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Letter to the Editor

A case of acute posterior multifocal placoid pigment epitheliopathy with aseptic meningitis and cerebral infarction

1. Case report

A 24-year-old male with no medical history presented with a bilateral blurred vision that progressively worsened for three days without influenza-like illness. Fundoscopy showed multiple cream-colored placoid lesions (Fig. 1A). Optical coherence tomography revealed multiple hyperreflective lesions located in the outer nuclear layer and subfoveal photoreceptor layer disruption in both eyes (Fig. 2). Fluorescein angiogram showed early hypofluorescent lesions with a staining in the late sequence (Fig. 1B). Indocyanine green chorioangiography angiograms showed bilateral multiple hypofluorescent lesions in the early phase which persisted throughout the late phase (Fig. 3). Based on clinical features, the patient has been diagnosed with bilateral acute posterior multifocal placoid pigment epitheliopathy (APMPPE).

The next day, the patient was referred to the neurology department for a thunderbolt headache. He did not show any meningeal symptoms nor signs of focal lesion. Magnetic resonance imaging (MRI) showed no abnormalities. The analysis of the cerebrospinal fluid (CSF) revealed a lymphocyte count of 34.10^6/L and a protein level of 0.46 g/L without any evidence of oligoclonal bands, a glucose level of 0.56 g/L. PCR for HSV, VZV and enterovirus in the CSF was reported negative. A lupus anticoagulant was detected in blood samples without any other immunological abnormalities. *Borrelia burgdorferi* serology, syphilis serology and tuberculin skin test were negative. Minor salivary gland biopsy and lung computed tomography were normal.

The diagnosis of APMPPE with aseptic meningitis was made. Intravenous methylprednisolone therapy was induced (3 g in 3 days) and followed by an oral steroid therapy of 1 mg/kg/day.

After two months, while decreasing steroid therapy, the patient presented with sudden-onset visual impairment, Weber's syndrome and impaired consciousness (NIHSS 20) requiring intensive care. Brain MRI showed mesencephalic and cerebral infarction in the territories of the posterior cerebral artery and its paramedian branches, of the middle cerebral artery and of the anterior cerebral artery. Magnetic resonance angiography and contrast-enhanced MRI showed signs of vasculitis and meningitis (Fig. 1C). Cardiovascular investigation was normal. Intravenous methylprednisolone therapy and monthly cyclophosphamide infusion were induced, resulting in a partial improvement (NIHSS 9).

2. Discussion

APMPPE is an uncommon chorioretinopathy, affecting both women and men aged 20–50 years, characterized by multiple cream-colored placoid lesions located in the posterior pole leading to scotomas and photopsia [1]. Fluorescein angiography shows an early hypofluorescence and a late hyperfluorescence due to an inflammatory choriocapillaritis. The more frequent systemic sign is headache sometimes in thunderclap preceded by visual signs from few days to several months. Although rare, its neurological complications are: stroke, cerebral haemorrhage and seizure [2,3] due to vasculitis. The natural history of APMPPE is often good with recuperation of visual acuity but some symptoms may remain, and recurrences may occur.

The histopathological findings in cerebral arteries showed a segmental giant cells vasculopathy of meningeal arteries [4]. These findings are not reported in Vogt-Koyanagi-Harada disease which is a differential diagnosis. APMPPE may rarely be associated with sarcoidosis, syphilis, Lyme disease, adenovirus type 5 infection, tuberculosis, necrotizing vasculitis, vaccination for hepatitis B virus and varicella [5]. Investigations should research these associated diseases.

Headache and pleocytosis in CSF seem to be predictive factors of stroke [3], justifying a neurological care. When these predictive factors are present, an aggressive treatment like an immunosuppressive medication (azathioprine or cyclophosphamide) could be used. Presently, there is no consensus for therapeutic management of neurological involvement.

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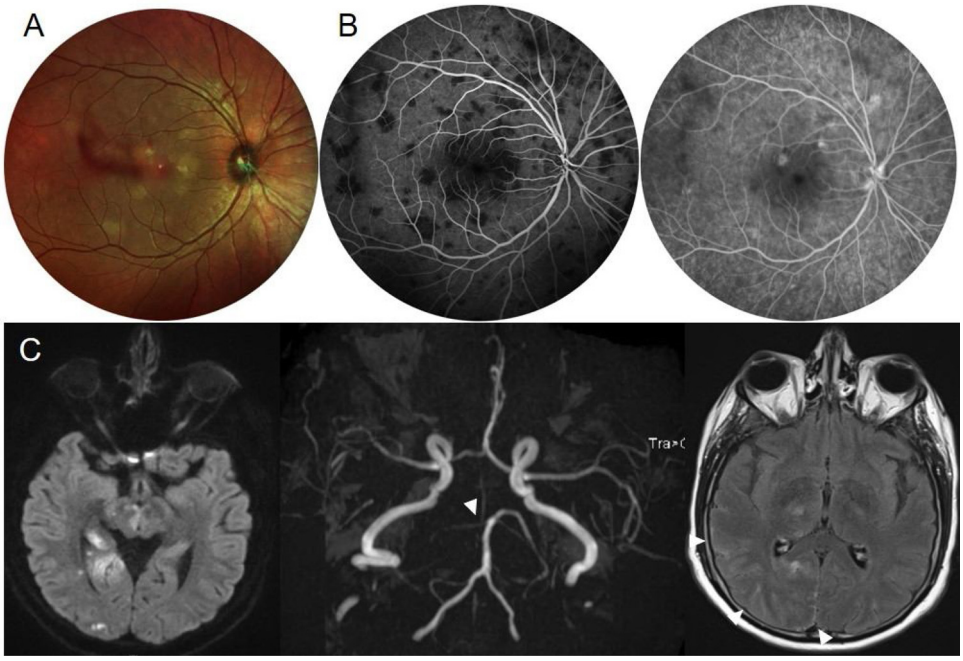

Fig. 1 – Ophthalmological investigation and Brain MRI. A: Right eye fundus color photography reveals multiple cream-colored placoid lesions. B: Fluorescein angiogram shows early hypofluorescence of the lesions (left) and later in the sequence, bright staining (right). C: Brain MRI: diffusion-weighted (left) image shows cerebral infarction of the posterior cerebral artery and its paramedian branches. MRA image (middle) shows an occlusion of the right posterior cerebral artery (arrow). Contrast-enhanced T2-FLAIR-weighted image (right) shows a temporo-occipital meningeal enhancement (arrow). Abbreviations. FLAIR: Fluid Attenuated Inversion Recovery. MRA: Magnetic Resonance Angiography.

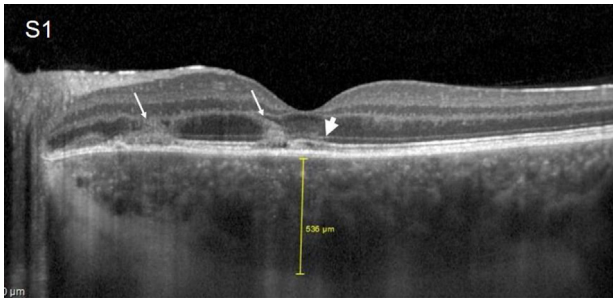

Fig. 2 – Optical coherence tomography. Optical coherence tomography reveals hyperreflective lesions located in the outer nuclear layer (white arrows) and focal photoreceptor layer disruption (white arrows head).

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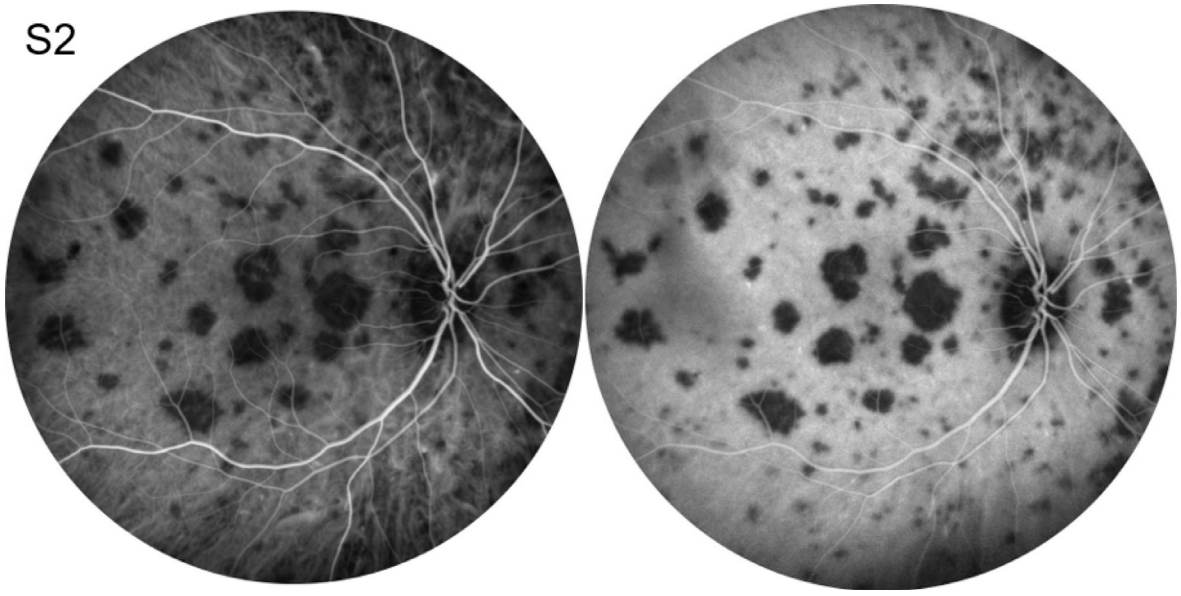

Fig. 3 – Indocyanine green chorioangiography angiograms. Right eye ICG angiograms demonstrates multiple hypofluorescent lesions in the early phase (left) which persisted throughout late phase (right). Abbreviations. ICG: Indocyanine Green Chorioangiography.

Prospective studies will be useful to determine predictive factors and define guidelines for treatment.

Disclosure of interest

The authors declare that they have no competing interest.

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