CASE REPORT

Intravitreal ranibizumab for bilateral choroidal neovascularisation in a patient with angioid streaks

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SUMMARY

Angioid streaks are described as irregular lines deep into the retina, configured in a radiating fashion which results from breaks in Bruch's membrane. Optic nerve head drusen are acellular, globular deposits located within the optic nerve head. Angioid streaks and optic nerve head drusen may coexist in patients with pseudoxanthoma elasticum. Both disorders may cause choroidal neovascularisation. In this case report we aimed to present a 48-year-old man with pseudoxanthoma elasticum. The patient had bilateral choroidal neovascularisation secondary to angioid streaks and was treated with intravitreal ranibizumab injections. Visual acuity was increased and maintained at the same level during the follow-up time. There was no complication related to the injection.

BACKGROUND

Choroidal neovascularisation (CNV) caused by angioid streaks (AS) is a rare cause of visual acuity loss. Recent reports showed that CNV caused by AS can be successfully treated with intravitreal anti-vascular endothelial growth factor (anti-VEGF) injections.

CASE PRESENTATION

A 48-year-old man was referred to us with a symptom of decreased visual acuity in both eyes since a week. The best-corrected visual acuity (BCVA) in the right eye (RE) was 1.7 logMAR with ability in the left eye (LE) to count fingers from 1 m. Ocular history was unremarkable. The patient had systemic hypertension which was under control medically and also pseudoxanthoma elasticum (PXE). Anterior segment examination and

intraocular pressure measurements were normal in both eyes. Dilated fundus examination revealed elevated optic discs with blurred margins which was compatible with optic nerve head drusen (ONHD) and narrow, irregular lines deep into the retina which were compatible with AS in both eyes (figure 1A, B). In addition, an elevated yellowish subretinal lesion which was compatible with CNV was observed between optic disc and fovea in both eyes (figure 1A, B). Ultrasound imaging demonstrated bright reflex at the optic disc area corresponding to ONHD in both eyes (figure 2A, B).

A cross-sectional SD-OCT scan of the macula showed CNV with high reflectivity and subretinal fluid in the RE (figure 3A). There were multiple intraretinal cysts (figure 3A). SD-OCT scans of the LE showed subretinal fluid (figure 3A).

The patient signed a consent form that explained the potential risks and benefits of intravitreal injections. Each 0.5 mg/0.05 mL intravitreal ranibizumab injection was then administered under local anaesthesia. After the first injection, the patient was called for monthly visits and treated on an as-needed treatment regimen. Additional treatment was administered when the lesion showed subretinal–intraretinal fluid on OCT, leakage on fundus fluorescein angiogram, intraretinal–subretinal haemorrhage in the examination, or when a decrease in visual acuity was detected. After a follow-up period of 11 months, five injections were administered for the RE and two injections for the LE.

OUTCOME AND FOLLOW-UP

After the first injections, BCVA increased to 0.4 logMAR in the RE and to 1.0 logMAR in the LE. SD-OCT scans showed no subretinal fluid at the same

Figure 1 Fundus images of the right and left eye (A and B). Arrows indicate angioid streaks and stars indicate yellow elevation of the choroidal neovascularisation.

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Figure 2 Ultrasound images of the right and left eye (A and B). Ultrasound scans showing focal hyper-reflectivity at the head of the optic nerve that indicates optic nerve head drusen.

Figure 3 SD-OCT images of the right and left eye before the treatment (A and B). SD-OCT scan of the right eye demonstrates a hyper-reflective subfoveal choroidal neovascular membrane with subretinal fluid and intraretinal cysts before treatment (A). SD-OCT scan of the left eye demonstrate subretinal fluid (B).

location at 11 months follow-up (figure 4A, B). No complication related to the injection was observed.

DISCUSSION

AS were first reported by Doyme.¹ Owing to the clinical appearance, the term ‘angioid streaks’ was used.² However, the histopathological findings revealed that the underlying abnormality was in Bruch’s membrane rather than a vascular structure. Lesion described as irregular lines radiating from the optic nerve head to the peripheral retina and result from rupture or dehiscence of Bruch’s membrane between the retinal pigment epithelium and the choroid.³

AS may accompany ONHD in patients with PXE.⁴ Munteanu and Chercotă⁴ reported the association between ONHD and AS in the context of PXE was 8.5%. The values are significantly higher compared with those from the normal population.⁴

Another study showed that 86.2% of patients with AS had PXE and 21.6% of those patients also had ONHD.⁵

In AS cases, the visual acuity remains unchanged unless a break, a CNV or a hemorrhage affects the macula. In the literature, it has been shown that AS may cause a subfoveal CNV.⁶ There is no proven effective treatment for breaks yet; however, laser photocoagulation and photodynamic therapy has been used as a treatment for CNV.^{7, 8} The visual outcomes of these treatments were generally poor.^{7, 8} With the introduction of anti-VEGF therapy in the recent years, the prognosis of CNV-associated retinal diseases has changed dramatically. In our case, CNV was successfully treated by multiple ranibizumab injections. We decided the number of injections according to clinical and imaging findings. The patient suffered from a greater and more serious CNV in his RE than his LE, therefore he required five injections for the RE and two for the LE.

Figure 4 SD-OCT images of the right and left eye at 11 months follow-up after treatment (A and B). SD-OCT scan of the right eye demonstrates the resolution of subretinal fluid and decreased intraretinal cyst volume (A). SD-OCT scan of the left eye demonstrates the resolution of the subretinal fluid (B).

Similarly Ozkaya *et al*⁹ used three consecutive bevacizumab injections to treat CNV caused by AS in two patients and they reported that a successful result was obtained and visual acuity improved from 20/40 to 20/25 in one patient and from 20/80 to 20/50 in the other patient. Zebardast and Adelman¹⁰ used 12 ranibizumab injections for a 51-year-old patient with PXE presenting with macular CNV secondary to AS and after 5 years follow-up they reported that once stabilisation of the CNV was achieved by initial injections, recurrent neovascular activity was minimal and visual acuity was preserved. Also similar successful results were reported by several authors.^{11 12}

Our result in this case indicates that an anti-VEGF treatment can be useful for the CNV caused by AS. The visual and anatomical success was achieved and preserved during 11 months of follow-up period. This report adds to the evidence base for the use of ranibizumab in a case of CNV caused by AS.

Learning points

- ▶ Angioid streaks and optic nerve head drusen may be seen in the same patient.
- ▶ Choroidal neovascularisation caused by angioid streaks may be treated by intravitreal ranibizumab injection.
- ▶ Number of injection may be decided according to clinical and imaging findings.

Competing interests None.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

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