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Treatment of a Pseudoxanthoma Elasticum Patient Initially Presented with Choroidal Neovascularisation Secondary to Angioid Streaks

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Abstract

Pseudoxanthoma elasticum (PXE) is a rare, systemic disease with typical ophthalmological findings including angioid streaks (AS) (represent breaks in the Bruch's membrane). A serious and common complication of AS is the development of choroidal neovascularization (CNV), which can result in significant and irreversible vision loss. The introduction of intravitreal agents that inhibit vascular endothelial growth factor (VEGF) has improved outcomes in patients with CNV. In this paper we aimed to present a 33-years-old woman with PXE. The patient had CNV in the left eyesecondary to AS and was treated with intravitreal anti-VEGF injections. The visual acuity of our patient remained stable during follow up period.

Keywords

Pseudoxanthoma elasticum, Angioid streaks, Ranibizumab, Bevacizumab intravitreal, Choroidal neovascular membrane

Introduction

Pseudoxanthoma elasticum (PXE) is a rare systemic cutaneous disorder that mainly affects the elastic fiber of the skin, retina, and cardiovascular system [1,2]. Characteristic ocular lesions include angioid streaks, peaud'orange, pattern dystrophy, comet-tail atrophic spots, and choroidal neovascularization (CNV) [1]. CNV is the most common cause of vision loss in 68%-86% of all cases [1,3]. The vascular endothelial growth factor (VEGF) blockage treatment is effective in CNV of the patients with PXE [4-7].

Herein we presented a PXE case with AS related CNV and her management with anti-VEGF injections.

Case

A 33-years-old woman presented with gradual visual loss and metamorphopsia for 2 months in her left eye. She had a history of intravitreal bevacizumab injection a month ago in a clinic with a diagnosis of idiopathic CNV. However her complaints did not disappear and she admitted to our clinic.

Her visual acuity was 20/20 OD and 12/20 OS.Ocular examination of the anterior segment was unremarkable. Fundus examination revealed bilateral AS. Moreover subretinal granular deposites, pigment epithelium changes in the macula were seen in the right eye, and CNV was observed in the left eye.

Fundus fluorescein angiography revealed the hyperfluorescence of the CNV in the left eye. Optical coherence tomography showed subretinal fluid and CNV in the left eye (Figure 1). Her systemic investigation revealed PXE with characteristics skin lesions, and mitral valve prolapse.

The patient signed a consent form that explained the potential risks and benefits of intravitreal injections. Intravitreal bevacizumab (1.25mg/0.05mL) injection was performed. The patient was asked to visit monthly and treated on pro re nata. The patient was treated with 3 more intravitreal bevacizumab injections during follow up period of 18 months, and the last injection was performed 2 months ago before last visit.

The final examination revealed a visual acuity 20/20 OD and 10/20 OS, and minimal granular deposits and no sign and findings of reactivation of CNV (Figure 2).

Discussion

Angioid streaks are characterized with irregular lines under the retina, configured in a radial fashion results from breaks in Bruch's membrane. AS are not specific for PXE and have also been encountered in Paget's disease, Ehlers–Danlos syndromes, Marfan's syndrome, or inherited haemoglobinopathies [2].

Choroidal neovascularization is a serious and common complication of AS [1,3]. Moreover, CNV due to AS in PXE are very hard to treat and carry a high risk of severe vision loss, especially in macular involvement [4,8]. The risk of developing CNV increases with age [9]. Our patient was 33 years old and she was diagnosed as idiopathic CNV in an external center. The diagnosis of PXE was given in our center based on the fundus findings (AS and CNV), typical skin lesions, and the presence of mitral valve prolapse. Although CNV was bilaterally seen in 80% of the PXE patients, our patient had unilateral membrane [10].

Anti-VEGF agents are one of the treatment options of CNV which carries minimal risk of damage to adjacent ocular tissue [10]. Favorable results have been reported with bevacizumab and ranibizumab [5,11,12]. Whereas PXE patients with CNV need relatively more intravitreal injections than the other CNVs. Our

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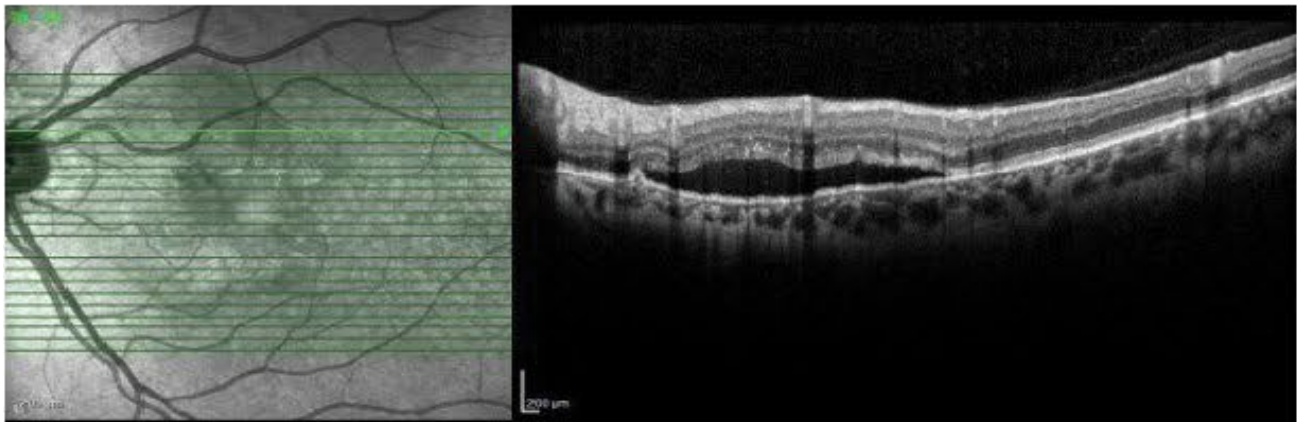

Figure 1: Optical coherence tompgraphy section shows the subretinal fluid and choroidal neovascular membrane.

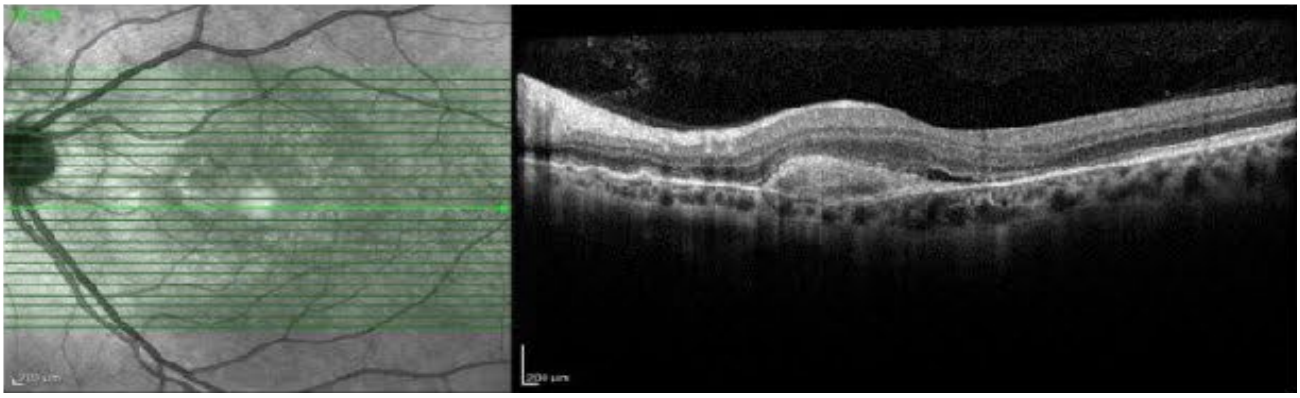

Figure 2: At 18th month the optical coherence tomography revealed the granular hyper reflective accumulation and the disappeared subretinal fluid.

patient received four intravitreal anti-VEGF injections during 18 months follow up.

Anti-VEGF therapy is the first treatment that can stabilize and even improve vision in AS associated CNV.In the management of CNV in PXE, anti-VEGF therapy should be started as soon as possible to achieve optimal results [5]. Some authors have reported stabilization or improvement of visual acuity in patients treated with intravitreal bevacizumab or ranibizumab for AS related CNV [5,10,11,13]. In this report, we also demonstrated that intravitreal anti-VEGF treatment is effective in our PXE patient with CNV. The visual acuity of our patient remained stable during follow up period.

It is of great importance to diagnosis PXE early in order toprevent morbidity and mortality from systemic complications. Although they are not frequent but PXE can be responsible for severe, life threatening complications such as myocardial infarction [14]. Therefore the management of the PXE patients requires a multidisciplinary approach of the dermatologist, ophthalmologist, and cardiologist [1].

Conclusion

In conclusion as in this case report, repeated intravitreal injections of anti-VEGFs have been shown to be effective in the treatment of CNVs, and stabilization of the visual acuity.

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