 in right eye and 20/400 in the left eye. We observed 2+ cells, mild flare in the anterior chamber, mutton-fat keratic precipitates on the corneal endothelium of both eyes and posterior synechia in the left eye (Figure 2). We found 3+ cells in the vitreous. Follow up history revealed that the patient had not suffered any eye problems. Treatment with 2-hourly topical prednisolone acetate 1% and cyclopentolate 1% twice daily and oral prednisolone (1 mg/kg/day) was initiated. The topical and oral medications were gradually reduced and were discontinued after six weeks. The patient's blood chemistry was within normal limits and the serology tests were negative for cat-scratch disease, Lyme titer, syphilis and toxocariasis. Tests for

rheumatoid factor, antinuclear antibody, and anti-neutrophil cytoplasmic antibodies were negative. The patient's chest X-ray, tuberculosis tests, angiotensin converting enzyme level and erythrocyte sedimentation rate were within the normal limits. Our clinical diagnosis was APMPE with granulomatous anterior uveitis. On the 5<sup>th</sup> day of the treatment, the patient's best corrected visual acuity was 20/20 in the right eye and 20/25 in the left eye. We observed gradual improvement of the anterior chamber reaction and the fundus lesions. On follow up six weeks later, the patient's symptoms regressed and the best corrected visual acuity was 20/20 bilaterally. The anterior segment was quiet and showed no keratic precipitates on the corneal endothelium. The ophthalmoscopic examination revealed multiple grayish lesions on the posterior pole. These lesions were flat and deeply located to



**FIGURE 2:** (A) Mutton-fat keratic precipitates on the corneal endothelium and posterior synechiae in the left eye at the pre-treatment period. (B, C and D) Illustrations of the left eye six weeks after the treatment are shown in other photographs.

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the retinal vessels. FFA revealed a large number of irregular lesions, some of them blocking the choroidal fluorescence, and others showing staining at the edges (Figure 2).

## DISCUSSION

APMPPE was first described by Gass in 1968.<sup>8</sup> The etiology and pathogenesis of this disease remains unclear. One-third of patients have reported a flu-like illness prior to the onset of APMPPE, suggesting that it may be caused by hypersensitivity to microbial antigens. This hypothesis is supported by reports of APMPPE development following hepatitis B vaccination, mumps, swine flu vaccination and bacterial infections.<sup>9</sup> Two hypotheses exist regarding the pathogenesis of APMPPE. Gass postulated primary involvement of the RPE because the lesions overlying the RPE were flat.<sup>8</sup> This hypothesis is supported by subnormal electrooculogram findings in the acute phase of the disease, suggesting widespread abnormality of the RPE.<sup>1</sup> Van Buskirk et al. have proposed that delayed filling of the choriocapillaris was caused by a focal choroidal vasculopathy, rather than a primary pigment epitheliopathy with masking of the underlying choroid.<sup>10</sup> Several studies support the role of decreased choroidal perfusion in the pathogenesis of APMPPE, including that of Howe et al., who used indocyanine green (ICG) angiography to study APMPPE.<sup>11</sup> It has been shown that there is an obstruction in the choriocapillaris at the level of the precapillary arterioles in the ICG angiography.<sup>1</sup> Those findings suggest that APMPPE is a primary choroidal vascular disease and affects RPE.<sup>4</sup> Our observation of inflammatory cells in the anterior

chamber of the eye provides further support for this hypothesis. The inflammatory reaction affects either the anterior or posterior uveal tissue, causing secondary changes in the RPE cell.

The present case of APMPPE with uveitis was most similar to multifocal choroiditis or Vogt-Koyanagi-Harada syndrome, especially with the presence of granulomatous anterior uveitis. These two disorders are included in the differential diagnosis of APMPPE.<sup>12</sup> FFA is essential for the diagnosis of APMPPE. The lesions are characterized by hypofluorescence in the early stages and hyperfluorescence in the later stages.<sup>1</sup>

It is not known how the association of granulomatous anterior uveitis is related to the pathogenesis of APMPPE, nor is it clear why only a limited number of patients with APMPPE develop anterior uveitis. Type IV hypersensitivity reaction might play a role in the inflammatory damage caused by APMPPE; for instance, several patients with APMPPE have shown positive tuberculin skin tests.<sup>12</sup> Furthermore, granulomatous inflammation was found in a renal biopsy of a patient with APMPPE who developed sarcoidosis, and it was observed in the arterial walls of two patients who had APMPPE and simultaneous cerebral vasculitis.<sup>13</sup>

Subtle or transient signs of anterior chamber inflammatory changes that have not been identified may be present during the course of APMPPE. Patients with APMPPE usually have a good visual prognosis. Treatment is not necessary for most cases of typical APMPPE; however, systemic and topical corticosteroids and/or cycloplegics may be useful in treating carefully selected cases of APMPPE, with atypical clinical features.

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