Intravitreal ranibizumab for type 3 choroidal neovascularization complicating adult onset foveomacular vitelliform dystrophy

Traitement par ranibizumab de la néovascularisation choroidienne compliquant la dystrophie vitelliforme de l’adulte

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Summary

Purpose. — To describe the results obtained with intravitreal ranibizumab injections in a patient with adult onset foveomacular vitelliform dystrophy (AOFVD) complicated by Type 3 choroidal neovascularization (CNV).
Methods. — A 78-year old man diagnosed with AOFVD presented at our department for decreased vision in his left eye (LE) (20/80). Upon a complete ophthalmologic examination, including fluorescein angiography, indocyanine green angiography, and spectral-domain optical coherence tomography, the patient was diagnosed with Type 3 CNV. Three monthly injections of ranibizumab 0.05 ml/0.5 mg were administered intravitreally without complications.
Results. — After the first injection, visual acuity of the LE improved (20/64) and regression of the Type 3 CNV was observed by fluorescein angiography, indocyanine green angiography and OCT. Six months after the final ranibizumab injection, a more-or-less complete resolution of the exudative retinal changes was observed.

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Introduction

Adult onset foveomacular vitelliform dystrophy (AOFVD) is a relatively uncommon macular disease that shares phenotypic features with best vitelliform macular dystrophy. The disease progression is generally characterized by a slow development of macular atrophy. In some cases, AOFVD may be complicated by choroidal neovascularization (CNV) [1]. Based on the classification proposed by Gass [1], CNV may be defined as Type 1, when the neovascularization is confined to the sub-retinal pigment epithelium (RPE) space, and as Type 2 when the neovascularization proliferates above the RPE in the subneurosensory space. In 2001, Yannuzzi et al. [2] described a different type of neovascularization characterized by intraretinal proliferation and proposed to refer to it as retinal angiomatous proliferation (RAP). Freund et al. recently proposed to refer to this peculiar type of neovascularization as Type 3 CNV [3]. Intravitreal ranibizumab (Lucentis, Genentech Inc, South San Francisco, California, USA) was the first therapy for each type of CNV (Type 1, Type 2 and Type 3) secondary to age-related macular degeneration (AMD) to show in 2 Phase III clinical studies an improvement in mean visual acuity [4,5]. Here we report on a patient with AOFVD, who developed type 3 CNV. The patient was treated by intravitreal ranibizumab injections.

Case report

A 78-year-old man diagnosed with AOFVD presented at our department for decreased vision in his left eye (LE). Best-corrected visual acuity (BCVA) was 20/40 in the right eye (RE) and 20/80 in the LE. On fundus biomicroscopy, both eyes showed a yellowish lesion (the vitelliform material) within the macula. On fundus autofluorescence (FAF), the vitelliform material appeared as hyper-autofluorescent in both eyes (Fig. 1); some hypo-autofluorescent changes were evident in the LE macula. Spectral-domain optical coherence tomography (SD-OCT) macular scans of both eyes showed the vitelliform material as a highly reflective dome-shaped lesion located between the photoreceptor layer (inner segments (IS)/outer segments (OS) junction) and retinal pigment epithelium (RPE). On SD-OCT, the left eye IS/OS junction appeared markedly disrupted in the LE. Interestingly, fluorescein angiography (FA) (Fig. 1) and indocyanine green angiography (ICGA) (Fig. 2) of the LE showed the presence of a vascular communication between the retinal and choroidal circulation, as well as a late focal hyperfluorescence (“hot spot”) in the upper perimacular area, suggesting the presence of Type 3 CNV. SD-OCT scan referenced on the ICGA hot spot confirmed the suspicion of Type 3 CNV in the LE, showing a small localized elevation of the RPE, the “flap sign” [6], and the axonal communication between...
Figure 1. Fundus autofluorescence (FAF) of the right eye (A) and left eye (B) showing the vitelliform material as hyperautofluorescent; some hypo-autofluorescent changes are evident in the left macula (B). Spectral-domain optical coherence tomography (SD-OCT) macular scans of both eyes (C and D) show the vitelliform material as a highly reflective dome-shaped lesion located between the photoreceptor layer (inner segments (IS)/outer segments (OS) junction) and retinal pigment epithelium (RPE). On SD-OCT, the left eye IS/OS junction appeared markedly disrupted. Fluorescein angiography (E and F) shows inhomogeneous hyper-fluorescence within the macula of both eyes, and a focal hyperfluorescence in the upper perimacular area of the left eye (arrowhead).

Discussion

CNV (Type 1/Type 2) is a rare complication of AOFVD. In this report, we describe an unusual association between AOFVD and Type 3 CNV in a 78-year-old man, treated by intravitreal ranibizumab injections. To the best of our knowledge, this case represents the first demonstration of Type 3 CNV associated with AOFVD. Recently we reported a case of Type 3 CNV complicating fundus flavimaculatus, which was treated by intravitreal ranibizumab injections [7]. For this reason we suggest that Type 3 CNV should not be considered as a complication exclusive of AMD.

Treatment of Type 3 CNV secondary to AMD generally shows poor anatomical and functional outcomes. Direct laser photocoagulation of the vascular lesion, laser photocoagulation of the feeder vessel, photodynamic therapy, and transpupillary thermotherapy rarely allow the anatomical closure of the lesion. Recently, encouraging short-term results have been reported using anti-VEGF drugs in the treatment of these lesions.

As suggested by Freund et al. [3], early in its evolution, Type 3 CNV appears exquisitely sensitive to intravitreal anti-VEGF, and a complete lesion regression is possible after the first injection.

In our patient, Type 3 CNV complicating AOFVD was diagnosed early in its evolution ("flap sign") [6], and, intravitreal ranibizumab succeeded in halting the progression of the CNV-related disease, at least in the short-term. Our report is limited by the short follow-up. In fact, efficacy of intravitreal ranibizumab is limited in time, especially in Type 3 CNV. Moreover, we acknowledge that no conclusion can be drawn from a single case, but based on our findings, intravitreal ranibizumab may be considered as a therapeutic option for the rare association between type 3 CNV and AOFVD.

Disclosure of interest

The authors declare that they have no conflict of interest concerning this article.
Figure 2. Indocyanine green angiography (ICGA) of the left eye (A and B) showing a late focal hyperfluorescence ("hot spot") (arrowhead) in correspondence of the Type 3 choroidal neovascularization (CNV). Spectral-domain optical coherence tomography (SD-OCT) scan (C) actually referenced on the ICGA (A) hot spot (Type 3 CNV) shows a small localized elevation of the retinal pigment epithelium (RPE) and the "flap sign" (asterisks), as well as the associated intraretinal exudative changes (C). Late ICGA frame showing resolution of the "hot spot" (open arrowhead) one month after the third and last injection of ranibizumab (D and E). SD-OCT (F) scan referenced on the ICGA frame (D) shows, one month after the third and last injection of ranibizumab, still a small elevation of the RPE (arrow), but complete resolution of the associated exudative retinal changes.

References