Short communication

Intravitreal bevacizumab for choroidal neovascularization associated with Best's disease


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ARTICLE INFO

Article history:
Received 6 January 2013
Accepted 5 July 2013
Available online 22 October 2014

Keywords:
Best's disease
Best's vitelliform macular dystrophy
Choroidal neovascularization
Bevacizumab
Intravitreal injection

ABSTRACT

Case report: A 27-year-old woman presented with loss of vision in the right eye (20/200). Ophthalmoscopic examination showed intraretinal hemorrhage in the macular region with neurosensory detachment in the right eye, and vitelliform deposit in the left eye. Fluorescein angiography and the electrooculogram confirmed the diagnosis of choroidal neovascularization associated with Best's disease. Four weeks after a single bevacizumab intravitreal injection, visual acuity was restored (20/25) and remained stable after a 12 month follow-up.

Discussion: Intravitreal bevacizumab appears to be an effective treatment for choroidal neovascularization associated to Best's disease.

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Bevacizumab intravitreo en neovascularización coroidea asociada a enfermedad de Best

RESUMEN

Caso clínico: Mujer de 27 años que presentaba disminución de visión en ojo derecho (20/200). El examen funduscópico reveló una hemorragia intraretiniana macular con desprendimiento neurosensorial en ojo derecho, y un depósito de material viteliforme en el ojo izquierdo. La angiografía fluoresceínica y el electrooculograma confirmaron el diagnóstico de neovascularización coroidea asociada a enfermedad de Best. Cuatro semanas después de una única inyección de bevacizumab intravitreo, la agudeza visual a la normalidad (20/25) y se mantuvo estable tras 12 meses de seguimiento.

Discusión: El bevacizumab intravitreo puede ser una opción terapéutica eficaz en la neovascularización coroidea secundaria a enfermedad de Best.

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Introduction

Best’s disease is a dominant autosomal inheritance macular dystrophy caused by a mutation in the bestrophin protein, a transmembrane protein that forms a chloride channel in the retinal pigment epithelium cells (RPE). The alteration of this protein accounts for electrophysiological findings (Arden index = 1.5) as well as for the accumulation of lipofuscin on the RPE. In the classification of Best’s disease, the appearance of choroidal neovascularization is an infrequent complication, associated to late stages of the disease.  

Clinic case

A female, 27 years, was referred due to diminished vision in right eye (RE) with 2 weeks evolution. The patient did not refer personal or familial ophthalmological antecedents. The initial visual acuity (VA) was 20/200 in RE and 20/25 in the left eye (LE). RE funduscopic examination revealed an intraretinal hemorrhage with neurosensory retina elevation. The rest of the examination was normal (Fig. 1). LE ocular fundus revealed macular pigment alteration together with yellowish material sedimentation at the subretinal level (Fig. 1). RE optic coherence tomography (OCT-SD; model 3D-OCT-2000, Topcon) (Topcon Corporation, Tokyo, Japan) revealed irregularities in the interface between RPE and Bruch membrane, with intraretinal edema and neurosensory detachment (central macular thickness: 540 μm) compatible with choroidal neovascularization (CNV) (Fig. 2). In the LE, OCT showed atrophy of the RPE and underlying choriocapillary (Fig. 3). Fluorescein angiography evidenced pattern hyperfluorescence at the subfoveal level from early angiogram stages leaking fluorescein in late stages, confirming the existence of membrane with active CNV in RE (Fig. 4).

With the clinical suspicion of CNV associated to Best’s disease, electrophysiological tests were requested which confirmed the suspected diagnosis after finding in the electrooculogram a pathological Arden index in both eyes (1.39 in RE and 1.45 in LE).

In this context, intravitreal bevacizumab injection in RE was indicated (0.5 mg/0.05 ml). Four weeks after the injection, the patient recovered 7 lines of vision to reach RE BCVA of 20/25 and evidencing disappearance of the hemorrhage and subretinal fluid (Fig. 5). OCT revealed complete macular edema regression (central macular thickness: 243 μm), as well as the absence of intra- and subretinal fluid (Fig. 6).

Twelve months after said injection, vision remained stable without signs of CNV reactivation or complications derived from the intravitreous treatment.

Discussion

Despite the typical macular lesions exhibited by patients with Best’s disease, the majority maintain good VA throughout
their lifetime and therefore the diagnosis can be a casual finding during an ophthalmological checkup. Fishman et al. demonstrated in their series that 75% of the patients under 50 maintained VA ≥ 20/40 in at least one eye up to late stages of their lives. For this reason, if said patients experience diminished VA the presence of CNV must be discarded. OCT is useful for identifying the presence of subretinal fluid although fluorescein angiography continues to be the test of choice to confirm the existence of active CNV.

The treatment of CNV associated to Best’s disease is controversial due to the absence of randomized studies comparing different therapeutic options.

For cases which do not associate significant vision reduction or extensive hemorrhage, simple observation could be a valid proposal as stated by Chung et al. in a series of 11 patients in which, after 49 months follow-up, the mean BCVA remained at 20/50.

Photodynamic therapy could be another efficient alternative as demonstrated by Ozdek et al. after achieving vision improvement in 4 out of 5 treated cases after a mean follow-up of 25 months.

There are very few documented cases about CNV associated to Best’s disease treated with antiangiogenic drugs. When associated to Best’s disease, CNV usually requires a lower number of antiangiogenic injections (one or two according to different published series) than other types of macular dystrophies (pattern dystrophy, Sorsby macular dystrophy) which generally require at least 3 injections.

The case reported herein required a single injection to deactivate the CNV, achieving rapid visual recovery without side effects after a follow-up of 12 months.

Prospective studies are required with a larger number of cases to establish the most recommendable therapeutic option for these patients, although the efficacy and swift VA recovery prompted the authors to recommend the use of antiangiogenics in CNV patients associating Best’s disease.

**Conflict of interest**

No conflict of interests has been declared by the authors.

**REFERENCES**