Short communication

Subretinal neovascular membrane in angioid streaks treated with intravitreal bevacizumab

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Abstract

Introduction: Angioid streaks are breaks in Bruch’s membrane that may be associated, among others, with pseudoxanthoma elasticum. Its most common complication is the development of subretinal neovascular membranes (SRNVM) and the decreased vision this entails.

Case report: A 28-year-old woman with angioid streaks and SRNVM in the left eye, who received 3 injections of intravitreal bevacizumab, with rapid improvement in vision and stability during 11 months follow-up. The finding of angioid streaks led to the diagnosis of pseudoxanthoma elasticum.

Conclusion: Intravitreal bevacizumab should be considered as an effective treatment option for choroidal neovascularization associated with angioid streaks.

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Membrana neovascular subretiniana asociada a estrías angioides tratada con bevacizumab intravítreo

Resumen

Introducción: Las estrías angioides son roturas en la membrana de Bruch que pueden asociarse, entre otras, a pseudoxantoma elástico. Su complicación más frecuente es el desarrollo de membranas neovascular subretinianas (MNVSR) con la disminución de visión que conlleva.

Caso clínico: Mujer de 28 años con estrías angioides y MNVSR en ojo izquierdo, que recibió tres inyecciones de bevacizumab intravitreo, con rápida mejora de la visión y estabilidad durante 11 meses de seguimiento. El hallazgo de estrías angioides permitió el diagnóstico de pseudoxantoma elástico.

Conclusion: El bevacizumab intravitreo debe ser considerado como una opción terapéutica eficaz para la neovascularización coroidea asociada a estrías angioides.

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Introduction

Angioid streaks are ruptures in Bruch’s membrane that appear in the ocular fundus in the form of radially extending grayish stripes around the optic nerve.\(^1\)

The majority of angioid streaks are idiopathic although they can associate to multiple systemic diseases, being pseudoxanthoma elasticum most frequent (85% of patients exhibit ocular involvement). Generally, angioid streaks are bilateral and accompany alterations such as “orange skin” and papillary drusen.

The evolution of angioid streaks is highly variable. The most frequent complication is the appearance of subretinal neovascular membranes (SRNVM) (72–86%), with poor visual prognosis if left untreated.\(^1\) Treatment includes a number of therapeutic possibilities.\(^1\)\(^-\)\(^3\) A case of choroidal neovascularization associated to angioid streaks is presented, treated with intravitreal bevacizumab in a patient with pseudoxanthoma elasticum.

Clinic case

Female, 28, examined in our service due to loss of vision and metamorphopsia in left eye (LE) with 2 days evolution. Relevant history included repeated renal colics and normal pregnancy and labor.

The initial exploration produced best corrected visual acuity (BCVA) of 0.5 in right eye (RE) and 0.16 in LE. Anterior pole biomicroscopy and intraocular pressure were normal. In both eyes, ocular fundus biomicroscopy revealed papillary drusen, angioid streaks and orange skin pigment alterations temporal to the macula. The RE macula exhibited subretinal fibrous scar and LE macula subfoveal grayish lesion with intraretinal hemorrhage (Fig. 1).

Optic coherence tomography (OCT) of both maculae was taken, which evidenced the presence of untreatable subretinal hyper-reflective lesions in RE and subfoveal hyper-reflective lesion with intraretinal fluid in LE (Fig. 2).

RE fluorescein angiography (FA) did not show contrasted diffusion at the macular level, which confirmed lesion inactivity. LE FE showed hyper-fluorescent area with subfoveal late times leak (Fig. 3), compatible with active SRNVM9.

LE was treated with intravitreal injection of bevacizumab (Avastin\(^®\) (1.25 mg/0.05 mL). After 3 injections, administered at 4-week intervals, LE BCVA improved to 0.9 while metamorphopsia diminished. Perilesional hemorrhage disappeared from the ocular fundus (Fig. 4) and OCT showed intraretinal fluid reabsorption (Fig. 5).

Systemic examination revealed hyperpigmented lesions and folds on the patient neck. Suspecting pseudoxanthoma elasticum, skin biopsy was requested from the Dermatology Service, which confirmed the diagnostic. Genetic study revealed the existence of homozygote mutation in gene ABCC6, which is responsible for the disease.

The ophthalmological examination of the father, siblings and daughter of the patient did not produce significant findings.

Discussion

Pseudoxanthoma elasticum is a hereditary disease characterized by fragmentation and calcification of elastic skin fibers and the middle tunic of arteries. Its inheritance is autosomic recessive even though there are some cases of pseudo-dominance. The disease is caused by mutations in gene ABCC6 (16p13.1), which encodes membrane-transporting proteins. The defect expresses in liver and kidneys, suggesting that the transport dysfunction could produce the accumulation of an unknown substance in blood and cause dystrophic changes in elastic fibers.

Clinically, it can involve various organs (cardiovascular, urological, gastrointestinal systems, among others). At the
Dermatological level, small yellowish papules appear with redundant lax skin in neck and armpits, among other areas.

At the ocular level, angioid streaks are observed in 85% of patients, which enhances the importance of diagnosing a systemic disease starting from an ocular disorder as in this case, with the ensuing repercussions for patients and family (genetic counseling).

Patients with angioid streaks are generally asymptomatic unless the lesions extend to the foveola or develop subfoveal SRNVM.\(^1\)

The literature reports that angioid streaks associated to pseudoxanthoma elasticum exhibit higher probabilities of developing macular choroidal neovascularization, in contrast with those associated to sickle cell anemia.\(^1\)

In the majority of cases, including the one discussed herein, FA confirms the presence of choroidal neovascularization. Indocyanine green angiography is used when it is difficult to determine the edges of the neovascular membrane in FA.\(^1\)

Historically, a range of treatments has been applied for angioid streak neovascularization. Initially, neovascularization was treated with argon laser photocoagulation\(^1\) but this was discarded due to higher relapse percentages. Photodynamic therapy (PTD) is controversial. Even so, Prabhu et al.\(^2\) reported positive treatment of SRNVM with low fluency PTD and intravitreal ranibizumab.

At present, intravitreal antiangiogenics are becoming the therapeutic option of choice.\(^3\)–5 Schiano Lomoriello et al.\(^3\) described effective treatment with intravitreal bevacizumab for choroidal neovascularization in angioid streaks and its deactivation in the long-term. More recently, Lee et al.\(^4\) described a case treated with 6 intravitreal bevacizumab injections with improvement of visual acuity. In the case reported here, the patient exhibited quick vision improvement after treating SRNVM with only 3 intravitreal bevacizumab injections. In addition, visual acuity has remained stable after 11 months follow-up. This result is in contrast with the evolution of the natural history of the disease as observed in the other eye which was not given treatment.

For the above reasons, intravitreal antiangiogenics should be considered as an efficient therapeutic option for choroidal neovascularization associated to angioid streaks.

Conflict of interest

The authors declare no conflict of interest.

References

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