Short communication

Bevacizumab treatment for acquired vitelliform detachment in patient with cuticular drusen

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ABSTRACT

Case report: We report a case of a 30-year-old male with acquired vitelliform detachment (AVD) secondary to cuticular drusen and suffering from metamorphopsia in his right eye. Intravitreal bevacizumab (Avastin) was administered, achieving successful results.

Discussion: It is an independent disease, of unknown genetic phenotype, caused by a generalized dysfunction of the retinal pigment epithelium (RPE). About 50% of patients develop AVD, and a correct diagnosis can be made with the help of new complementary tests. With no effective treatment currently available, and because of the incidence of developing choroidal neovascularization (NVC), treatment with anti-VEGF could help stabilize or improve the disease functionally and/or anatomically.

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Desprendimiento viteliforme adquirido en paciente con drusas cuticulares tratado con bevacizumab

RESUMEN

Caso clínico: Varón de 30 años, diagnosticado de desprendimiento viteliforme adquirido (DVA) secundario a drusas cuticulares que presentaba metamorfopías en su OD. Se trató con inyecciones intravitrales de bevacizumab (Avastin), respondiendo favorablemente.

Discusión: Enfermedad independiente, de fenotipo genético aún desconocido, debida a una disfunción generalizada del epitelio pigmentario retiniano (EPR). Evoluciona en un 50% a DVA; con la ayuda de nuevas pruebas complementarias llegamos a un diagnóstico certero. Sin tratamiento efectivo hasta el momento. Dada la frecuencia con la que se desarrolla


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Introduction

Pseudo-vitelliform detachment is an acquired vitelliform-like detachment located in the macula in the context of multiple diseases (pattern dystrophies of the retinal pigment epithelium [RPE], basal cuticular drusen, reticular pseudo-drusen, idiopathic polymorphic exudative vitelliform dystrophy and various macular dystrophies associated to macular disease).\(^1\)

The acquired vitelliform detachment (AVD) associated to basal cuticular or laminar drusen was described by Gass in 1985. Cuticular drusen would be expressed as abundant small yellowish and semi-translucent lesions in the macular area of young adult patients. Their evolution associates to AVD, formation of choroidal neovascularization and chorioretinal atrophy Plates.\(^3\)

Clinical case

Male, 30, who visited the emergency section due to sudden onset metamorphopsia in RE.

No relevant personal and familial antecedents. All first degree relatives were assessed without finding signs of the disease. Best corrected visual acuity (BCVA) was of 0.6 in both eyes (BE). Ocular tension and anterior pole biomicroscopy were normal.

Funduscopy: abundant small yellowish lesions (drusen) disseminated throughout the anterior pole in BE. Central raised injury with yellowish cloudy material (AVD) in LE.

Autofluorescence (FA): small hypoautofluorescent lesions surrounded by hyperautofluorescent ring (drusen). The pseudo-vitelliform material appears hyperautofluorescent as it contains lipofuscin or a similar material.

Fluorescein angiograph (FA): hyperfluorescent lesions (drusen) next to an initially hypofluorescent inferior lesion (due to the obstruction produced by the pseudo-vitelliform material) and subsequently hyperfluorescent due to slow and late filling (Figs. 1 and 2).

Optic coherence tomography (OCT; Cirrus HD-OCT [Carl Zeiss Meditec]): hyper-reflective sawtooth image (cuticular drusen), next to soft drusen in the pigment subepithelial space. Neuroepithelium detachment in BE, more extensive and with more reflective material in its core in LE (Figs. 3 and 4).

Fig. 1 – Small yellowish lesions disseminated throughout the posterior pole, shown in FA as hypoautofluorescent lesions surrounded by hyperautofluorescent ring, showing transmission hyperfluorescence (drusen) in FA over a dark background.
Fig. 2 – Similar appearance to RE, next to a central yellowish cloudy material (AVD); this pseudo-vitelliform material is shown in FA as hyperautofluorescent as it contains lipofuscin or similar material and in FA: inferior hypofluorescent lesion due to pseudovitelliform material obstruction with slow and late filling hyperfluorescence.

Fig. 3 – evolution OCT before and after RE treatment, showing sawtooth hyperreflectiva RPE-CC hyper-reflective strip modulations. Diminished AVD.
Due to the extensive metamorphopsia exhibited in RE, it was decided to treat with 3 anti-VEGF intravitreal injections (bevacizumab [Avastin]), observing functional and anatomical improvement. After verifying a positive result in the RE, the same treatment was repeated in LE, observing slight functional but not anatomical improvements. BCVA improved to 0.8 in RE and 0.7 in LE, while metamorphopsia disappeared in RE (Figs. 3 and 4).

Discussion

Basal cuticular or laminar drusen are described as a general RPE dysfunction which express ophthalmoscopically as multiple and small lesions (25–75 μm) disseminated throughout the posterior pole. Histologically, it was believed these were nodular excrescences of the internal portion of Bruch’s membrane. Russell et al. demonstrated that their location, structure and composition are similar to the drusen associated to age-related macular degeneration (ARMD). Their appearance is earlier (young adults) and half of cases develop acquired vitelliform detachment of a size exceeding 1, 500 μm larger than that appearing in the course of adult pseudo-vitelliform dystrophy. New imaging techniques (FA and OCT) enabled diagnostic precision. OCT allowed optimum identification of AVD walls, nodular images (drusen in the pigment subepithelial space) in the external and internal walls (neurosensory retina) undamaged over the drusen and AVD. Amorphous vitelliform material is generally highly reflective and even so OCT enabled positive visualization of adjacent structures. It is believed that this autofluorescent vitelliform material is made up of retinoids derived from phagocytosis of photoreceptor external segments.

Half of said AVD associated to cuticular drusen resolve spontaneously in a period of one or 2 years. However, the persistence of this AVD facilitates the development of CNV in one third of cases or could leave geographical atrophy plates with the ensuing loss of vision.

In 2007, Montero et al. and, in 2009 Lee et al. reported the efficacy of bevacizumab treatment in adult pseudo-vitelliform foveomacular dystrophy clinical cases. As the patient of this case was young and exhibited significant and sudden metamorphopsia in RE, and even though we did not objectively verify signs of CNV in any of the supplementary tests, we decided to treat with intravitreal bevacizumab injections. As in the above mentioned reports, we observed anatomical improvements and discrete functional improvement.

It has been demonstrated “in vitro” that RPE cells express the vascular endothelial growth factor (VEGF-A) and cuticular drusen are due to a generalized dysfunction of these cells. Accordingly, the beneficial role of anti-VEGF medication could be suggested. On the other hand, in this specific case, the enhanced anatomical response in RE could be due to greater RPE integrity and therefore of its function as a metabolic pump.

The authors consider that this treatment could be effective for speeding up the reaplication of AVD, diminishing atrophy eschars and possibly avoiding CNV formation.

Conflict of interests

No conflict of interests has been declared by the authors.

References

2. Russell SR, Mullins RF, Schneider BL, Hageman GS. Location, substructure and composition of basal laminar drusen compared with drusen associated with aging and

