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Short communication

Surgical treatment in combined hamartoma of the retina and retinal pigment epithelium☆

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ABSTRACT

Case report: The case is presented of a 39 year-old man with a combined hamartoma of the retina and retinal pigment epithelium, who experienced progressive visual loss and worsening of metamorphopsia. The patient underwent vitrectomy and epiretinal component peeling, with improvement in visual acuity, metamorphopsia, and retinal architecture, assessed by optical coherence tomography.
Discussion: Selected patients with combined hamartomas of the retina and retinal pigment epithelium may benefit from surgical management.

Tratamiento quirúrgico en un hamartoma combinado de retina y epitelio pigmentario

Palabras clave:
Hamartoma combinado de retina y epitelio pigmentario de retina
Membrana epirretiniana
Vitrectomía
Tomografía de coherencia óptica

RESUMEN

Caso clínico: Presentamos el caso de un paciente varón de 39 años, con un hamartoma combinado de retina y epitelio pigmentario de retina. El paciente refería disminución progresiva de la visión y empeoramiento de la metamorfopsia. Se realizó una vitrectomía con pelado de la membrana epirretiniana, consiguiendo una mejoría de la agudeza visual y de la metamorfopsia, así como de la arquitectura retiniana en la tomografía de coherencia óptica.
Discusión: Algunos pacientes seleccionados con hamartomas combinados de retina y epitelio pigmentario podrían beneficiarse de la realización de una vitrectomía.

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Introduction

Combined hamartoma of the retina and retinal pigment epithelium (CHRRPE) is an infrequent, benign and presumptively congenital tumor[1] that can produce significant visual acuity loss. CHRRPE expresses as a solitary and unilateral lesion located in the optic disc or posterior pole, presenting slightly raised tumors with variable amounts of pigmentation, retinal vessel tortuosity and formation of epiretinal membranes (ERM).[2] In addition, CHRRPE associates a variable amount of gliosis on the retinal surface that can give rise to ERM, retinal distortion and tractional retina detachment.[3]

The use of surgery in CHRRPE is controversial. Very few cases reporting vitrectomy for resolving the retinal traction have been published.[2–5]

The case of a male, 39, who referred progressive visual acuity (VA) loss and increased metamorphopsia is presented. Vitrectomy with ERM peeling and elimination of the superficial glial component was carried out.

Clinic case report

Male, 39 years, diagnosed a few years earlier with CHRRPE in the left eye (LE), who referred progressive vision loss and increased metamorphopsia. Maximum corrected visual acuity (AVMC) was 1.0 in the right eye (RE) and 0.4 in the LE. The previous visual acuity of the LE was unknown as the patient was new, although he insisted on referring diminished vision in said eye.

The LE ocular fundus (OF) showed the presence of CHRRPE in the form of one elevated and pigmented lesion with its center in the superior temporal arch, surrounded by retina pigment epithelium (RPE) alterations, with dilated, tortuous and telangiectasic vessels, as well as a significant tractional component. Hard macular exudates were also observed (Fig. 1).

Spectral domain optic coherence tomography (SD-OCT) (Topcon® 3D OCT-1000, Tokyo, Japan) revealed a hyper-reflective and compact mass with posterior hyporeflective shadow, and a significant tractional component with macular involvement (Fig. 2).

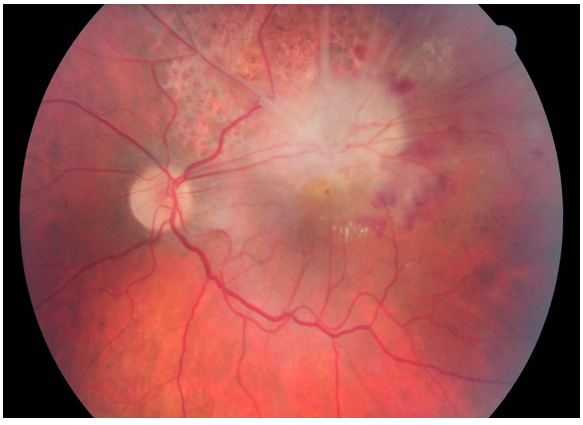

Fig. 1 – Left eye posterior pole retinography, showing the presence of CHRRPE in the superior zone of the macula, as an elevated mass with vascular dilatation and significant glial component on the surface. The lesion is surrounded by RPE alterations with associated hemorrhagic component and hard exudates in the macula.

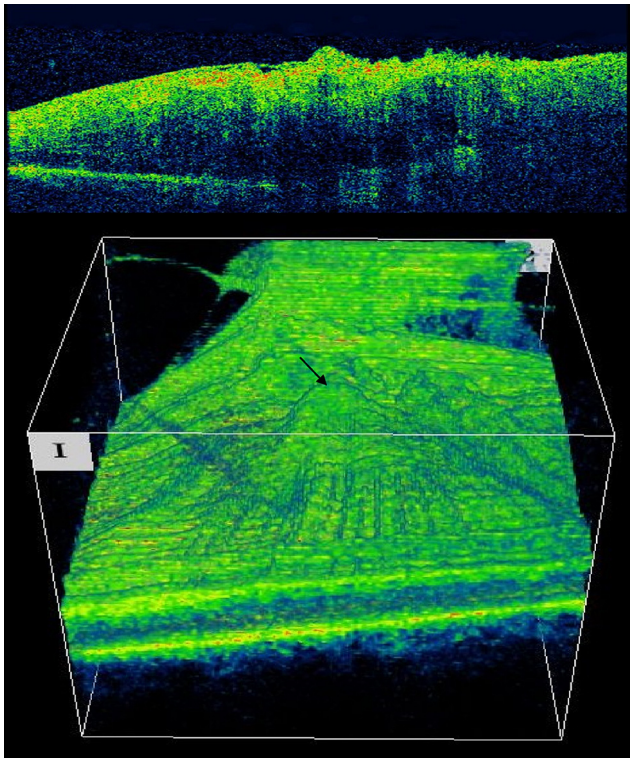

Fig. 2 – SD-OCT of CHRRPE: hyper-reflective mass with posterior shadow. In 2-D image ERM can be observed whereas 3-D image shows the adherence of the vitreous to the lesion.

Fluorescein angiography (FAG) evidenced said vascular and RPE alterations, with slight diffusion in late times (Fig. 3). The rest of the exploration produced normal results.

Vitrectomy was performed, including peeling of ERM and internal limiting membrane (ILM), and extraction of a large glial component which was firmly adhered to the cusp of the tumor, achieving partial relaxation of the lesion.

Fifteen months after surgery, BCVA improved to 0.63 and metamorphopsia diminished. The appearance of the fundus

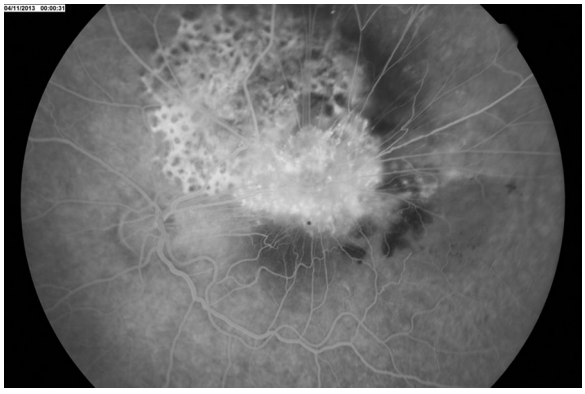

Fig. 3 – Fluorescein angiography of CHRRPE, showing RPE alteration and traction exerted over the macula.