



Case report

Amalric triangular sign in a case of central retinal artery occlusion combined with posterior ciliary artery occlusion – Case report

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ABSTRACT

Purpose: Amalric triangular sign is a rare phenomenon indicating choroidal ischemia. In this study, we reported a typical Amalric triangular sign in a case of central retinal artery occlusion (CRAO) combined with posterior ciliary artery (PCA) occlusion.

Observations: A 49-year-old women developed sudden visual loss in her left eye for one day. Ocular examination revealed rubeosis iridis, macular retinal edema followed by multiple whitish triangular patches in the peripheral four days later. Fluorescein angiography (FAG) revealed delayed choroidal filling time, delayed arteriovenous transit time, choroidal non-perfusion areas and triangular lesions of hyperfluorescent corresponding to the hypopigmented patches on the fundus. Carotid Doppler and magnetic resonance angiography (MRA) then disclosed 90% stenosis of left internal carotid artery (ICA), causing ischemia of the central retinal artery and posterior ciliary artery.

Conclusions and importance: The Amalric triangular sign indicates the occlusion on the main trunk of PCA. The sign might combine with CRAO or branch retinal artery occlusion (BRAO) as presented in our case and therefore is a strong indication of possible systemic vascular risk.

1. Introduction

Amalric triangular sign was first described by Pierre Amalric¹ as an unusual triangular pigmentary disturbance in the fundus due to choroidal ischemia. Combined central retinal artery occlusion (CRAO) and posterior ciliary artery occlusion is an uncommon vascular disease causing the loss of blood supply to the inner retina and choroid that results in severe vision loss. In this study, we report a typical Amalric triangular sign presentation in a case of central retinal artery occlusion combined with posterior ciliary artery occlusion.

1.1. Case report

A 49-year-old diabetic woman presented to our outpatient department with sudden onset of blurred vision in her left eye for one day. Ocular examination showed visual acuity was only counting finger (CF) in the left eye and intra-ocular pressure (IOP) was 25.0 mmHg with rubeosis iridis. Ophthalmoscopy discovered that the disc was pink, and there was retina edema over the posterior pole with an indistinct cherry-red spot. Increased reflectivity and thickness in inner retina were

seen on Optical coherence tomography (OCT; Stratus, Zeiss, Model 3000, USA) [Fig. 1]. Four days later, multiple triangular hypopigmented subretinal lesions were detected with triangle vertices toward the posterior pole. Fluorescein angiography (FAG) showed delayed choroidal filling time, delayed arteriovenous transit time, choroidal non-perfusion areas and triangular lesions of hyperfluorescent with the apex pointing toward the macula and the base facing the periphery in the late phase corresponding to the hypopigmented patches on the fundus [Fig. 2]. Systemic investigations revealed an elevated HbA1c 9.9%, increased triglyceride 239 mg/dL, and cholesterol 202 mg/dL. Carotid Doppler and magnetic resonance angiography (MRA) disclosed 90% stenosis of the left internal carotid artery (ICA) [Fig. 3]. Pan retinal photocoagulation therapy (PRP) and anti-vascular endothelial growth factors (anti-VEGF) injection were performed, resulting in a complete regression of iris neovascularization and a decrease of IOP to 19 mmHg. Additionally, the patient was referred to a cardiologist for the left carotid artery stenting. Two months later, the whitish triangular lesions on the fundus evolved to mottled atrophic patches with interweaving hypo and hyper-pigmentation [Fig. 4].

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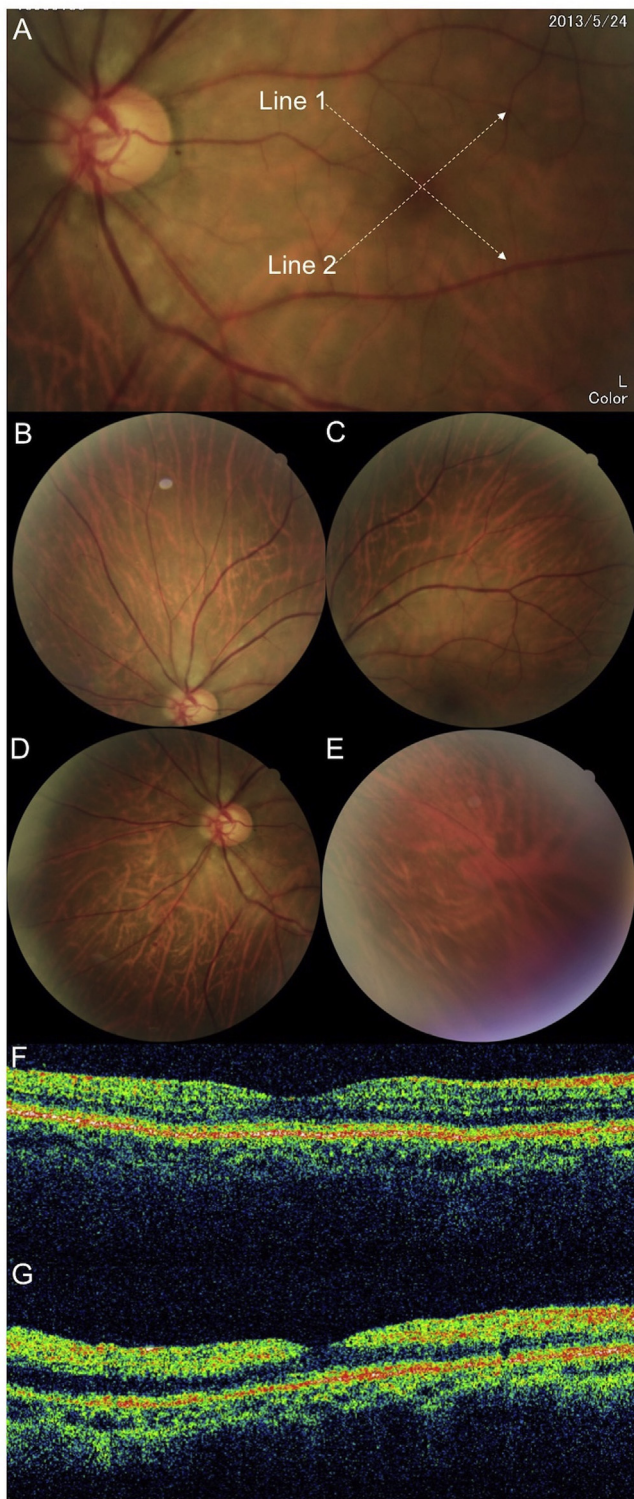


Fig. 1. Left color fundus photographs and optical coherence tomography scanning of the patient at the initial visit.

(A) Obvious retinal edema with an indistinct cherry-red spot in the posterior pole. (B) (C) (D) (E) showed the peripheral retina; (F) (G) Optical coherence tomography from line 1 and line 2 showed increased reflectivity and thickness of the inner retina. (For interpretation of the references to color in this figure legend, the reader is referred to the Web version of this article.)

1.1.1. Discussion

Central retinal artery occlusion (CRAO) is an ophthalmic emergency equivalent to an acute stroke of the eye causing severe vision

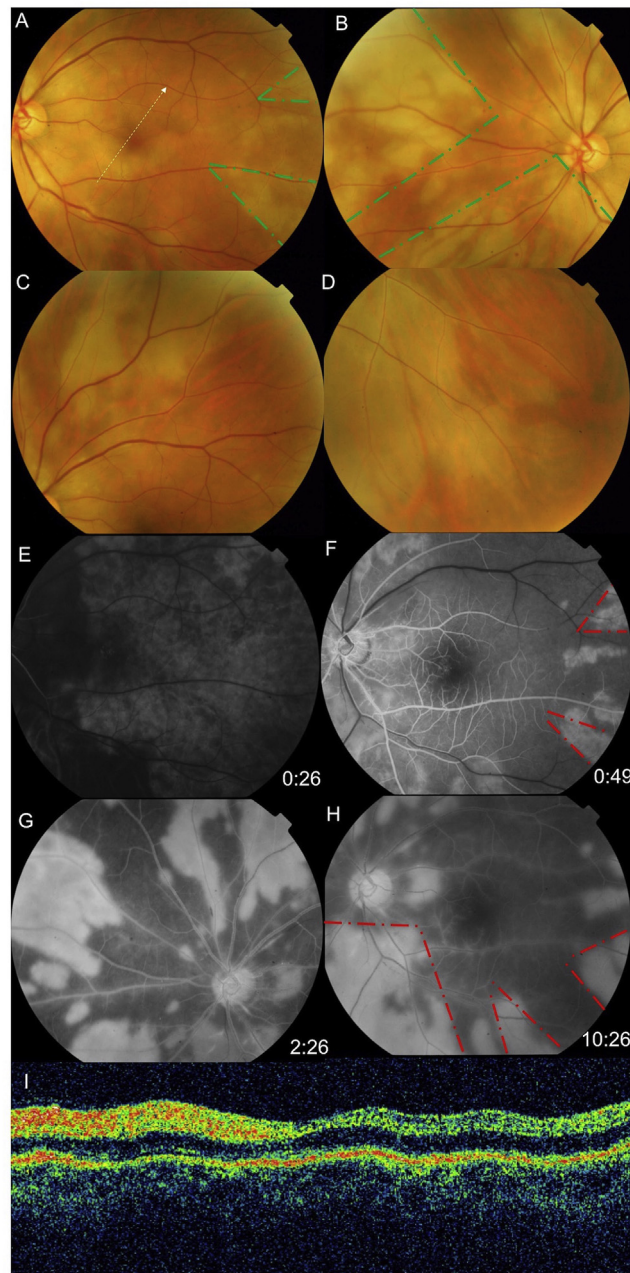


Fig. 2. Left color fundus photograph, fluorescein angiography and optical coherence tomography scanning four days after the initial visit.

(A)~(D) Color fundus photograph revealed the triangular hypopigmented subretinal lesions (the green lines highlight the contour). Fluorescein angiography revealed (E) choroidal nonperfusion area in the early phase and (F) (G) (H) triangular hyperfluorescent lesions in the late phase (the red dashed lines highlight the contour). (I) Optical coherence tomography corresponding to the area from white dash line in (A) still showed inner retinal edema. (For interpretation of the references to color in this figure legend, the reader is referred to the Web version of this article.)

impairment. Most cases of CRAO have underlying atherosclerotic risk factors, which also are the risk factors of cerebral vascular complications and ischemic heart disease in the future.²

The posterior ciliary artery (PCA), one of the branches of ophthalmic artery, is the key source of blood supply to the optic nerve head (ONH), the choroid to the equator, the retinal pigment epithelium (RPE), the outer 130 μm of retina, and the medial and lateral segments of the ciliary body and iris.³ Any ischemic disturbance in the PCA circulation can result in a variety of ocular and ONH ischemic lesions,



Fig. 3. Carotid magnetic resonance angiography.

(A)The carotid magnetic resonance angiography showed severe stenosis of left internal carotid artery (B)The red box: amplification of stenosis area. (For interpretation of the references to color in this figure legend, the reader is referred to the Web version of this article.)

causing varying degrees of visual loss.

Pierre Amalric was the first to describe an unusual triangular pigmented disturbance in the fundus of patients with ischemic ocular disease.^{1,4} He postulated that this disturbance was caused by choroidal ischemia. According to Hayreh's animal model, cutting the medial or lateral posterior ciliary arteries in rhesus monkeys produced similar fundus lesions in a typical time course.⁵ In the initial stage (within 1 h after PCA occlusion), no detectable fundus change could be noted. After 18–24 hours, sectoral-shaped or triangular whitish patches of varying size became apparent in the fundus. These triangular patches mostly pointed toward the posterior pole or disc and separated by narrow strips of normal color, compatible with the distribution of choroidal branches of short posterior arteries. In the early phase of fluorescein angiography (FAG), delayed ciliary-retinal artery filling and absence of background choroidal fluorescence could be noted, while in the mid to late phases intense hyperfluorescence in the regions corresponding to the triangular patches developed, which may represent a transmission defect from RPE necrosis. After two to three weeks, these patches developed as granular, grayish depigmented scars.⁶ Eventually, the lesions were discovered to be triangular, depigmented scars as revealed in our patient's fundus [Fig. 4].

Histopathological examination revealed that these patches were caused by infarction lying deep within the retina.⁶ Outer nuclear layer and RPE necrosis develop first, followed by degeneration of outer nuclear layer and RPE within three weeks. RPE proliferation with irregularly scattered pigment granules in the necrotic area could also be found at around day 21. Approximately two months later, these areas evolved to become several atrophic patches with some pigment clump accumulation in the fundus as presented in our case [Fig. 4 (I)]. More destructive RPE degeneration developed at day 100. The choroid usually had no demonstrable change during the whole course.

There are several reports of retinal artery occlusion and choroidal vessel occlusion after subcutaneous or nasal mucosal injection of particles including corticosteroid powder or cosmetic facial filler, such as hyaluronic acid or autologous fat.^{7–14} Most cases had typical presentations of sudden-onset, painless visual loss or visual field defects after injection. The cases possibly accompanied ptosis, ophthalmoplegia or positive relative afferent pupillary defects. In some cases, initial fundus examination found branch or central retinal artery

occlusion with single or multiple visible emboli. Embolic shadow could also be found in the choroidal vasculature on ophthalmoscopic examination. The embolic shadow was caused by retrograde blood flow to the ophthalmic artery and then to the retinal artery after accidental intra-arterial injection of particles into the collateral connections of ophthalmic arteries. While these small and multiple embolic occlusions by small particles could cause severe visual impairment, the occlusions would usually not induce the typical Amalric triangular sign, since this sign indicates infarction of a large area of choroidal tissue supplied by large choroidal vessels, such as the main trunks of short PCA.

In 2009, Sribhargava reported a case of left CRAO and left PCA accompanied with left distal ICA total occlusion, which also presented Amalric sign in FAG.¹⁵ As in our case, the 49-year-old female with left eye CRAO combined with PCA occlusion resultant from 90% occlusion of left ICA, also presented typical Amalric triangular sign. Amalric triangular sign indicates the occlusion of the main trunk of the PCA and might combine with a CRAO or a BRAO. Therefore, Amalric triangular sign is a strong indication of possible systemic vascular risk.

Patient consent

The patient consent had not been obtained since the patient had been lost follow up. This article do not contain any personal information of the patient.

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Authorship

All authors attest that they meet the current ICMJE criteria for authorship.

Conflicts of interest

None.

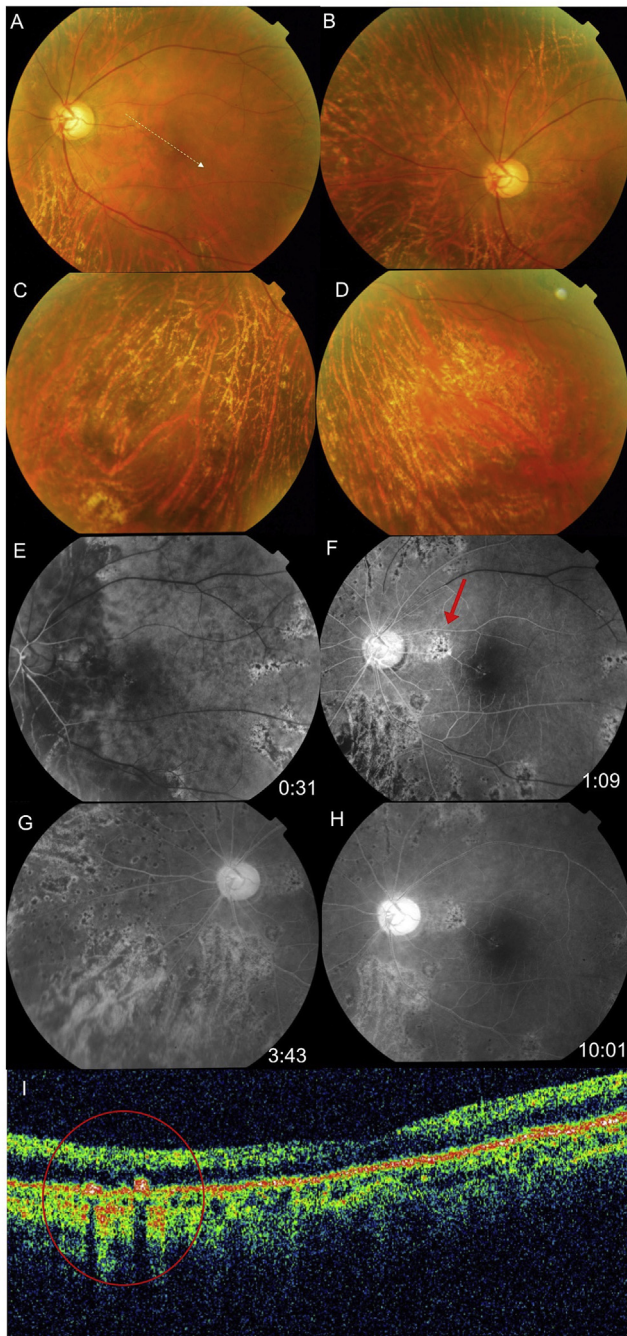


Fig. 4. Left color fundus photograph, fluorescein angiography and optical coherence tomography scanning two months after the initial visit.

(A)–(D) The whitening triangular lesions on the fundus were evolved to as atrophic patches. Fluorescein angiography revealed (E) delayed choroidal perfusion in early phase and (F) (G) (H) late staining. (I) Optical coherence tomography revealed retinal pigment epithelium proliferation (red circle) in the area of atrophic patches corresponding to the area of hypofluorescence in fluorescein angiography (Figure F, red arrow). (The multiple small round pigmented spots in mid-peripheral and peripheral retina with late staining in fluorescein angiography were panretinal photocoagulation scars). (For interpretation of the references to color in this figure legend, the reader is referred to the Web version of this article.)

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