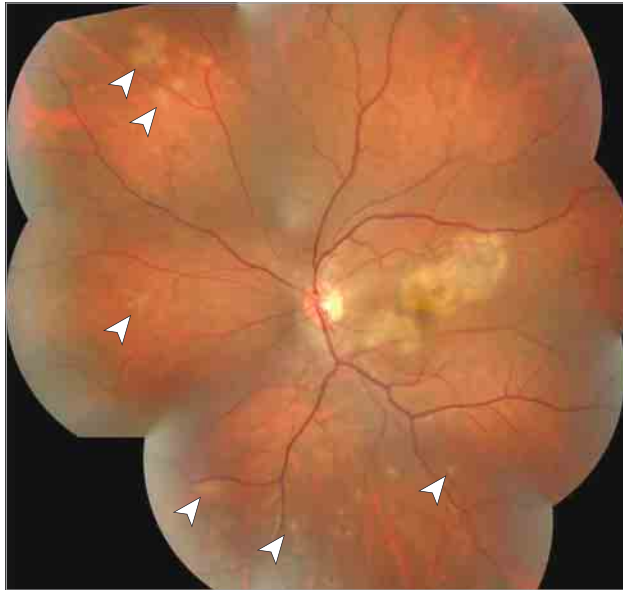


## JAMA Ophthalmology Clinical Challenge

## Vision Loss and Paresthesias in a Young Man

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A Fundus photograph



B Fluorescein angiogram



**Figure 1.** A, Subretinal and retinal pigment epithelial lesions (arrowheads) are visible in the left eye. B, Late staining of the macular lesion. The arrowheads outline the border of the area that was hypofluorescent in the early frames (not shown). The right eye showed similar findings.

**A young man** presented to the emergency department with a 1-day history of vision loss in the left eye. He had experienced eye redness for 1 week, which resolved with use of artificial tears; a mild posterior headache; and paresthesias of the left thumb and index and middle fingers.

His medical history was significant for epilepsy as a child, although he had been seizure free for more than 10 years without medication. He had no ocular history, was not taking any medications, and had no known drug allergies. He smoked 2 to 4 packs of cigarettes per day, consumed 1 to 3 alcoholic drinks per day, and denied illicit drug use. He denied recent travel, vaccinations, systemic illnesses, or influenza-like symptoms.

His best-corrected visual acuity was 20/20 OD and 20/100 OS. Amsler grid testing revealed a central scotoma in the left eye. No afferent papillary defect was apparent. Intraocular pressures were 16 mm Hg OU. Results of the external and anterior segment examinations were unremarkable, and no vitreous inflammation was noted. **Figure 1** shows findings of the posterior segment examination. Results of the neurologic examination revealed decreased sensation to light touch, vibration, and temperature in his left upper and lower extremities.

## WHAT WOULD YOU DO NEXT?

- A. Initiate high-dose corticosteroid therapy
- B. Obtain magnetic resonance imaging of the brain
- C. Perform a retinal biopsy
- D. Obtain computed tomography of the chest

## Diagnosis

Acute posterior multifocal placoid pigment epitheliopathy (APMPPE) with presumed associated cerebral vasculitis

## What To Do Next

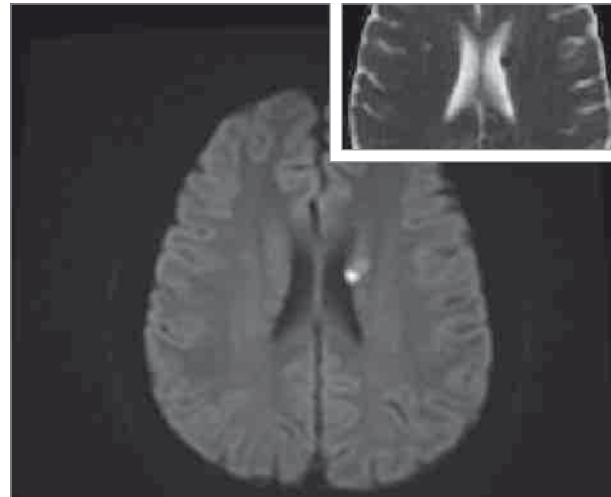
### B. Obtain magnetic resonance imaging of the brain.

Classic clinical findings such as bilateral postequatorial creamy gray-yellow placoid lesions at the level of the retinal pigment epithelium demonstrating early blockage and late staining on fluorescein angiography in a young, otherwise healthy patient are often sufficient for establishing a diagnosis of APMPPE. Before considering therapy, however, pertinent associated signs and symptoms should be addressed. This patient described a headache that, in the setting of a prodromal viral illness, has been described with APMPPE and does not always warrant neuroimaging.<sup>1</sup> However, the presence of peripheral sensory deficits should prompt urgent magnetic resonance imaging (MRI) with diffusion-weighted imaging and apparent diffusion coefficient sequences to evaluate for signs of cerebral vasculitis.

## Discussion

The rare inflammatory eye disease APMPPE has been associated with multiple systemic conditions, including Wegener disease, systemic lupus erythematosus, and erythema nodosum, although causative relationships have not been established. Central nervous system involvement, ranging in severity from headaches to diffuse cerebral vasculitis, stroke, and death, has also been described, with at least 17 cases of APMPPE-related cerebral stroke reported.<sup>2</sup>

Optimal therapeutic regimens for APMPPE have not been established because the disease is so rare; however, corticosteroids and immunomodulatory agents are used variably. Some authors<sup>3,4</sup> have posited that isolated APMPPE in an otherwise healthy young adult follows a self-limited clinical course and carries an excellent visual prognosis without treatment. Others<sup>2,5,6</sup> have reported substantial rates of persistent visual field defects, metamorphopsia, and incomplete visual recovery while remaining equivocal regarding the role of systemic therapy. Several<sup>1,7,8</sup> agree that despite the paucity of high-quality clinical evidence, rapid initiation of therapy is justified in patients with central nervous system involvement and that long-term immunomodulatory therapy may be necessary.



**Figure 2.** Representative section of magnetic resonance imaging, which demonstrated foci of marked hyperintensity on diffusion-weighted imaging sequences and corresponding hypointensity on apparent diffusion coefficient sequences (inset), consistent with acute stroke.

## Patient Outcome

The MRI in our patient revealed multiple hyperintense foci on diffusion-weighted imaging sequences and corresponding hypointensities on apparent diffusion coefficient sequences, consistent with acute stroke (Figure 2). Results of the cerebrospinal fluid sample analysis were normal. Results of the laboratory workup for tuberculosis, sarcoidosis, and syphilis were negative. The patient received a 3-day course of intravenous methylprednisolone sodium succinate (Solu-Medrol); soon after therapy was transitioned to oral prednisone, he developed new symptoms and was found to have new cerebral infarcts. Oral cyclophosphamide was added, and he continued this regimen for 7 months before the prednisone dose was tapered gradually. Five months after treatment initiation, the patient's visual acuity was 20/20 OD and 20/25 OS, with no recurrence of the disease.

Most patients with APMPPE can expect an excellent overall prognosis. However, physicians of patients who demonstrate any neurologic signs or symptoms should consider prompt neuroimaging because delayed treatment of APMPPE-associated cerebral vasculitis can result in substantial morbidity or death.

## ARTICLE INFORMATION

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