PIGMENTARY EPITHELIOPATHY MULTIFOCAL ACUTE PLACOID ASSOCIATED WITH PARALYSIS OF VI CRANIAL PAR

EPITELIOPATÍA PIGMENTARIA PLACOIDE MULTIFOCAL AGUDA ASOCIADA A PARÁLISIS DEL VI PAR CRANIAL

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ABSTRACT

Clinical case: We report the case of a 24-year-old patient who attended our hospital with an acute posterior multifocal placoid pigment epitheliopathy (APMPPE) which was later confirmed by fluorescein angiography. One month after presentation the patient developed a right VI nerve palsy.

Discussion: APMPPE is an acute-onset bilateral inflammatory disease causing impaired vision. Although it is thought to be benign, neurologic manifestations have been described even months after presentation. There is no previous report of APMPPE associated with VI nerve palsy (Arch Soc Esp Oftalmol 2009; 84: 159-162).

Key words: Acute posterior multifocal placoid pigment epitheliopathy, VIth nerve palsy

INTRODUCTION

Acute posterior multifocal placoid pigment epitheliopathy (APMPPE) is referred to as one of the white dots syndromes. It affects young and middle-aged adults, with equal incidence in both genders. Its clinical manifestation is a decrease in visual acuity (VA) which tends to be bilateral and asymmetrical. It is generally considered to have a benign prognosis and resolves without treatment in

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6 to 12 weeks, leaving behind areas of pigment change at the pigment epithelium level. The present article describes a bilateral APMPPE in a 24-year-old patient with neurological manifestations after one month follow-up.

**CASE REPORT**

A 24-year-old male arrived in the ER reporting decreased bilateral VA and cephalgia of two days evolution. Relevant personal history included flu during the previous week treated with Ibuprofen 600 mg. VA exploration of the right eye reveals lumps; left eye VA is 0.05. Pupils are isochoric and normoreactive and biomicroscopy of the anterior pole is normal. Ophthalmoscopy reveals white-grayish confluent lesions in the posterior pole in both eyes affecting the fovea (fig. 1). Suspecting APMPPE, a fluorescein angiography (FAG) was performed, revealing hypofluorescence areas at an early time (fig. 2) which become hyperfluorescence areas at a later stage (fig. 3). Indocyanine green angiography (ICG) shows hypofluorescence areas in the choroid corresponding to placoid lesions related to the choroidal hypoperfusion accompanying this pathology (fig. 4). The optic coherence tomography shows retinal thickening with hyper-reflectivity of the external retinal layers. Oral corticoids treatment (prednisone 60 mg) was prescribed over a week. On the other hand, the patient reported an intense cephalgia which improved with the corticoid treat-

![Fig. 1: Ophthalmoscopy: white-grayish lesions in the posterior pole.](image1)

![Fig. 2: Fluorescein angiography: hypo-fluorescent areas in early times.](image2)
ment and reappeared after suspending medication, thus suspecting cerebral vasculitis, sometimes associated with this condition. A magnetic nuclear angiographic resonance (MNAR) did not reveal any signs of vasculitis.

The requested histocompatibility study for HLA B-7 and HLA DR2 was positive.

Regarding evolution, visual acuity gradually improved though one month later during follow-up the patient reported diplopia at distance viewing. Exploration revealed paralysis of the right VI cranial par, with slight limitation of abduction in the right eye, a fact which was confirmed with the Hess-Lancaster test. Botulinum toxin was injected in the medial rectum of the right eye and diplopia disappeared within a week.

At present, diplopia has disappeared and VA is 0.7 in the right eye and 0.8 in the left eye. OCT reveals recovery of the foveal architecture, although there was an increase of retinal reflectivity in both eyes.

**DISCUSSION**

Posterior multifocal placoid pigment epitheliopathy is a rare entity included among the white dots syndromes. It is generally considered a benign condition with spontaneous resolution, although one should always bear in mind that full vision recovery will not be achieved when lesions involve the fovea.
40% of cases report a pseudo-flu condition during the week prior to the decrease in VA.

Fluorescein angiographies reveal the presence of hypofluorescence areas in early times with hyperfluorescent areas in late times. Indocyanine green angiographies (ICG) show hypofluorescence areas matching the placoid lesions, which are related to choroidal hypoperfusion secondary to vascular occlusion areas (4). Optic coherence tomography in these patients reveals retinal thickening with increased reflectivity in the retina’s external layers (5). In the present case, retinal thickness has disappeared, while retinal hyper-reflectivity persists.

A small percentage of cases are accompanied by neurological manifestations which may be more or less severe. Cephalea is related to the association between APMPPE and aseptic meningitis, which does not require treatment, although it is important to perform adequate neurological explorations (1). Patients may present different degrees of cerebral vasculitis and meningoencephalitis, which may appear weeks or months after APMPPE (1,3). The patient presented diplopia after one month follow-up which during exploration turned out to be paralysis of the right VI cranial par. It resolved favorably after injecting botulinum toxin in the medial rectum of the right eye. This neurological complication has not been confirmed so far in the literature.

REFERENCES