Ranibizumab for choroidal neovascularization following atrophic involution of adult-onset foveomacular vitelliform dystrophy.

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Abstract
We report a case of adult-onset foveomacular vitelliform dystrophy complicated with atrophic involution and subsequent choroidal neovascularization and treated with intravitreal ranibizumab.

A 71-year-old woman diagnosed with adult-onset foveomacular vitelliform dystrophy experienced a progression of the vitelliform lesion to the atrophic stage in her left eye immediately after cataract surgery. Best-corrected visual acuity decreased from 0.5 to 0.16. Ten months later, the patient was referred with new worsening of her visual acuity (0.05) and metamorphopsia. The spectral-domain optical coherence tomography confirmed the presence of type 2 choroidal neovascularization next to the atrophic area. The patient was treated with 2 monthly injections of ranibizumab. Spectral-domain optical coherence tomography images improved significantly and her best-corrected visual acuity increased to 0.20 associated with subjective improvement of metamorphopsia.

Ranibizumab may be effective for choroidal neovascularization in patients with adult-onset foveomacular vitelliform dystrophy.

Key words: Adult-Onset Foveomacular Vitelliform Dystrophy; Retinal Atrophy; Choroidal Neovascularisation; Ranibizumab

Resumen
Presentamos el caso de una paciente con distrofia foveomacular viteliforme del adulto que presentó una evolución atrófica complicada con neovascularización coroidea tratada con ranibizumab.

Mujer de 71 años diagnosticada de distrofia foveomacular viteliforme del adulto, que tras la cirugía de catarata en el ojo izquierdo, presentó una evolución atrófica en dicho ojo. Su mejor agudeza visual corregida disminuyó desde 0.5 a 0.16. Diez meses más tarde, la paciente viene referida de nuevo por empeoramiento de su visión, siendo 0.12, y metamorfopsia. La tomografía de coherencia óptica de dominio espectral confirmó la presencia de una neovascularización coroidea tipo 2 que se acompañaba de una leve exudación próxima a la zona de atrófia. La paciente fue tratada mediante 2 inyecciones mensuales de ranibizumab.

Las imágenes tomográficas mejoraron significativamente y su agudeza visual corregida mejoró hasta 0.2, así como una mejora subjetiva de la metamorfopsia.

El tratamiento con ranibizumab de la
neovascularización coroidea en pacientes con distrofia foveomacular del adulto puede ser efectivo, requiriendo de controles periódicos.

**Palabras clave:** Distrofia foveomacular viteliforme del adulto, atrofia, neovascularización coroidea, ranibizumab

**Introduction**

Adult-onset foveomacular vitelliform dystrophy (AOFVD) is a rare macular disease that shares certain phenotypic features with the vitelliform stage of Best dystrophy. However, electrophysiological changes in AOFVD usually evidence normal or subnormal responses, whereas Arden index alteration is characteristic of Best dystrophy. Dilated fundus examination (DFE) typically shows a bilateral yellowish ovoid subretinal lesion, frequently larger than one papillary diameter, which usually results in slow progressive moderate vision loss. Further visual impairment may be severe when the lesion progresses to the atrophic stage or less frequently when the patient develops choroidal neovascularization (CNV).

Herein we report a case of AOFVD with atrophic involution complicated with CNV and treated with intravitreal ranibizumab.

**Case Report**

A 71-year-old woman clinically and electrophysiologically, electrooculogram (EOG) showed normal results (Arden index was 1.95), making the difference with the severe alteration of this electrophysiological test present in Best disease, diagnosed with AOFVD, experienced a progression of the vitelliform lesion to the atrophic stage in her left eye (OS) following immediately an uneventful cataract surgery. Best corrected visual acuity (BCVA) decreased from 0.5 preoperatively to 0.16 after surgery in her OS. DFE showed complete disappearance of the vitelliform lesion which was replaced by a geographic atrophy of the foveal retinal pigment epithelium.

Ten months later, the patient was referred to our department complaining of new worsening of her BCVA OS (0.05) associated with metamorphopsia. DFE revealed the presence of macular haemorrhages and...

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**Figure 1:** A. Colour fundus photograph showed foveal atrophic changes with subretinal fibrosis within the temporal edge of the lesion and laminar haemorrhages. B. Fundus autofluorescence image highlighted the hypoautofluorescent atrophic area, surrounded by fibrous tissue. C. Fluorescein angiography revealed the presence of a predominantly classic choroidal neovascularization (CNV). D. Spectral-domain optical coherence tomography images showed type 2 CNV in close relationship to the temporal edge of the atrophic area. E. Following two intravitreal injections of ranibizumab, SD-OCT showed complete resolution of the intraretinal fluid.
exudation due to the development of CNV next to the temporal margin of the atrophic area. Spectral-domain optical coherence tomography (SD-OCT) confirmed the presence of type 2 CNV. Treatment with 2 consecutive monthly intravitreal injections of ranibizumab was administered. SD-OCT images evidenced reduction of the retinal swelling, and BCVA improved to 0.20 with subjective improvement of metamorphopsia (Figure 1).

Discussion

The use of vascular endothelial growth factor (VEGF) inhibitors has been reported in cases of AOFVD with variable results.3-4 It has also been reported the treatment of CNV with ranibizumab in many other macular conditions associated with CNV such as high myopia5, angioid streaks6, pattern dystrophy7, retinal astrocytic hamartoma8, choroidal hemangioma9, traumatic Bruch membrane rupture10, fundus flavimaculatus11, or Stargardt disease12. They all share the same tomographic appearance of type 2 neovascularization (growing above the RPE within the subretinal space). The use of ranibizumab for CNV related to AOFVD has previously been reported13,14 with positive outcomes, therefore offering a possible therapy for this condition.

In conclusion anti-VEGF drugs may be effective for CNV related to AOFVD albeit cases with severe disruption of the outer retinal layers or atrophic changes should have a limited visual improvement. Extended follow-up is warranted to assess possible recurrences and determine the need of retreatments in order to achieve the control of the CNV associated with AOFVD.

REFERENCES