Short communication

Pigmented paravenous retinochoroidal atrophy with macular involvement

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ABSTRACT

Case report: A 65-year-old female, with poor visual acuity in right eye since childhood, was referred to our hospital. Funduscopic examination revealed bone-spicule pigmentation and choroidal and retinal pigment epithelium atrophy along the vascular arcades, with macular involvement in right eye. The patient was diagnosed with pigmented paravenous retinochoroidal atrophy (PPRCA).

Discussion: PPRCA is a rare disease, of unknown origin, characterised by retinochoroidal atrophy and retinal pigmentation along retinal veins, generally without macular involvement. Latest publications show a slow progression. No effective treatment is available.

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Atrofia coriorretiniana pigmentada paravenosa con afectación macular

RÉSUMÉ

Caso clínico: Mujer de 65 años que acude al hospital por mala agudeza visual por ojo derecho desde la infancia. El fondo de ojo revelaba pigmentación espiculada y atrofia del epitelio pigmentario retiniano y coroides a nivel de arcadas, afectándose además la mácula en ojo derecho. La paciente fue diagnosticada de atrofia coriorretiniana pigmentada paravenosa (ACRPP).

Discusión: La ACRPP es una enfermedad poco frecuente, de etiología desconocida, caracterizada por atrofia coriorretiniana y acúmulo de pigmento a lo largo de las venas retinianas, generalmente sin afectación macular. Las últimas publicaciones demuestran progresión lenta. No existe tratamiento efectivo.

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Introduction

Pigmented paravenous retinochoroidal atrophy (PPRCA) is a disease characterized by retinochoroidal atrophy and accumulation of pigment, generally in the form of bone spicules, along retinal veins. Involvement is usually bilateral even though it can be asymmetric. Macular alteration is not frequent. PPRCA is an infrequent entity of unknown cause of which only about 100 cases have been described.  

This paper presents a PPRCA case with macular involvement.

Case report

A 65-year-old female visited our hospital in 2004 due to poor visual acuity (VA) in the right eye (RE) since childhood, without history of ocular trauma or inflammation. Personal antecedents were not relevant.  

The patient exhibited a corrected VA of finger counting in the RE and of 1.0 in the left eye (LE). The anterior pole of both eyes (BE) was normal and the intraocular pressure (IOP) was of 16 mmHg in BE. Ocular fundus exploration revealed retinal pigment epithelium (RPE) and choroidal atrophy with spiculated pigmentation. In the LE the disease was restricted to the territory of the superior and inferior temporal arch whereas in the RE all the arches and the macular area were affected (Figs. 1 and 2). No signs of vitritis were found. Optic coherence tomography of the macular area confirmed the RPE atrophy and the accumulation of pigment in the RE as well as the absence of LE involvement (Figs. 3 and 4).

In the 24-2 visual field scotoma were observed, corresponding to the retinochoroidal atrophy areas. Electroretinogram revealed reduced response amplitude in the RE.

A systemic study was carried out, comprising physical assessment, thorax X-ray and serology for virus herpes and syphilis, with all results being normal. The patient was diagnosed of PPRCA.

In successive assessments of the past 3 years no progression has been observed in funduscopic alterations or VA reduction.

Discussion

PPRCA is an uncommon disease of unknown etiopathogenic. Some cases have been associated with inflammatory diseases such as Behcet’s disease and infectious conditions such as syphilis, tuberculosis and rubeola. Recently, a mutation in gene CRB1 has been detected in PPRCA patients. This gene is involved in various retinal dystrophies. Even though the majority of cases are sporadic, some familial cases have been described.

For some authors, PPRCA is a response pattern to an inflammatory or infectious disease, while for others it is retinal dystrophy. Other authors consider that PPRCA could be an incomplete form of pigmentary retinitis.

The macular area is nearly always respected. For this reason, the vast majority of patients with PPRCA are asymptomatic and the diagnosis is usually a casual finding. However, the present case exhibited an important macular involvement of the RE with poor VA in said eye.

The progressive or stationary nature of this disease is the subject of controversy although recent published papers have demonstrated a slow progression through the years.
To this date there is no effective treatment for this disease.

Conflict of interests

No conflict of interests has been declared by the authors.

REFERENCES