

Angioid streaks – clinical course and photodynamic therapy

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Abstract

Angioid streaks develop as a result of damage to the Bruch's membrane. They may occur as idiopathic angioid streaks or in association with pseudoxanthoma elasticum, Paget's disease and the haemoglobinopathies. Other pathologies of the fundus of eye may coexist together with angioid streaks, such as drusen of an optic disc and granular-like retina in posterior pole known as "peau d'orange". Subretinal neovascularization as well as atrophy in macula are also observed in the cases of long-lasting pathologies. The diagnosis is based mainly on clinical manifestations. Photodynamic therapy in treatment patients with secondary choroidal neovascularisation appeared to be effective.

INTRODUCTION

Angioid streaks are lesions in the elastin-filled tissue in the Bruch's membrane associated with atrophic degeneration of the overlying retinal pigmented epithelium. Increased vulnerability of the basal layer of the retinal pigmented epithelium is secondary to degeneration process and calcium deposition (Sato *et al.*, 2009).

Angioid streaks were first described in 1889 by Doyne. In 1892, Knapp was the first one who introduced the term "angioid streaks" as their appearance suggested a vascular origin.

Angioid streaks have a typical appearance as irregular lines which are broader than blood vessels. They radiate out in a cruciate pattern from the optic disc and are visible under the retinal vessels. The color of angioid streaks, which is usually red, brown or yellow depends on the background coloration of the fundus. Optic disc drusen are usually associated finding.

The most common retinal feature associated with angioid streaks is the presence of peau d'orange, which classically precedes the appearance of the streaks. It is described as relatively symmetrical, bitemporal drusen-like retinal pigment lodgements.

Other retinal change associated with angioid streaks include focal chorioretinal scarring – ("salmon patches") which typically appear in the peripheral area of the fundus. Choroidal hemorrhage may occur after even a minor trauma as a consequence of increased brittleness of the Bruch's membrane.

Choroidal neovascularization is a common complication which may lead to reduced visual acuity. It becomes symptomatic when the angioid streaks involve the foveola.

Angioid streaks may occur as an isolated phenomenon or in association with systemic disease,

such as pseudoxanthoma elasticum (Grondblad-Strindberg syndrome) (Buteică *et al.*, 2008; Finger *et al.*, 2009), Ehlers-Danlos syndrome, Paget disease or hemoglobinopathies which are found in approximately 50% of patients with angioid streaks.

The diagnosis of angioid streaks is usually made on the basis of fluorescein angiography (FA). Angioid streaks have a 'window defect' due to atrophy of the retinal pigment epithelium, which occurs over the streaks (Sayanagi *et al.*, 2009).

OCT (optical coherence tomography) (Charbel Issa *et al.*, 2009) and ultrasound examination may be useful diagnostic tools to diagnose angioid streaks in cases with other coexisting lesions.

CASE REPORT

A 39-year old female with no significant medical history had a routine eye examination which revealed the following:

Vod=1.0sc, Snod=0.5sc, VOs=1.0csc, Snos=0.5sc. In anterior parts of both eyes no abnormalities were detected. In both eyes optic disc drusen were found. Around each optic disc a dark grey ring of irregular angioid streaks with radiations to circumference of the fundus was found (Figures 1 and 2).

The visual field test was normal for each eye. Presence of drusen was confirmed by ultrasound examination (Figures 3 and 4) as well as in fluorescein

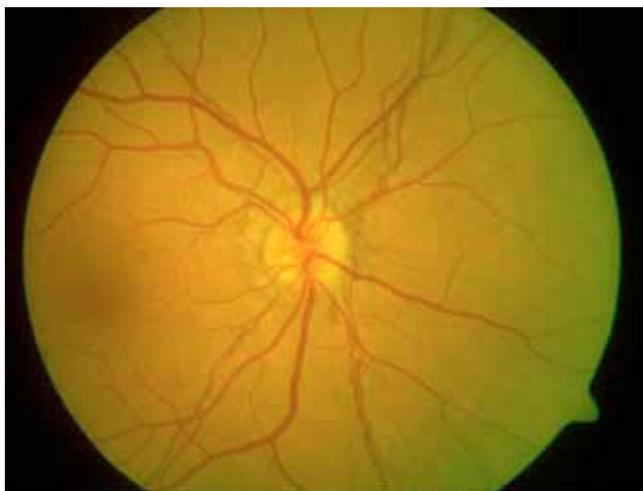


Fig. 1. Right eye: angioid streaks, optic nerve drusen.



Fig. 2. Left eye: angioid streaks, optic nerve drusen.

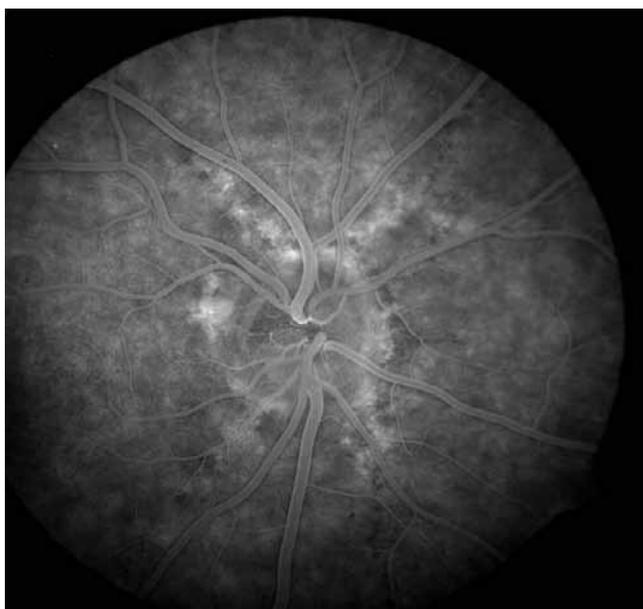


Fig. 3. Fluorescein angiogram of the right eye: drusen, angioid streaks

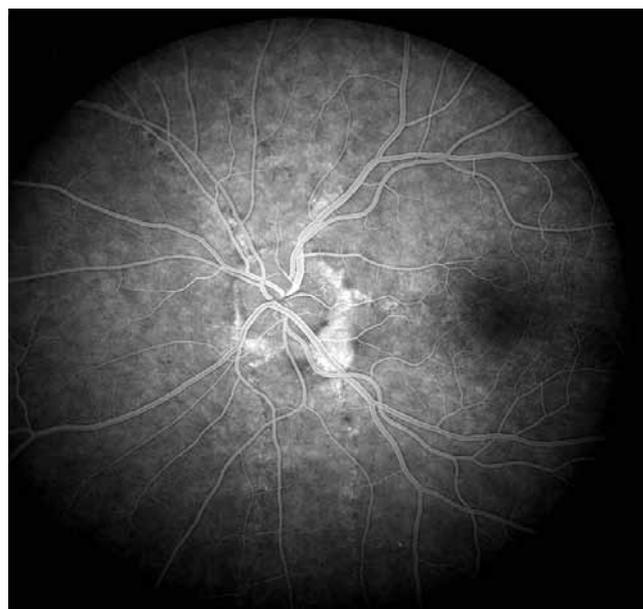


Fig. 4. Fluorescein angiogram of the left eye: drusen, angioid streaks

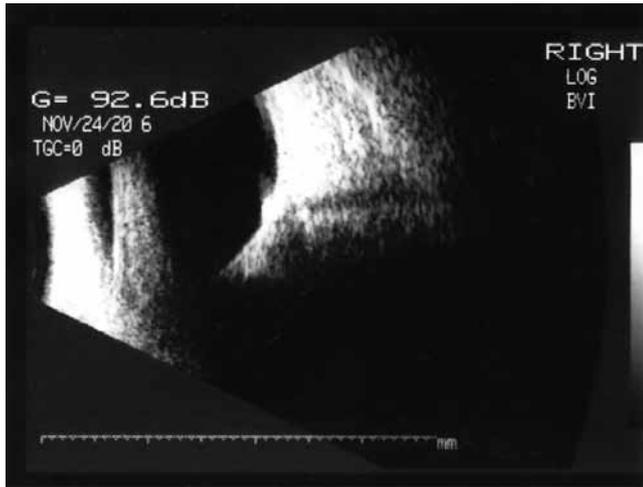


Fig. 5. Ultrasonogram of the right eye: optic disc drusen.



Fig. 7. Skin lesions.



Fig. 6. Ultrasonogram of the left eye: optic disc drusen.



Fig. 8. Skin lesions.

angiography (Figures 5 and 6). Skin lesions typical of pseudoxanthoma elasticum were found on general physical examination (Figures 7–9).

After approximately 1.5 year, the patient presented visual acuity reduction in the left eye ($V_{os}=0.4$). The secondary choroidal neovascularization (CNV) in the macula was confirmed by clinical examinations and imaging studies (FA, OCT) (Figure 10). No additional lesions were identified in the right eye.

The patient was treated by photodynamic therapy (PDT) in the left eye. After 3 months of treatment a scar was formed in the area of macular neovascularisation (Figure 11). Satisfactory visual acuity was sustained ($V=0.2$).

The patient was advised to have regular ophthalmological examinations.



Fig. 9. Skin lesions.

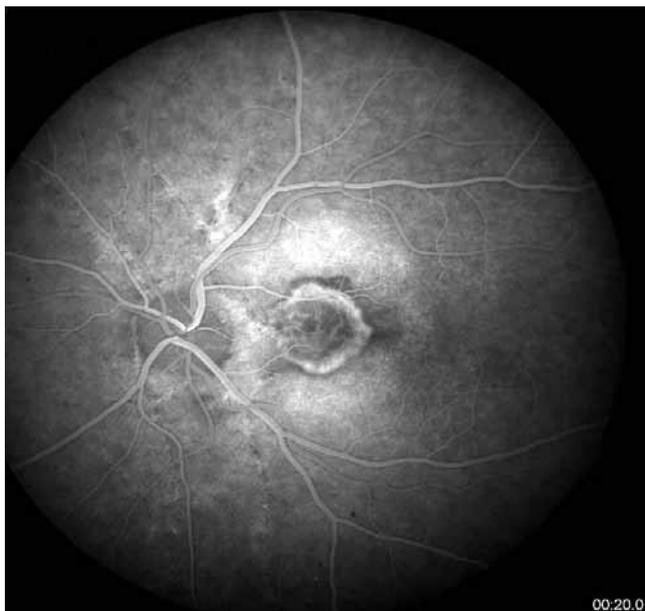


Fig. 10. Choroidal neovascularisation in macula.

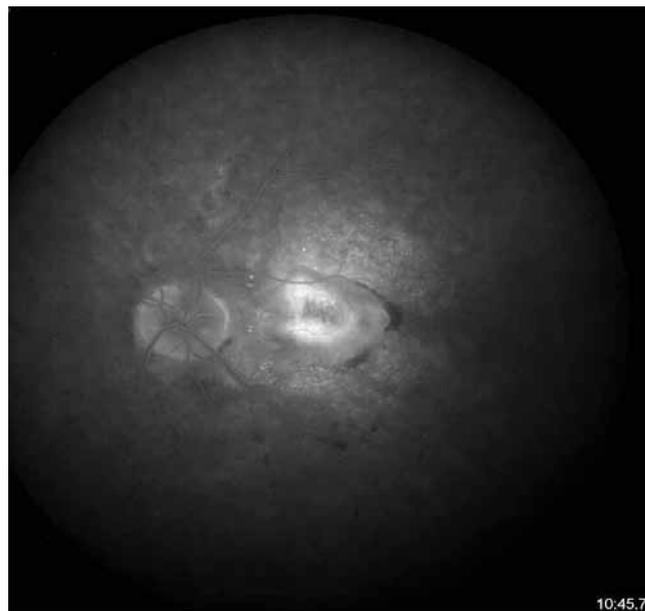


Fig. 11. Left eye: scar in macula.

DISCUSSION

Angioid streaks are considered a rare eye disorder with diversified clinical picture. They are usually an accidental finding in patients who remain asymptomatic.

There is no specific treatment for asymptomatic angioid streaks, but it is crucial to warn the patients that even minor trauma may lead to retinal hemorrhage.

The secondary choroidal neovascularisation which may result in reduced visual acuity is the most serious consequence of angioid streaks.

The most common treatment for choroidal neovascularisation has been laser photocoagulation of the retina. However, it can be applied only for the treatment of extra- and perifoveal membranes. New treatment methods are currently being evaluated. PDT has been assessed in clinical trials and approved for age-related macular degeneration (AMD) treatment (Soucek and Cihelkova, 2002). The PDT treatment, however cannot restore sight that has already been damaged by macular degeneration.

Photodynamic therapy with verteporfin appears to be highly selective in destroying neovascularisation associated with angioid streaks, which was proven in the described case report (Figurska, 2007; Lee *et al.*, 2007). Treatment with anti-VEGF drugs is now applied in CNV treatment (Liu *et al.*, 2009; Neri *et al.*, 2009; Schiano *et al.*, 2009; Wiegand *et al.*, 2009).

Clinical effectiveness and low risk of adverse effects seem to make this method useful in patients with angioid streaks.

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