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Short communication

Unilateral retinitis pigmentosa. A case report☆

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ABSTRACT

Clinical case: A 27-year-old woman with a history of nyctalopia and constriction of visual field of the right eye. The ophthalmological examination showed a visual field and electroretinogram that were compatible with unilateral retinitis pigmentosa (RP). After a one year follow-up, the unilateral condition remained.

Discussion: Unilateral retinitis pigmentosa is a rare condition, with a frequency between 0.2% and 5% of the RP. It mainly affects women and older age groups than bilateral RP. For a definitive diagnosis, it is necessary to have a funduscopy and electroretinogram (ERG) altered unilaterally, and exclude infectious, inflammatory, and vascular causes.

Retinitis pigmentosa unilateral a propósito de un caso

Palabras clave:
Retinitis pigmentosa
Retinitis pigmentosa unilateral
Distrofia retiniana
Distrofia bastón-cono
Nictalopía

RESUMEN

Caso clínico: Mujer de 27 años con historia de nictalopía y constricción de campo visual del ojo derecho. El examen oftalmológico, campo visual y electrorretinograma (ERG) fueron compatibles con una retinitis pigmentosa (RP) unilateral. Al seguimiento de un año, permanecía la afección unilateral.

Discusión: La RP unilateral es un trastorno infrecuente, con una frecuencia entre el 0,2-5% de las RP. Afecta principalmente a mujeres, y en edades más avanzadas que las bilaterales. Para dar un diagnóstico definitivo tiene que haber un fondo de ojo y ERG alterados unilateralmente, y excluir causas infecciosas, inflamatorias y vasculares.

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Introduction

Retinitis pigmentosa (RP) is the most frequent retinal dystrophy, characterized by being hereditary, causing progressive dysfunction and loss of cells, first compromising rods and then cones. RP is also known as rod–cone dystrophy.[1]

Generally, RPE expresses bilaterally within the first three decades of life and is characterized by nyctalopia and progressive loss of peripheral visual field. In the early stages, central vision is preserved although it deteriorates with the progressive loss of photoreceptors and for this reason it is diagnosed in more advanced stages. RP is associated to cystoid macular edema (20% of cases) and posterior subcapsular cataracts (50% of cases).[1,2]

Ocular fundus examinations reveal pigment changes known as bone spicules due to their shape, a typical characteristic of the disease. Additional signs include optic nerve paleness, retina pigment epithelium atrophy and vascular attenuation.[2,3]

RP may express sporadically or follow a hereditary pattern with genetic heterogeneity. Its inheritance mechanism can be autosomal dominant (30–40% of cases), autosomal recessive (50–60%) or X-linked (5–15%).[4,5]

In RP, alterations are usually limited to the eyes. However, extraocular involvement has been described in the context of some syndromes, approximately 20–30% of cases.[2,6]

The therapeutic options are limited. Vitamin A supplementation in diet has been proposed, although its role is controversial.[2]

A clinic case of a patient with unilateral RP is described below.

Clinic case report

Female, 27, with a history of nyctalopia and right eye (RE) visual field constriction of 2 years evolution was admitted. No history of traumatism or other ophthalmological events, use of medicaments, systemic, pneumatic or vasculitic diseases or familial antecedents.

Ophthalmological examination showed a visual acuity of 20/20 in both eyes, preserved color vision and RE relative afferent pupillary defect. Biomicroscopy revealed poorly vitreous pigments in the RE, while funduscopy showed a paler right papilla with generalized attenuation of vasculature and bone-shaped pigment in the periphery with perifoveal macular atrophy (Fig. 1). Left eye (LE) biomicroscopy and funduscopy were normal (Fig. 2).

The patient was evaluated with a range of examinations, i.e., computerized visual field that showed a tubular field in the RE, being normal in the LE (Fig. 3). RE autofluorescence evidenced increased fluorescence at the parafoveal level (Fig. 4). In addition, OCT showed increased granular hyper-reflectiveness, collapse of nuclear layers, particularly the internal one, generalized retinal thickness thinning and increased internal limiting membrane signal (Fig. 5). Standard and multifocal electroretinogram (ERG) did not show response of rods or cones in the RE. However, these were normal in the LE (Fig. 6).

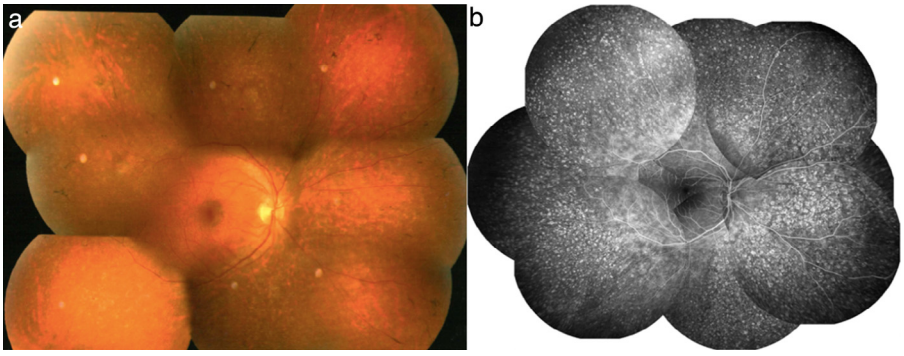

Fig. 1 – (a) Right eye fundus, showing the optic nerve paleness, vascular attenuation and osteous pigment; (b) right eye angiography in 3–4 min, showing window-type defects up to the periphery.

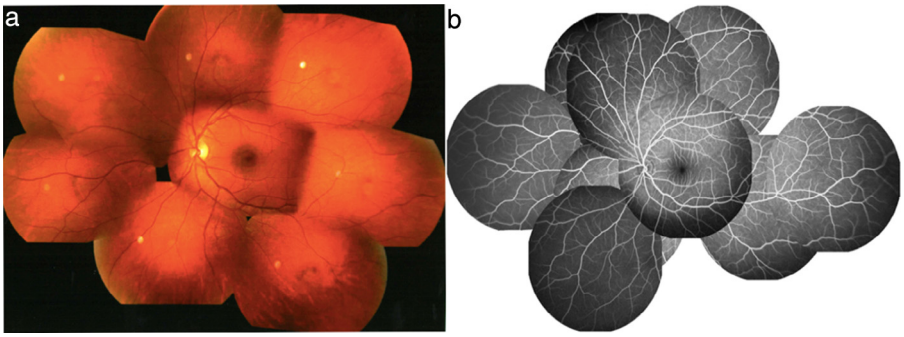

Fig. 2 – (a) Lefts eye fundus, and (b) left eye angiography within normal limits.