Short communication

Acute retinal pigment epitheliitis. Diagnosis using optical coherence tomography

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ABSTRACT

Case report: A 17 year-old female presented with a bilateral and acute visual loss. On ophthalmoscopic examination, there was a subfoveal deposit with a vitelliform appearance. Optical coherence tomography revealed a hyperreflective and homogeneous material located at the photoreceptor external segment layer. A month later, vision had spontaneously recovered and macular appearance was normalized. On tomography, the subretinal material had completely disappeared.

Discussion: Acute retinal pigment epitheliitis is a rare condition that usually causes a transient visual loss, with a good prognosis in young subjects.

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Epitelitis pigmentaria retiniana aguda. Diagnóstico mediante tomografía de coherencia óptica

RESUMEN

Caso clínico: Una mujer de 17 años de edad consultó por una disminución visual aguda y bilateral. Oftalmoscópicamente se le observó un depósito subfoveal de aspecto vitelliforme. En la tomografía de coherencia óptica, aparecía como un material hiperreflejivo y homogéneo acumulado en la capa de los segmentos externos de fotorreceptores. Al mes de evolución, se produjo la recuperación visual espontánea, con la normalización del aspecto macular. Tomográficamente se comprobó también la desaparición de dicho material subretiniano.

Discusión: La epitelitis pigmentaria retiniana aguda es una enfermedad infrecuente, que suele causar una pérdida visual transitoria y de buen pronóstico, en sujetos jóvenes.

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Introduction

Acute retinal pigment epithelitis (ARPE) or Krill’s disease is an infrequent retinal disorder characterized by the appearance of blurred vision or central scotoma.\(^1\) Its onset is unilateral or bilateral and generally affects healthy young adults. It is sometimes preceded by a fever-like infection produced by picornavirus.\(^2\) This disorder has no gender preference and it generally resolves spontaneously in a few months with good individual prognosis.\(^2\) Typically, it is attributed to retina pigment epithelium (RPE) alterations although there is no agreement about the initial location of the process.\(^3\) At the ophthalmoscopic level, the disease is characterized by dotted pigment alterations associated to yellowish lesions located in the RPE of the central macula.\(^1-3\)

Clinic case

A female patient, 17-years-old, visited due to visual difficulty in both eyes (BE) that began a few days earlier. In addition, she referred for recent upper airways infection. No relevant personal history was noted with the exception of the usage of oral contraceptives (OC) beginning 1 year earlier. The composition of the contraceptive was 0.02 mg ethinyl estradiol and 0.075 mg gestodene.

In the ophthalmological examination, the best-corrected visual acuity in the right eye (RE) was of 0.8 and the left eye (LE) was of 0.9. No significant alterations were found in the anterior segments and the ocular pressure was normal in both eyes. The funduscopic examination revealed a yellowish deposit circumscribed to the fovea in BE without edema or exudation but with associated dotted pigment alterations (Fig. 1). The retinal vessels were normal and the papilla exhibited physiological cups with normal colors and clearly defined. Amsler’s grid did not exhibit apparent metamorphopsia although it showed a small central scotoma which was larger in the RE.

By means of spectral domain optic coherence tomography (SD-OCT) an accumulation of dense and homogeneous hyper-reflective material was observed between the retina and the RPE with preservation of the foveal depression. The hyper-reflective lines corresponding to the external limiting membrane (ELM) and the union of the internal and external photoreceptor segments (IS/OS) were preserved even though those of the RPE were poorly defined (Figs. 2 and 3). The patient was requested to visit the following week for fluorescein angiography (FA), recommending her to suspend the OC. The patient did not turn up for the other FA and appeared a month later for a checkup. The best-corrected visual acuity was of 0.9 in RE and 1.0 in LE. The subfoveal deposit had disappeared and the macula had recovered normality with funduscopic examination with the exception of small and discreet dotted alterations in the RPE (Fig. 4). SD-OCT confirmed the disappearance of the subretinal material in BE. The IS/OS lines and ELM exhibited...
normal appearance although the RPE was slightly irregular (Fig. 4).

Discussion

ARPE is a rare foveal disorder which affects young adults, with acute but temporary and self-limited course.\(^1\)\(^2\)\(^3\) Diagnostic in the acute phase is considered to be difficult and usually based on clinical suspicion, on funduscopic examination and angiographic characteristics.\(^4\) In this case FA was not performed due to the patient missing the appointment. However, the recent introduction of optic coherence tomography facilitated the definition of the morphological characteristics of this condition throughout its clinical course and to develop a hypothesis relating to physiopathology.\(^1\)\(^2\) Hsu et al.\(^1\) were the first to study ARPE by means of optic coherence tomography in the temporal domain mode (TD-OCT). The 3 cases described by the said author found increased foveal reflectiveness in the photoreceptor and external nuclear layers which also affected the RPE (in 2 of the 3 cases), with total absence of serous intra- or subretinal accumulations. With clinical resolution, the hyper-reflective thickening of the photoreceptor layer decreased.

Subsequently, applying SD-OCT in the study of an additional case, Puche et al.\(^2\) described the preservation of the IS/OS line, the elongation of photoreceptor external segments (OS) due to the accumulation of dense material and RPE thickening, which suggested a temporary RPE dysfunction. In the course of a few weeks the said authors observed the normalization of the fovea with only residual RPE dysfunction. In our case, at baseline the IS/OS line was preserved with accumulation of a dense and homogeneous subfoveal material which in addition did not allow us to differentiate the RPE line at that level. The lesions were subfoveal and roundish, consisting in the accumulation of yellowish material about the RPE with discrete associated adopted pigment alterations. Subretinal liquid was not apparent and the appearance was vitelliform. In a large series of 90 cases, Freund et al.\(^5\) described various adult diseases which can express vitelliform lesions in the central macula. These are adult foveomacular dystrophy, cuticular drusen, non-exudative ARMD, vitreomacular traction and central serous chorioidopathy. These vitelliform deposits would represent the external segments (OS) of FR not digested by the dysfunctional RPE in these diseases.\(^5\) The subfoveal localization of this deposit could be due to the higher metabolic activity of the FR at that level which would bring out the RPE insufficiency. In the said series,\(^5\) the spontaneous resolution of the vitelliform lesions took place in 13.3% of these cases not before the 6 months follow-up. In addition, it was observed that the visual prognostic after resolution was related to the integrity of the IS/OS and the ELM in the foveal center.\(^5\) In our case, the clinical and morphological resolution of the condition was very fast, with an evolution of 1 month. This suggests acute RPE functional insufficiency which interfered with the normal phagocytosis and exchange of OS for a very short period of time. It was also observed that after the subfoveal material disappeared the external retina tomography returned to normal, with good functional recovery. It could be that suspending the OC facilitated this recovery.

By way of conclusion, this case is acute retinal epitheliitis in a young and healthy patient which coursed with a deposit of vitelliform material exclusively at the subfoveal level. Visual recovery was quick and complete, observing by means of SD-OCT the disappearance of said deposits and the anatomic recovery of the hyper reflective external retina layers.

Conflict of interest

No conflict of interest has been declared by the authors.

REFERENCES

