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Short communication

Multiple retinal pigment epithelial detachments: A case report[☆]

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ABSTRACT

Case report: A 47-year-old female who presented with a bilateral idiopathic multiple pigment epithelial detachment (PED) in a routine visit. This pathology is shown as a rare clinical manifestation, where the outcome is resolution of localized atrophy of the pigment epithelium, with a good functional prognosis.

Discussion: PED is a common clinical manifestation in several chorioretinal diseases, particularly in macular degeneration associated with age. Idiopathic PED can be considered as a kind of central type II serous chorioretinopathy. Fundus fluorescein angiography (FFA) and optical coherence tomography (OCT) are complementary tests to study the number, extension, and nature of these PED.

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Múltiples desprendimientos de epitelio pigmentario idiopático: a propósito de un caso

RESUMEN

Caso clínico: Mujer de 47 años de edad presenta múltiples desprendimientos de epitelio pigmentario (DEP) idiopáticos bilaterales en una revisión rutinaria. Se muestra esta enfermedad como una manifestación clínica rara cuya evolución será la resolución, atrofia localizada del epitelio pigmentario y el buen pronóstico funcional.

Discusión: El DEP es una manifestación clínica frecuente en múltiples enfermedades coriorretinianas. La degeneración macular asociada a la edad (DMAE) es donde con mayor frecuencia se manifiesta. La forma idiopática se puede englobar dentro de la coriorretinopatía central serosa (CCS) tipo II. La angiografía (AFG) y la tomografía de coherencia óptica (OCT) son pruebas complementarias en el estudio del número, la extensión y la naturaleza serosa de estos DEP.

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Palabras clave:

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Introduction

Retinal pigment epithelium detachment (PED) is a frequent clinical expression in a range of chorioretinal diseases. Age-related macular degeneration (ARMD) is the disease in which PED expresses most frequently. It consists a separation between the basal lamina of the pigment epithelium cell and the innermost collagen layer of Bruch's membrane.¹

Possible causes for PED include inflammation, ischemia, as well as degenerative and idiopathic causes. When the latter occurs in patients under 50 years of age a variant of serous central chorioretinopathy (SCC) must be considered. It could be due to alteration in the vascular permeability of the choroids due to ischemia² and usually courses with good visual prognosis.

Clinical case

A female, 47, with personal history of hyperlipidemia, asthma and stable *angina pectoris* presented in a routine ophthalmological examination multiple bilateral PED without visual symptoms.

Visual acuity (VA) was of one in both eyes (BE), intraocular pressure and biomicroscopy were normal while funduscopy revealed multiple bilateral lesions of different sizes, slightly raised and hypopigmented without confluence, with well-defined edges and dotted pigment changes surrounded by an orange ring and distributed in the posterior pole closer to the fovea in the right eye (RE) (Fig. 1).

BE autofluorescence exhibited roundish hypoautofluorescent lesions surrounded by hyperautofluorescent halos, corresponding to PED (Fig. 1).

Optic coherence tomography (OCT) (HD OCT Cirrus, Zeiss) showed multiple small dome-shaped hyper-reflective lesions which did not affect the fovea in BE, compatible with PED (Fig. 2).

Angiofluoresceingraph (FA) revealed multiple bilateral lesions which were hyperfluorescent in early times in well defined forms and shapes and in greater numbers than revealed by funduscopy. Said lesions were more numerous and superior in LE and close to the fovea in the RE, without evidencing neovascularization, vasculitis or vascular occlusion (Fig. 3).

The patient was referred to Internal Medicine for study, where she underwent general analytics, serology for syphilis, herpes virus and cytomegalovirus among others, Mantoux, chest X-ray, autoimmunity tests, globular segmentation rates and angiotensin conversion enzyme. As all the results were negative, systemic disease was discarded. Accordingly, the diagnosis was multiple PED of idiopathic etiology.

Six months later the patient was examined again and exhibited VA of 0.8 in RE, and of one in LE. Funduscopy revealed in addition to PED, neuroepithelium detachment in the macular area of RE with elevation and no variations in LE. In RE, OCT showed lesions compatible with small size dome-shaped hyper-reflective PED involving the fovea, with some larger sized lesions above the fovea together with neuroepithelium detachment without modifications in LE (Fig. 2).

The patient was asked to return one month later for examination, exhibiting diminished neuroepithelium detachment

thickness in RE, although small PED persisted in the macula. VA remained at 0.8 in RE and 1 in LE (Fig. 2).

Two months later, the patient continued with the same VA and funduscopy showed some EP alterations in the RE macula without elevation and a lower number of PED in BE (Fig. 2).

At present, the patient is being examined regularly in our service.

Discussion

The majority of RPE detachments occur in males between 20 and 60 years of age. Although they can appear as an isolated finding, they could be the beginning of the development of serous detachment.³

The close apposition of the retinal pigment epithelium, neurosensory retina, Bruch membrane and choroids are extremely important to enable retina metabolism and normal function. Adhesion takes place on one side between the villi of the pigment epithelium and the external photoreceptor segments (supplemented by active transport through pigment epithelium and greater osmotic pressure in the choroids that forces continued extraction of liquid from the subretinal space) and, on the other side, due to the union of the pigment epithelium basal membrane and Bruch membrane, which is carried out through laminin filaments, collagen type IV and V and proteoglycans in the hemidesmosome regions. Any disorder which destabilizes this balance could have an effect on the retina and the pigment epithelial adhesion. Several hypotheses have been proposed but the most important reason is the composition of the choroidal interstitial fluid, which is determined by the degree of choroidal vascular permeability. Any inflammatory, infectious, vascular, degenerative, malign or genetically determined process that can give rise to such changes is able to produce alterations in the pigment epithelium adhesion. Isolated PED develops due to the disintegration of the retinal pigment epithelium union with the collagen layer of Bruch's membrane. If this disintegration persists in time it could lead to serous detachment.³

Quite frequently, history reveals ocular or systemic concomitant diseases such as ARMD, angiod striations, presumed ocular histoplasmosis syndrome, polypoidal choroidal vasculopathy, Vogt-Koyanagi-Harada syndrome, sarcoidosis, hyperviscosity, kidney and collagen diseases, malign hypertension, cytomegalovirus infections or retinal neoplasia such as primary ocular lymphoma and acute myeloid leukemia.^{4,5}

In the patient of this report, infectious systemic disorders were discarded due to negative serology against tuberculosis, syphilis, cytomegalovirus, HIV and herpes simplex among others, as well as due to the absence of choroidal infiltrates which are typical of fungal infections. Inflammatory diseases such as sarcoidosis were also discarded due to the absence of choroidal granuloma, thorax X-ray and normal angiotensin conversion enzyme, Vogt-Koyanagi-Harada (the patient did not exhibit sudden loss of vision or ocular pain or other symptoms such as fever, headache, nausea, neurological symptoms such as stiff neck or hearing alteration) or other systemic inflammatory diseases such as rheumatoid arthritis, polyarteritis nodosa, Wegener due to negative autoimmunity analysis, and absence of other symptoms which arise with

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