Ranibizumab in the treatment of choroidal neovascularization on the border of an inferior staphyloma associated with tilted disc syndrome

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Purpose: To describe a case of choroidal neovascularization (CNV) on the border of an inferior staphyloma associated with tilted disc syndrome treated with intravitreal ranibizumab.

Patients: Observational case report.

Methods: A patient with CNV on the border of an inferior staphyloma associated with tilted disc syndrome was imaged using fluorescein angiography, autofluorescence and spectral domain optical coherence tomography, and treated with intravitreal injections of ranibizumab.

Results: The patient received 3 ranibizumab injections during the 9-month follow-up. The visual acuity improved from 20/40 to 20/32 and the foveal thickness reduced from 470 microns to 248 microns. The angiograms showed resolution of leakage associated with CNV. There were no adverse events.

Conclusion: Intravitreal ranibizumab is an efficacious and safe treatment in the management of choroidal neovascularization on the border of an inferior staphyloma associated with tilted disc syndrome.

Keywords: intravitreal ranibizumab, choroidal neovascularization, tilted disc syndrome, inferior staphyloma

Introduction

The main features that characterize eyes with tilted disc syndrome are oval optic disc with oblique axis, inferonasal crescent, situs inversus of the retinal vessels, myopic astigmatism and visual field defects.1–3 In addition, an inferior staphyloma is often associated with this syndrome and severe complications such as choroidal neovascularization (CNV) can occur when the border of the staphyloma lies across the macula.4–7 This CNV usually shows a classic pattern on fluorescein angiography (FA) resulting in significant visual loss.

Ranibizumab (Lucentis®; Genentech Inc, South San Francisco, California, USA) is a humanized antibody fragment designed to bind and inhibit all vascular endothelial growth factor-A (VEGF-A) isoforms. It was approved by the Food and Drug Administration (FDA) for the treatment of wet age-related macular degeneration in June 2006. It has shown a great efficacy in the management of all angiographic subtypes of lesions in different clinical trials.8,9

We present a case of CNV on the border of an inferior staphyloma associated with tilted disc syndrome successfully treated with intravitreal ranibizumab.

Case report

A 77-year-old woman complained about visual acuity (VA) loss in her left eye in the previous 3 weeks. She had an uncomplicated cataract surgery in both eyes 4 years...
At the moment of the examination her refractive error was 90° −1.00 + 0.25 in her right eye and 90° −0.75 + 0.50 in her left eye. The axial length was 22.9 mm in the right eye and 23.2 mm in the left eye. Her VA measured with ETDRS

Figure 1 At baseline, the fundus photograph of the left eye revealed a tilted disc and the border of an inferior staphyloma lying across the macula and a subretinal lesion (a). Autofluorescence image showed mild hyperautofluorescence at the temporal border of the macula (b). Fluorescein angiography showed a well-defined predominantly classic choroidal neovascularization (CNV) in an early phase with leakage in a late phase (c, d). Spectral domain optical coherence tomography scanning showed an irregular and elevated retinal pigment epithelium band with overlying CNV associated with intraretinal and subretinal fluid (e).
(“Early Treatment of Diabetic Retinopathy Study”) charts was 20/20 in the right eye and 20/40 in the left eye. The anterior segment examination showed transparent corneas and clear intraocular lenses located in the posterior chamber. The intraocular pressure was 16 mmHg in both eyes. The fundus examination revealed tilted discs and the border of an inferior staphyloma lying across the maculas in both eyes. Imaging with FA (Imagenet®; Topcon Corporation, Tokyo, Japan) showed an active predominantly classic CNV and a window defect on the border of the staphyloma. OCT scanning (OCT 3-D, Topcon Corporation, Tokyo, Japan) showed an irregular and elevated retinal pigment epithelium (RPE) band with

![Image](image_url)

**Figure 2** After three intravitreal injections of ranibizumab, the fundus photograph of the left eye revealed a flat macula with pigmentary changes (a). Autofluorescence image showed mild hyperautofluorescence at the temporal border of the macula (b). Fluorescein angiography showed staining but no leakage from the choroidal neovascular membrane at the early phase and the late phase as well (c, d). Spectral domain optical coherence tomography scanning showed no intraretinal or subretinal fluid with restoration of the foveal contour (e).
over any additional injection during the 9-month follow-up period. Nevertheless, it is likely that further treatment will be required with longer follow-up. We did not have any ocular or systemic side effect related to the drug or the route of administration.

The VA improved by one line, from 20/40 to 20/32. In comparison, a much more substantial anatomical benefit was observed, with resolution of the angiographic leakage and the intraretinal and subretinal fluid detected on OCT scans. However, it is known that visual gaining is often limited in predominantly classic lesions growing between the photoreceptors and the RPE.

Photodynamic therapy with verteporfin could have been used as an alternative treatment in this patient. However, potential RPE damage could have limited visual recovery. Bevacizumab could have been used as an alternative treatment. However, it is not an approved drug for intraocular use in Spain. RPE rips have been reported with ranibizumab and bevacizumab as well. To our knowledge, no data support reduced risk of RPE rips with either ranibizumab or bevacizumab.

In summary, ranibizumab therapy can be used as an efficacious and safe treatment in the management of CNV associated with tilted disc syndrome. Nevertheless, this is only supported by a single case report. It would be desirable to accumulate more data to demonstrate potential safety and efficacy of ranibizumab in this condition.

**Disclosures**

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Dr Arias had full access to all the data in the study and takes responsibility for the integrity of the data and the accuracy of the data analysis as well as the decision to submit for publication.

**References**


