• Letter to the Editor •

Primary vitreoretinal lymphoma masquerading as acute posterior multifocal placoid pigment epitheliopathy

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Dear Editor,

Primary vitreoretinal lymphoma (PVRL) is a rare form of ocular malignancy and it is usually considered as a subset of primary central nervous system lymphoma (PCNSL). An insidious onset associated with delays in diagnosis is common. PVRL presents with nonspecific symptoms including painless blurred vision, floaters and decreased visual acuity[1-2]; clinical signs vary significantly between patients and at times can mimic other ocular diseases. Acute posterior multifocal placoid pigment epitheliopathy (APMPPE) is a self-limited inflammatory disorder typically affecting young patients, following a flu-like syndrome in approximately one-third of the cases[3].

Here, we report an atypical case of PVRL masquerading as APMPPE. In July 2016, a 42-year-old Caucasian woman presented at the Foundation Adolphe de Rothschild Hospital in Paris, complaining of sudden decreased vision and scotomas in her right eye. She reported an episode of mild fever and headache one month prior to the onset of the diminished vision. Based on clinical examination and a first multimodal imaging evaluation (Figure 1A, 1B), the physicians made a first diagnosis of APMPPE with a delayed involvement of the fellow eye, and an intravitreal steroid injection was performed in the right eye within a few days. Two weeks later, whilst holidaying in Sardinia, Italy, persistent symptoms called for a second ophthalmological evaluation at the Eye Clinic, University of Cagliari. Best corrected visual acuity was 0.3 logMAR in the right eye and 0 logMAR in the left eye. Anterior segment slit lamp examination of both eyes was unremarkable, whereas fundus examination showed some subretinal infiltrates appearing as yellowish lesions more evident in the right eye. Suspecting a white dot syndrome a multimodal imaging was required. Fluorescein angiography, indocyanine green angiography and spectral domain optical coherence tomography (SD-OCT) findings (Figure 1C-1J) were compatible with the diagnosis of APMPPE. A three-week oral steroid therapy was dispensed. In October, at the scheduled follow up visit, the new examination revealed a worsening of the right eye retinal lesions, and suspecting a PVRL further examinations were sought, for which patient opted to have carried out to completion at the Institut Curie in Paris. In November 2016 the right eye analysis of aqueous disclosed an increased interleukin (IL)-10/IL-6 ratio, and vitreous biopsy revealing the presence of large atypical lymphocytes confirmed the final diagnosis of PVRL with large B-cell. All medical and neurologic examinations including neuro-imaging and lumbar puncture showed no systemic or central nervous system (CNS) involvement. Four weekly intravitreal injections of methotrexate in the right eye were administered, resulting in a complete regression of retinal active lesions. Moreover, a systemic chemotherapy was also initiated. In this case there were several elements that could have led to a misleading diagnosis of a white dot syndrome. The patient reported a flu-like episode and ocular symptoms which are common in APMPPE. Furthermore the latter mostly affects young adults, whereas intraocular lymphoma usually arises at the average age of 60 years. Added to which, PVRL can present with CNS involvement which was absent in this patient. Finally, the SD-OCT examination carried out in Cagliari revealed in right eye lesions compatible with a stage 1 APMPPE according to the classification proposed by Goldenberg *et al*[4].

In conclusion, PVRL can masquerade as intermediate and posterior uveitis, vitritis or retinitis[1]; and as APMPPE as demonstrated in this paper for the first time. In the case of suspected PVRL a diagnostic vitrectomy or retinal biopsy is needed particularly in the absence of a prior CNS disease. Additionally, considering that prognosis is related to an early diagnosis, the management of patient necessitate a multidisciplinary team to achieve a prompt and correct diagnosis, and to commence appropriate therapy.

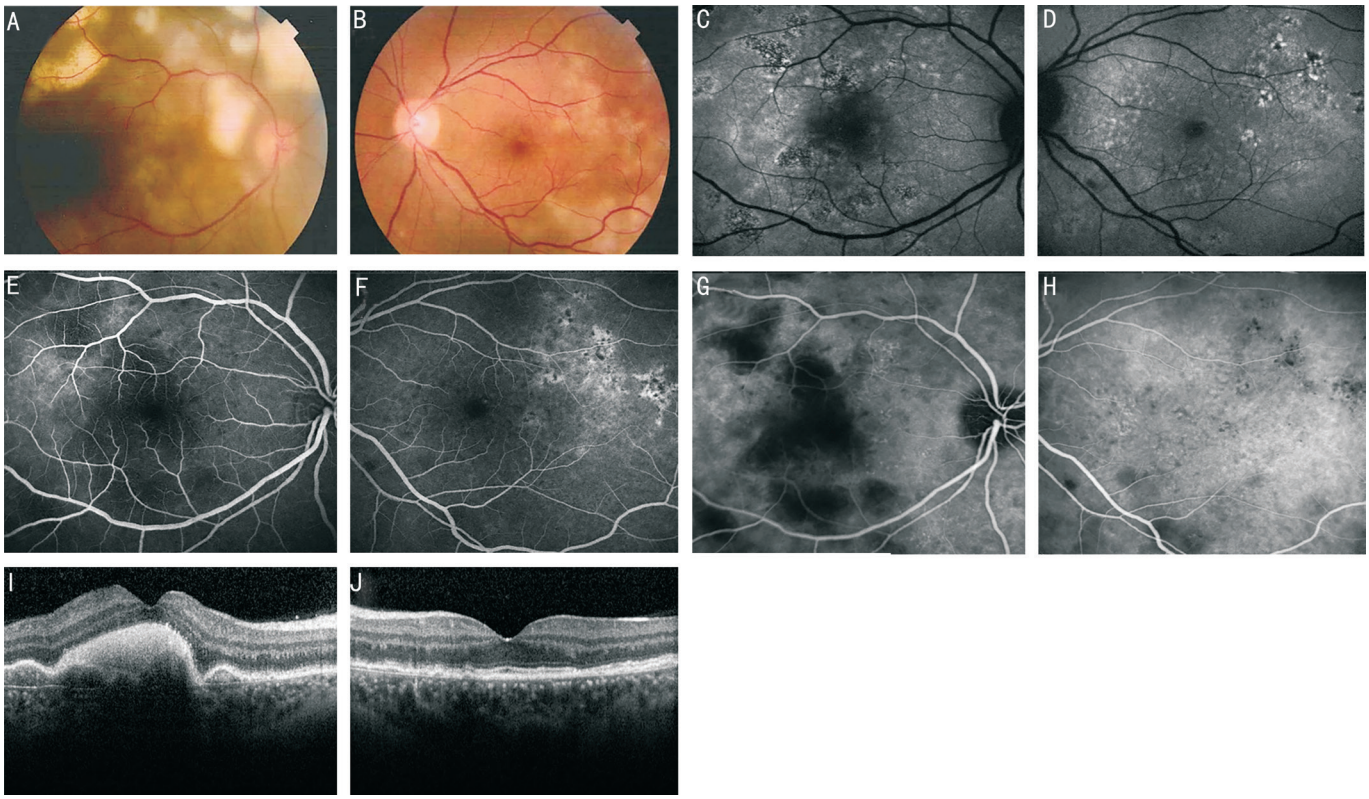

Figure 1 Baseline evaluation of both eyes made at the Foundation Adolphe de Rothschild Hospital in Paris A-B: Colour fundus picture showing yellowish-white lesions located primarily at posterior pole and varying in size. Second ophthalmological evaluation at the Eye Clinic, University of Cagliari; C-D: Fundus autofluorescence displaying some areas of hypoautofluorescence with hyperautofluorescent borders; E-F: Early phase fluorescein angiography showing some areas of hypofluorescence in right eye and mottled hyperfluorescence in left eye; G-H: Mid-late phase indocyanine green angiography confirming the dye blockage areas better visible in the right eye; I-J: Spectral domain optical coherence tomography showing a macular dome-shaped elevation with hyperreflective sub-RPE material in the right eye, and small RPE elevations with an IS/OS junction disruption in the left eye.

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