PHOTODYNAMIC THERAPY IN ADULT-ONSET FOVEOMACULAR VITELLIFORM DISTROPHY

TERAPIA FOTODINÁMICA EN DISTROFIA FOVEOMACULAR VITELIFORME DEL ADULTO

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ABSTRACT

Case report: We present the case of a patient who underwent photodynamic therapy (PDT) for adult-onset foveomacular vitelliform dystrophy (AFVD) at another clinic, having been misdiagnosed as having a choroidal neovascular membrane. Two years post-treatment, the patient had severe central visual loss and showed angiographic signs of choroidal hypoperfusion.

Discussion: AFVD is a rare clinical entity, usually having a fair visual prognosis, although some cases may show episodes of severe visual loss during their clinical course. Optical coherence tomography (OCT) is valuable in revealing the distinct images of AFVD from the features of a choroidal neovascular membrane. PDT using verteporfin may have negative long term effects if used in patients with AFVD (Arch Soc Esp Oftalmol 2007; 82: 117-120).

Key words: Photodynamic therapy, adult-onset foveomacular vitelliform dystrophy, optical coherence tomography, age-related macular degeneration, choroidal neovascular membrane.

RESUMEN

Caso clínico: Se describe el caso de un paciente al que se le realizó terapia fotodinámica con Verteporfin (TFD) en una distrofia foveomacular viteliforme del adulto (DFVA) en otro servicio al ser confundido con una membrana neovascular coroidea. La tomografía de coherencia óptica (OCT) es de gran ayuda en el diagnóstico diferencial. El paciente sufrió una disminución de la agudeza visual (AV) severa a los dos años del tratamiento con signos angiográficos de hipoperfusión coroidea.

Discusión: La DFVA es una entidad poco frecuente caracterizada por tener un buen pronóstico visual si bien existen casos con limitación de la AV severa en cortos periodos de tiempo en su historia natural. La OCT presenta una imagen claramente diferente respecto a la neovascularización coroidea. La terapia fotodinámica con Verteporfin puede no tener un efecto positivo a largo plazo en pacientes con DFVA.

Palabras clave: Terapia fotodinámica, distrofia foveomacular viteliforme del adulto, tomografía de coherencia óptica, degeneración macular asociada a la edad, membrana neovascular coroidea.
INTRODUCTION

Adult vitelliform foveomacular dystrophy (AVFD) is an infrequent disease with a dominant autosomic inheritance pattern. It is characterized by exhibiting a moderate reduction of vision in the adult age with good prognosis in most cases. The involvement is bilateral although unilateral cases have been described. The electrooculogram is normal or slightly altered. Biomicroscopically it is characterized by exhibiting a subretinal yellowish deposit in the macular area. This organization exhibits an angiographic image which can be confused with choroidal neovascularization. The optical coherence tomography (OCT) is helpful in the differential diagnosis.

CASE REPORT

A 65 year-old man with a history of two photodynamic therapy (PDT) sessions with Verteporfin and proposed for a third session visits the practice for obtaining an assessment. His present visual acuity is of 0.7 in right eye (RE), which is a single eye since the left eye (LE) is in phthisis bulbi due to a traumatic childhood accident. The patient did not refer changes since the beginning of the condition and did not noticed either visual acuity variations before the PDT sessions. The funduscopic image revealed a raised vitelliform injury in the foveal area with hard drusen in the posterior pole (fig. 1). The fluorescein angiography (FAG) revealed a progressive and intense ring hyperfluorescence in delayed time with a hypofluorescent central zone (figs. 2a and 2b). The indocyanine green angiography (IGA) did not exhibit alterations of interest. The OCT revealed the presence of a cuneiform structure of mid-high reflectivity between the pigmented epithelium and the neurosensory retina with detachment of the neurosensory retina surrounding the injury (fig. 3). An electrooculogram was performed, with results within the limits of normality.

The patient is diagnosed of AVFD and the decision is taken to follow the patient closely with regular checkups.
In the 3, 6 and 12 month checkups the test results did not exhibit variations with respect to the first visit and the initial visual acuity remains stable.

In the 2-year check the patient’s visual acuity was 0.02. The eye fundus exploration did not reveal the vitelliform injury but a rounded orange lesion is observed in the foveal area (fig. 4). Both the FAG and the AVI revealed an intense hypofluorescence maintained throughout the angiographic time (figs. 5a, 5b and 5c). The OCT showed a normal foveal depression with absence of the mean reflectivity mass identified in the first visit and an increase of posterior reflectivity at the height of the chorio-capillary epithelium complex (fig. 6).

Fig. 3: Mean reflectivity Injury located between the pigmented epithelium and the retina. A neurosensory wedge-shaped elevation can be seen next to the subfoveal material.

Fig. 4: Round orange injury in the foveal area where the vitelliform structure was located.

Fig. 5: Hypofluorescence maintained throughout the angiographic time due to choroidal hypoperfusion.
DISCUSSION

The angiographic behavior of the first visit can lead us to consider the existence of a subfoveal neovascular membrane due to the intense hyperfluorescence in delayed times. However, the OCT pattern is very characteristic of AVFD where the neurosensory elevation could be caused by the functional limitation of the pigmented epithelium due to the presence of the vitelliform injury. The OCT provides a clearly differentiating image between the AVFD and choroidal neovascularization (1,2). The absence of any detachments of pigmented epithelium and signs of neovascular activity, together with the presence of middle-high reflectivity material below the neurosensory retina leads us to think about AVFD in the presence of a neovascular membrane. Also, the absence of a high reflectivity structure between the pigmented epithelium and the resulting neurosensory retina of the fibrosis of the supposed choroidal neovascular membrane after two years confirmed the diagnosis.

The presence of a severe reduction of vision with intense angiographic hypofluorescence and the absence of the vitelliform material in the OCT suggests a choroidal hypoperfusion. It is difficult to know the degree of the PDT’s contribution in this condition since the visual decrease is detected after 2 years of performing the second session of treatment. There are bibliographical references describing the reabsorption of subretinal material after PDT and without reduction of visual acuity (3). The natural history of this disease also indicates the possibility of visual acuity reduction in short periods of time and not in a gradual way, as is more frequently observed in this condition (4). Some authors affirm that this treatment is innocuous, although their series solely cover one year of follow-up. In our case, after one year the patient did not exhibit any change with respect to the first visit. There are more extensive series with higher follow-up spans which were able to demonstrate a severe loss of visual acuity by choroidal hypoperfusion (5).

Therefore, we must clearly determine the angiographic and tomographic behavior of AVFD because to confuse it with a choroidal neovascularization and to establish treatment with PDT could possibly cause a severe and irreversible visual acuity reduction.

REFERENCES