Amalric sign: An augur of ophthalmic artery occlusion

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A 75-year-old man presented to us with sudden onset of profound vision loss in his right eye and was identified as suffering from an ophthalmic artery occlusion. Apart from the retinal whitening and box-carring of the retinal arteries, there were characteristic triangular patches of retinal whitening in the midperipheral temporal fundus indicating a previous lateral posterior choroidal artery occlusion. The patient was a chronic smoker and had dyslipidemia. The carotid Doppler study showed complete occlusion of the internal carotid artery. The presence of these triangular patches of retinal whitening or amalric sign can therefore herald a more proximal vessel occlusion. Hence such patients require evaluation on an emergency basis. The characteristic features of the patches on fluorescein angiography and indocyanine green angiography are discussed here.

Key words: Amalric sign, artery occlusion, dyslipidemia, triangular patch

Amalric first described the occurrence of triangular patches of retinal whitening in the fundus of patients with posterior choroidal artery occlusion.[1] These patches were reproduced in experimental animals by Hayreh and Baines by cauterizing the posterior ciliary arteries.[2] Several case reports have reported the occurrence of these patches in various scenarios, including cocaine inhalation[3] and polyarteritis nodosa-associated vasculitis.[4] Our current discussion is about a patient who presented with complete ophthalmic artery occlusion but had these patches of retinal whitening in the midperiphery which could possibly indicate a previous posterior ciliary artery occlusion.

Case Report

A 75-year-old man presented to us with sudden onset of diminution of vision in his right eye since 5 days. He had transient episodes of obscuration of vision in the same eye a few weeks back. He also complained of claudication pain in his lower limbs a month earlier for which he was started on treatment with antiplatelet and hypolipidemic drugs. The patient provided a history of smoking for the last 40 years. He was not a known diabetic or hypertensive.

The best corrected visual acuity was no light perception in the right eye and 20/80 in the left eye. The right eye showed afferent pupillary defect, fine keratic precipitates over the endothelium, anterior chamber flare 1+, cells 1+ and significant cataractous changes. The left eye showed a briskly reacting pupil and early cataractous changes. Intraocular pressure was 28 mmHg in the right eye and 8 mmHg in the left. Extraocular movements of both eyes were normal.

Fundus of the right eye showed box-carring of the retinal arteries, dilated retinal veins with sluggish circulation, whitening of the retina in the posterior pole but no cherry red spot, triangular white patches in the midperipheral fundus in the superior, inferior, and temporal retina and numerous cotton wool spots in the posterior pole [Figs. 1-3]. The left eye fundus was normal except for a few hard drusen.

Fluorescein angiography showed a gross delay in the arm-retina transit time in the right eye. The cilioretinal artery fluoresced by 57 s, followed by a minimal filling of the proximal arterial trunk, but the major part of

Figure 1: Fundus picture of the right eye showing box-carring of the retinal arteries, banking of the veins, and retinal whitening of the posterior pole with few cotton wool spots

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the retina remained nonperfused [Fig. 4]. The triangular patches seem to fluoresce from approximately 1 min and showed leakage in the late frames [Fig. 5]. More of these patches were picked up on fluorescein angiography than on indirect ophthalmoscopy. The left eye showed a normal fluorescence picture.

Indocyanine green angiography showed delayed filling of the large choroidal vessels, but the striking features of the triangular patches seen on fluorescein angiography was absent [Fig. 6]. The optical coherence tomography showed hyperreflectivity of the inner retinal layers with a hyporeflective band in the outer retina correlating with the opaque ischemic retina [Fig. 7].

The total cholesterol level was 380 mg/dl (ref range: 130–250 mg/dl) and the serum triglyceride level was 350 mg/dl (ref: up to 150 mg/dl). Acute phase reactants, blood sugars, and ECG were normal. The color Doppler study of both carotid and vertebral arteries showed diffuse irregular
circumferential plaques in the right carotid bulb causing 50%–60% stenosis with complete thrombotic occlusion of the entire internal carotid artery. The echocardiography showed normal valves and chambers. The arterial Doppler of both lower limbs showed occlusion of bilateral external iliac artery, high-grade stenosis of the right common femoral artery and monophasic flow and low velocity in nearly all the major arteries including popliteal, anterior tibial, and posterior tibial.

**Discussion**

This case scenario presents some unique features of embolic phenomenon in the ophthalmic arterial tree. The presence of cotton wool spots and the triangular patches in the fundus indicate embolic occlusion of the end arteries in the retinal and choroidal circulations. This might have preceded the complete ophthalmic artery occlusion with which the patient presented to us. The distribution of the patches indicates a possible occlusion of the short distal branches of the lateral posterior choroidal arteries.

Hayreh and Baines produced similar lesions in rhesus monkeys by cauterizing the posterior ciliary arteries. Hayreh describes them as elongated or wedge-shaped lesions of various shapes and sizes but they often take the shape of triangular sector-shaped lesions located in the peripheral fundus with bases toward the equator and apices toward the posterior pole.[5]

In a case report by Sribhargava *et al.*, similar patches were found in the temporal fundus of a chronic smoker.[6] Reddy *et al.*[3] reported similar triangular lesions in a patient following cocaine inhalation, but the lesions show initial hypofluorescence and late hyperfluorescence on angiography. The near-complete absence of perfusion in the adjacent regions may not have highlighted the early hypofluorescence of these patches in our case.

The presence of extensive thrombosis in the arterial tree of the lower limb and the carotids with an abnormal lipid profile in a smoker indicates a highly morbid condition. The presence of cotton wool spots in the retina and the presence of amalric sign in the midperipheral fundus are telltale signs of previous embolic events in the retinal and choroidal vascular tree. In hindsight, the presence of these patches in a patient should alert the clinician to image the carotids to possibly prevent a catastrophic event such as an ophthalmic artery occlusion.

The triangles of amalric were better evaluated on fluorescein angiography in this patient. These patches showed early hyperfluorescence due to transmission defects in the damaged RPE, and the late frames show considerable leak from the necrotic tissue which traps the slowly perfusing fluorescein. The indocyanine green angiography was not very useful in this case as the molecules, being more intravascular, did not highlight the patches in either the early or the late frames.

This is a singular case of ophthalmic artery occlusion in a chronic smoker with triangular sign of amalric in the peripheral fundus indicating a previous embolic occlusion of the lateral posterior choroidal arteries. Thereby, the presence of amalric sign can herald the development of a larger proximal vessel occlusion and should be evaluated on an emergency basis.

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**Conflicts of interest**

There are no conflicts of interest.

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