Short communication

Choroidal neovascularization secondary to angioid streaks: A familial case report


Servicio de Oftalmología, Hospital Universitario Príncipe de Asturias, Alcalá de Henares, Madrid, Spain

A R T I C L E   I N F O

Article history:
Received 15 February 2012
Accepted 7 November 2012
Available online 6 August 2014

Keywords:
Angioid streaks
Choroidal neovascularization
Ranibizumab
Lucentis
Pseudoxanthoma elasticum

A B S T R A C T

Case report: We report a familial case of 2 siblings that suffered choroidal neovascularization (CNV) secondary to angioid streaks. They were both treated with a monthly intravitreal injection of ranibizumab (Lucentis®) for 3 months. Visual acuity was stabilized and fluorescein angiography revealed complete resolution of CNV. Neither recurrent CNV lesion nor new hemorrhages were reported during the follow-up period.

Discussion: The use of intravitreal ranibizumab for the treatment of CNV in patients with angioid streaks has shown favorable results. However, further studies with a longer follow-up and larger number of patients are necessary to more precisely determine the results of this therapy.

© 2012 Sociedad Española de Oftalmología. Published by Elsevier España, S.L.U. All rights reserved.

Neovascularización coroidea secundaria a estrías angioides: un caso familiar

R E S U M E N

Caso clínico: Se expone el caso de dos hermanos que presentaron neovascularización coroidea asociada a estrías angioides. Ambos pacientes fueron tratados con una inyección mensual de ranibizumab (Lucentis®) intravitrea durante tres meses. La visión se estabilizó y la angiografía fluoresceínica mostró resolución completa de la neovascularización coroidea. Durante el seguimiento, no observamos recurrencia del cuadro.

Discusión: El uso de ranibizumab intravitreo en caso de neovascularización coroidea asociada a estrías angioides ha mostrado resultados favorables. No obstante, serán necesarias series de casos mayores que nos permitan conocer la verdadera eficacia de este tratamiento.

© 2012 Sociedad Española de Oftalmología. Publicado por Elsevier España, S.L.U. Todos los derechos reservados.


* Corresponding author.
E-mail address: jbenitezherreros@hotmail.com (J. Benitez-Herreros).

2173-5794/$ – see front matter © 2012 Sociedad Española de Oftalmología. Published by Elsevier España, S.L.U. All rights reserved.
Introduction

Angioid streaks (AS) are a dehiscence of the Bruch membrane which radiate from the papilla, the main complication being the development of choroidal neovascular membranes (CNVM). AS could be associated to systemic diseases, although the most frequently associated disease of AS is pseudoxanthoma elasticum (PXE), an autosomic recessive disease caused by the mutation of gene ABCC6. Other diseases which AS could associate include the Ehlers-Darlos syndrome, of dominant autosomic inheritance, Paget’s disease (of unknown origin) and sickle cell anemia of recessive autosomic inheritance.

Clinical cases

Two siblings with AS and PXE who developed CNVM in 3 out of 4 eyes.

Case 1

Female, 51 years, who referred metamorphopsiae in left eye (LE). Visual acuity (VA) was of 20/20 in both eyes (BE). Left ocular fundus (OF) exhibited extrafoveal hemorrhages and hard drusen above the superior temporal arch, while angiofluorograph (FA) revealed extrafoveal hyperfluorescent area corresponding to CNVM, and macular optic coherence tomography (OCT) showed absence of intraretinal liquid (Fig. 1). The therapeutic options were explained to the patient, who decided to maintain an expectant attitude. Nine months later, when the condition progressed and affected the foveal area (Fig. 2) and VA diminished to 20/200, she accepted treatment with ranibizumab (Lucentis®; Genentech, Inc., South San Francisco, CA, USA). After one injection per month during 3 months, LE VA remained at 20/200 and FA revealed inactivity in the lesion. Six months later, LE remained stable but metamorphopsiae began to appear in the right eye (RE). VA was 20/20. OF exhibited extrafoveal hemorrhages which, under FA, corresponded to CNVM, while OCT revealed absence of intraretinal liquid (Fig. 3A–C). On this occasion, the patient accepted treatment with ranibizumab. After 3 injections, VA continued at 20/20, metamorphopsiae had disappeared and FA exhibited no activity (Fig. 3D). Thirty months later, the condition remained stable in BE.

Case 2

Male, 57 years, brother of the Case 1 patient, referred metamorphopsiae in LE and finger-counting VA at 1m in LE. OF exhibited intense chorioretinal atrophy and sub- and extrafoveal hemorrhages with absence of drusen, while FA revealed sub- and extrafoveal hyperfluorescent sites and OCT revealed the presence of intraretinal fluid (Fig. 4A–C). Three ranibizumab injections were prescribed after which metamorphopsiae disappeared, VA remained stable and both FA and OCT showed neovascular inactivity (Fig. 4D–E). The patient remained stable during the 18-month follow-up.

Discussion

The main complication of AS is the development of CNVM. Photodynamic therapy, transpupilar thermal therapy and

![Fig. 1](image-url) – Case 1, left eye: (A) retinograph: extrafoveal hemorrhages in LE fundus. (B) FA: hyperfluorescence at the AS level, in addition to extrafoveal loci which leaks contrast in late times. (C) OCT: preserved foveal depression; absence of intraretinal liquid.
surgery have not obtained good results in the treatment of said CNVM. In recent years, the injection of anti-VEGF drugs has become the predominant treatment. The use of bevacizumab seems to be effective even though there are very few published reports on the use of ranibizumab or pegaptanib.

The age of the 2 siblings with PXE and AS who developed CNVM was within the range in which CNVM develops in AS, but an earlier age in which neovascularization appears in ARMD. In addition to AS, the examination revealed macular RPE atrophy and some drusen. In this regard, other authors have described the presence of broad chorioretinal atrophy areas and drusen in patients with AS. Treatment with 3 ranibizumab injections stabilized the situation of the 3 eyes, deactivating neovascular lesions and maintaining previous VA. In addition, during the follow-up the condition did not reactivate.

The results in this case are similar to those obtained by Lazaros and González-Gómez, who prescribed a...
Accordingly, opting for a charge dose of 3 ranibizumab injections in smaller series of patients with AS and CNVM, the lesions were resolved and remained inactive during the follow-up of both studies (27 and 12 months, respectively); none of the cases exhibited worsened VA. Mimoun treated 35 eyes affected by AS and CNVM with a dose of ranibizumab, re-injecting the drug if the lesion became active. The average amount of injections was higher than in the present case (5.7 injections, 24 months follow-up). However, the 3 injections of this study are within the range of injections applied by said author (2–14). Other authors opted for one monthly injection of ranibizumab during one year (12 injections) in a series of eyes with AS and CNVM. However, their visual results did not improve those obtained in the studies in which, after an initial pattern of one or 3 injections, only one case of CNVM activity was reported. Accordingly, in order to optimize the number of injections it seems beneficial to repeat treatments with ranibizumab only in case of CNVM activity.

As can be seen, ranibizumab appears to be a useful therapeutic strategy to stabilize CNVM in AS. Even so, larger series are necessary to determine the true efficacy of said treatment.

**Conflict of interest**

The authors declare no conflict of interest.

**REFERENCES**


---

**Fig. 4** – Case 2, left eye: (A) pretreatment retinograph: intense chorioretinal atrophy, subfoveal and juxtafoveal hemorrhages. (B) Pretreatment FA: macular atrophy areas, hyperfluorescence at the level of AS, sub- and extrafoveal loci which leak contrast in late times. (C) Pretreatment OCT: intraretinal fluid in the form of cysts. (D) Posttreatment FA: macular atrophy areas and hyperfluorescence at the level of AS; contrast leak suggesting neovascular activity is not observed. (E) Posttreatment OCT: chorioretinal atrophy without accumulation of retinal fluid.