Short communication

Intravitreal ranibizumab for choroidal neovascularisation associated with adult-onset vitelliform dystrophy


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ABSTRACT

Case report: A 70-year-old male patient diagnosed with bilateral adult-onset vitelliform dystrophy presented with a sudden decrease of vision in his left eye (LE) associated with the appearance of an occult type of neovascular membrane. It was treated with intravitreal ranibizumab due to juxtafoveal location of the membrane. Two injections were needed to induce total regression of the lesion.

Discussion: Intravitreal ranibizumab may be effective to induce morphological and functional improvement in cases of choroidal neovascularization secondary to adult-onset vitelliform foveomacular dystrophy. Further case series are required to confirm this observation.

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Ranibizumab intravitreo en neovascularización coroidea secundaria a distrofa foveomacular vitelliforme del adulto

RESUMEN

Caso clínico: Varón de 70 años diagnosticado de distrofa foveomacular vitelliforme del adulto (DFVA), que en el curso de su enfermedad presenta disminución brusca de visión en OI coincidiendo con la aparición de una membrana neovascular oculta. Dada la localización juxtafoveal de la membrana, se decidió tratar con ranibizumab intravitreo, siendo necesarias 2 inyecciones para lograr el cierre completo de la lesión neovascular.

Discusión: El uso de ranibizumab intravitreo puede ser una opción de tratamiento eficaz en la neovascularización coroidea secundaria a DFVA, siendo necesarias series de casos más amplias, para poder confirmar esta observación.

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Introduction

Adult-onset foveomacular vitelliform dystrophy (AOFVD), also known as pseudo-vitelliform dystrophy, is an infrequent disease characterized by the appearance of a yellowish subretinal lesion in the macular area, similar to the lesion of Best’s disease but of smaller size (0.5–1 papillary diameters). The lesion is caused by the deposit of eosinophile material and PAS(+) between the Bruch membrane and the retina. It usually involves both eyes although asymmetrically\(^1\) and it appears in the adult age (35–55 years). In addition, it responds to a dominant autosomic inheritance pattern although many sporadic cases have been described.\(^2\)

The visual acuity (VA) of these patients is usually preserved although it slowly deteriorates with the passage of years up to the first stages of the disease when the degradation of the vitelliform lesion finally produces retinal atrophy, significantly compromising vision in this stage.\(^1\)

Macular dystrophies, particularly those who give rise to alterations at the level of the Bruch membrane, can be considered as a risk factor for the development of choroidal neovascular membranes (CNVM).\(^3\) We present the case of a patient diagnosed with AOFVD, treated with intravitreal ranibizumab due to choroidal neovascularization associated to satisfactory anatomic and functional results.

Clinic case

Male, 70, referred due to progressive vision reduction in right eye (RE) and relative central scotoma with 6 months evolution. No personal or familial ophthalmological history was referred. Diabetes mellitus type 2 with 3 years of evolution and good metabolic control, and arterial hypertension in treatment.

The best corrected VA in the initial exploration was 4/10 in RE (+1.75; −1.00 × 95°) and 8/10 in the LE (+1.75; −1.50 × 85°).

Fig. 2 – Right eye fluorescein angiography: hyper-fluorescent lesion with central hypo-fluorescence.

Fig. 3 – (A) Right eye retinography: retinal atrophy patch after the disappearance of the vitelliform lesion. (B) Optic coherence tomography (OCT) showing the retinal pigment epithelium (RPE) thinning and atrophy with increased reflectiveness of the underlying choroids.

Fig. 1 – (A) Right eye retinography: vitelliform-like raised macular lesion. (B) Optic coherence tomography: macular subretinal cup-shaped thickening with foveal depression raising and rectification.
Anterior pole biomicroscopy was normal. The ocular fundus presented a yellowish lesion, raised in the macular area of the RE (Fig. 1A) and macular pigment alteration in the LE. No signs of diabetic retinopathy were found in either eye. Optic coherence tomography (OCT) revealed in the macular area of the RE the presence of a mass with average reflectiveness above the pigment epithelium strip with raised neurosensory retina, without evidencing subretinal or intra-retinal fluid (Fig. 1B). Fluorescein angiography (FAG) evidenced the presence of a hyper-fluorescent lesion with a hypo-fluorescent center, with increased intensity in late stages but without contrast dispersion (Fig. 2), which confirmed the AOGFVD diagnostic. Electrophysiological tests, including electoretinogram and electro-oculogram, were normal.

Regular ophthalmological checkups were established, which evidenced the progressive regression of the vitelliform lesion and the evolution toward atrophy of the macular pigment epithelium in the RE (Fig. 3). Vision remained stable in the LE (7/10), even though the macular pigmentary alterations were increasingly evident.

Two and a half years after the first visit, the patient visited the practice due to sudden vision reduction in the LE (2/10), together with the appearance of a serous-hemorrhagic macular detachment in the same eye. OCT revealed an important macular raising with pigment epithelium detachment, intraretinal cysts and presence of subretinal liquid. FAG confirmed the existence of a hidden neovascular membrane with late stain diffusion (Fig. 4). At this point it was decided to treat the LE with intravitreal ranibizumab at the dosis routinely applied for treating exudative macular degeneration (0.5 mg/0.05 ml).

Four weeks after the injection, VA had improved to 3/10 in the LE with a virtually complete reabsorption of the macular hemorrhage. A small amount of SRL was evidenced in the tomography (Fig. 5). After 8 weeks, VA was of 4/10 and the OCT showed an important reduction of the central macular thickness with a minimum of subretinal fibrosis. In the three-month checkup, vision in the LE had diminished (2/10) while the OCT evidenced an increased macular thickness exceeding 100μm, with presence of SRL. In the light of these findings, a second injection of ranibizumab was decided. The patient response to this second injection was favorable and at week 12 the subretinal fluid had reabsorbed completely, leaving a small subretinal fibrosis derived from the closure of the neovascular membrane (Fig. 6).

Twelve months after the last injection, VA remains stable in the LE (6/10), without signs of membrane reactivation or complications derived from the intravitreal treatment.

Fig. 4 – (A) Left eye retinography: macular sero-hemorrhagic detachment. (B) Left eye fluorescein angiography image (FA): hidden neovascular membrane. (C) LE optic coherence tomography (OCT): significant macular raising with significant PED, intraretinal cysts and presence of subretinal liquid. PED, pigment epithelium detachment.
The angiographic pattern of AOFVD, in cartwheel shape with hyper-fluorescence ring surrounding a central hypofluorescent area is quite reminiscent of—and can be confused with—a hidden neovascular membrane. For this reason, OCT has become an essential tool in the differential diagnostic of AOFVD and age related macular degeneration (ARMD) because it demonstrates the exact location of the pseudo-vitelliform material as a structure with medium-high reflectiveness above the retinal pigment epithelium (RPE), separating it from the photoreceptor layer. In contrast with serous detachments which appear in exudative ARMD, our case exhibited raised neurosensory retina without RPE raising, which could be clearly identified under the deposit although slightly thinner when compared to the adjacent retina. In addition, in the presence of neovascular lesions, vitelliform material does not produce the screen or cascade effect over the underlying choriocapillary.

Recently, the use of antiangiogenic drugs (anti-VEGF) has been described in the treatment of vitelliform lesions with contradictory results and without correlation between the anatomic and functional results obtained. The appearance of CNVM in the context of AOFVD is an infrequent complication. In the few documented cases found in the literature on choroidal neovascularization associated to macular dystrophies, such as Best disease, treatment with antiangiogenics has given good results.

This is the first case describing the use of intravitreal ranibizumab (Lucentis®) for treating choroidal neovascularization associated to AOFVD. In our case, a limited number of injections sufficed to achieve the complete closure of the neovascular lesion. However, larger series with longer follow-up periods are necessary in order to confirm this observation.

Conflict of interests

No conflict of interests was declared by the authors.

REFERENCES