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Multiple retinal pigment epithelial detachments: what should we do? A propos of an idiopathic case

Vários descolamentos do epitélio pigmentar da retina: o que devemos fazer? A propósito de um caso idiopático

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Abstract

The purpose is to report the management of multiple retinal epithelial detachments (PEDs) in a 50-year-old male with bilateral PEDs not associated with chorioretinal or systemic pathologies after a complete study. Idiopathic multiple PEDs could be a variation of central serous chorioretinopathy, but other causes as well as other pathological conditions which could produce similar images, are required to be ruled out.

Keywords: *Central serous chorioretinopathy; Fluorescein angiography; Indocyanine green; Tomography, optical coherence; Retinal detachment*

Resumo

O objetivo é relatar o manejo de múltiplos descolamentos epiteliais da retina (DEPs) em um homem de 50 anos de idade com DEPs bilaterais não associado a patologias coriorretinianas ou sistêmicas após um estudo completo. PEDs idiopáticos múltiplos podem ser uma variação da coriorretinopatia serosa central, mas precisam excluir outras causas, bem como outras condições patológicas que podem produzir imagens semelhantes.

Descritores: Coriorretinopatia serosa central; Angiofluoresceinografia; Verde de indocianina; Tomografia de coerência óptica; Descolamento retiniano

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This study was carried out according to principles and basic ethical regulations originated in the Declaration of Helsinki (Fortaleza, Brazil version; October 2013) approved by the World Medical Association.

Os autores declaram não haver conflito de interesses.

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Introduction

Retinal pigment epithelial detachment (PED) is a non-specific anatomical alteration, produced by compromised adhesion of the retinal pigment epithelium (RPE) basal membrane to the inner layer of Bruch's membrane. It may be single or multiple and it frequently derives from chorioretinal pathologies, among which age-related macular degeneration (AMD) has an outstanding prevalence.[1-4]

It has also been related to systemic inflammatory processes such as sarcoidosis and Vogt-Koyanagi-Harada syndrome, infections such as Cytomegalovirus (CMV), and hyperviscosity, hypercortisolism and malignant hypertension syndromes.[1-4]

An idiopathic origin is uncommon in multiple PEDs. In this regard, it exists a theory which states that multiple PEDs could be a variation of central serous chorioretinopathy (CSC).[1-4]

Case Report

A 50-year-old male was referred to our service for screening of diabetic retinopathy. He presented as antecedents of interest type 2 diabetes, obesity, arterial hypertension (AHT), dyslipidemia, stress and smoking habit.

On physical examination, visual acuity was 20/20 (Snellen visual acuity ratios) in both eyes, the patient did not refer visual alteration and the Amsler test result was negative. The anterior segment was normal; fundus examination showed PEDs in the temporal retinal arcade area in both eyes (Figure 1A).

In view of this finding a number of additional tests were run to identify a triggering disease. The optical coherence tomography (Cirrus HD-OCT; Carl Zeiss Meditec, Dublin, CA) showed the presence of several smaller PEDs in the superior temporal arcade in addition to those detected during fundus evaluation in the left eye, without any associated neurosensory retinal detachment (NSD) (Figure 1B).

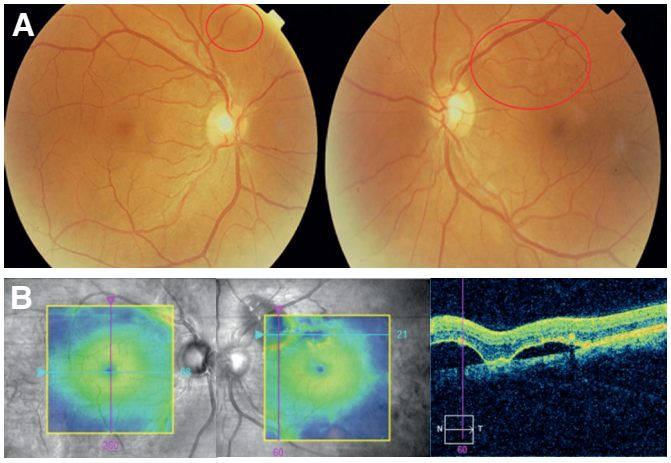

Figure 1: A) Fundus images showing PEDs in both superior temporal retinal arcades. B) OCT Cirrus of the both eyes showing several PEDs without any associated NSD in the left eye

The fluorescein angiography (FAG) by Topcon TRC 50 IX (Topcon Medical Systems, Inc) revealed an early hyperfluorescence of the PEDs, without objective signs of neovascular membrane (CNV), or any other pathological findings (Figure 2).

Then, an indocyanine green angiography (ICGA) by Topcon TRC 50 IX (Topcon Medical Systems, Inc) (Figure 3) was carried out mainly in order to exclude polypoidal choroidal vasculopathy (PCV). Neither polyps nor hot spots which may suggest retinal angiomatous proliferation (RAP) were found. Finally, an ocular ultrasonography ruled out choroidal tumors.

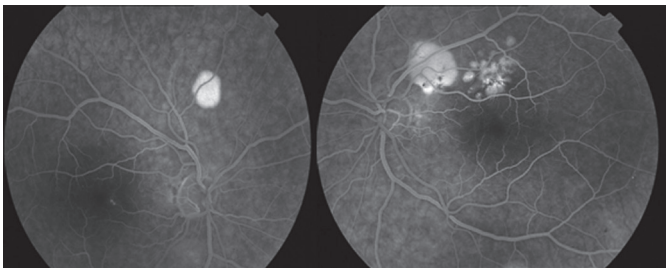

Figure 2: Image of the PEDs in the FAG of both eyes

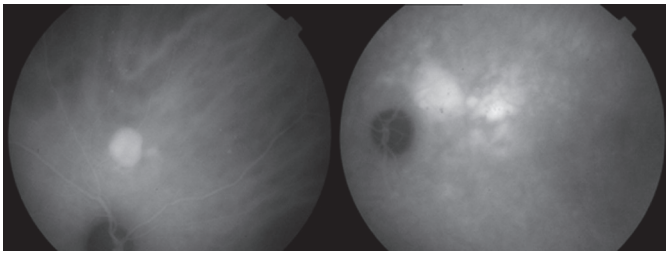

Figure 3: Image of the PEDs in the ICGA of both eyes

As these findings did not show any related chorioretinal pathology, it was decided to consult with the Internal Medicine service for systemic examination to identify any systemic aetiology. The medical evaluation included a comprehensive systemic history and exploration, a complete blood test (including cortisol, leukocytes, ACE, coagulation, ESR, protein, C-reactive protein tests and autoimmune markers); a chest radiograph, CMV, syphilis, herpes simple and HIV serologies and a Mantoux test. Infectious diseases, collagen diseases, sarcoidosis, kidney disease, hyperviscosity syndrome and malignant hypertension were ruled out.

Given the negative results of all tests, the patient was diagnosed with multiple chronic serous PEDs of unknown etiology and it was decided to run periodical tests only, as the visual acuity was not compromised. After a thirty -month follow-up, the lesions remained stable in size and number, and the patient remained asymptomatic.

Discussion

Regarding the presence of PEDs, it is necessary to run a complete set of tests because it is a non-specific alteration frequently related to pathology. It is vital to determine the aetiology because it will provide the guidelines for the evolution of the process and for the therapeutic approach 1. The authors of this manuscript recommend the following general guidelines in the presence of multiple PEDs, keeping in mind that the management of each patient should be individualized. First, a proper anamnesis and a complete ophthalmologic examination, including OCT, FAG and sometimes ICGA and ocular ultrasonography to exclude associated chorioretinal pathology as AMD, polypoidal choroidal vasculopathy, angioid streaks, CSC and lymphoma.[1-4] Another important point is the differential diagnosis with other apparently similar conditions, such as vitelliform dystrophy, where OCT demonstrates the hyper-reflective material in the subretinal space. The pathology is confirmed by an abnormal electrooculogram, with an acute reduction of darkness-light ratio and a pathological

index of Arden.[2, 4]

If an associated systemic disease is suspected or if the ophthalmologic study was not able to identify the cause, a systemic exam, ideally in conjuction with an internist, must be carried out with a correct history and exploration, with a chest radiograph and blood tests (including cortisol, leukocytes, ACE, coagulation, ESR, protein, C-reactive protein tests and autoimmune markers).

Serologies of CMV, syphilis, herpes simplex and HIV and a Mantoux test for tuberculosis should be done to exclude infectious disease. Systemic and inflammatory diseases such as sarcoidosis, rheumatoid arthritis, Vogt-Koyanagi-Harada disease, vasculitis like Wegener and node polyarteritis, disseminated intravascular coagulopathy, malignant hypertension, pregnancy associated with hypertension, lupus, Goodpasture's syndrome, hyperviscosity, hypercortisolism, Waldenstrom's disease, cryoglobulinemia and lymphoproliferative disorders must be ruled out.[1-8]

Idiopathic multiple PEDs are very similar to CSC regarding the characteristic profile of patients: males nearing their middle age and presenting a significant level of emotion-related stress. Both entities can appear simultaneously and they have been etiopathogenically associated, caused by an alteration in vascular permeability of the choroid, [3] considered as two forms of the same process.[4] In asymptomatic cases, the best procedure is the follow-up.[2] When there is a visual dysfunction, either photodynamic therapy 5 or focal argon laser photocoagulation 4 are useful, although there is not a treatment protocol established for these cases. Antiangiogenic therapy is indicated in cases of neovascular membrane associated to PEDs.[3]

In conclusion, according to the presence of multiple PEDs, a choroidal neovascular membrane must always be suspected, especially in patients older than 55, and it is essential to run an AFG test.[3] Most patients under 55 presenting with multiple PEDs have a good prognosis if the appropriate tests show no chorioretinal or systemic pathologies, even more so if the fovea is unaffected and there is no neurosensory retinal detachment. In those cases, the best therapeutic option is periodical follow-up.[3]

The theory postulated here is that idiopathic multiple PEDs could be a variation of CSC [1-8] but, like every idiopathic process, it requires to exclude other causes as well as other pathological conditions which could produce similar images.

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