

A case of central retinal artery occlusion due to atrial myxoma with excellent visual recovery

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Key words: Central retinal artery occlusion, emergency management, left atrial myxoma, leading fluorescein sign, systemic thromboembolism, visual recovery

Central retinal artery occlusion (CRAO) is an ocular emergency characterized by acute vision loss in one eye, with an incidence of approximately 1 to 10 in 100,000 people.^[1] The source of an embolus in elderly individuals is carotid artery atherosclerosis whereas it is a cardiac disease in young individuals.^[1] CRAO has been rarely caused by cardiac myxoma.^[1-8] In a case of CRAO caused by myxoma, the imminent management involves that of CRAO and definitive management is the resection of myxoma.^[1-10] Here, we report a case of CRAO secondary to left atrial myxoma managed by surgical resection with excellent visual recovery. A high index of suspicion is needed to diagnose a left atrial myxoma after CRAO and the myxoma should be resected surgically to prevent further embolic events.^[9,10]

A 45-year-old hypertensive and hypothyroid male patient who had a recent (6 months) history of left-sided hemiparesis, presented with sudden painless loss of vision to counting fingers in his only seeing left eye of 6 h duration. The other eye (right eye) had no light perception due to traumatic optic neuropathy following a road traffic accident that happened 14 years back. In the left eye, the anterior segment was unremarkable except for relative pupillary afferent defect (RAPD) and fundus examination showed severe arteriolar attenuation, retinal opacification with cherry-red spot, and cattle trucking of vessels consistent with the diagnosis of CRAO [Fig. 1a and b]. Immediate management involved globe massage with a 4-mirror gonioscope (Volk, USA) under slit-lamp guidance, with enough pressure to cause optic disc blanching and release of pressure in a ratio of 15:5 s to dislodge the causative embolus and restore retinal

circulation. Paracentesis was done with aseptic precautions and oral acetazolamide 500 mg stat was given. After the emergency management, findings were documented by fundus fluorescein angiography [Fig. 2a-d] and optical coherence tomography angiography [OCT] [Fig. 3a-c]. The patient was referred to a cardiologist to identify the source of the embolus, keeping in mind the two back-to-back thromboembolic events 6 months apart. He was found to have a large left atrial mass of mixed echogenicity measuring 35 × 32 mm on transthoracic echocardiogram [Fig. 4], consistent with the diagnosis of left atrial myxoma. To prevent further embolic events, surgical removal of the irregular gelatinous friable mass was done through a medial sternotomy approach. Histopathological diagnosis reiterated the diagnosis of myxoma. He was advised to continue blood thinners and advised periodic follow-up. Three weeks later, his unaided vision improved to 6/12 in the left eye. Fundus examination revealed a decrease in the retinal opacification (likely recovery of retinal circulation) with disc pallor and the repeat OCT macula showed macular atrophy [Fig. 5].

Discussion

CRAO is characterized by acute, painless loss of monocular vision, mostly seen in elderly patients with systemic co-morbidities.^[1] Although CRAO may be caused by cardiac diseases in the young, the incidence of CRAO due to atrial myxoma is rare.^[1]

Cardiac myxomas are the most frequent cardiac tumors of endocardial origin.^[9,10] Although histologically benign, myxomas may be lethal, due to impairment of cardiac dynamics and their thromboembolic potential.^[9] Myxomas occur more often in women between the third and sixth decades of life.^[9] They are usually solitary and originate in the left atrium (75%) or the atrial septum, close to the foramen ovale.^[1,9,10] When myxoma is located in the left atrium, the risk of embolism increases because of high blood flow dynamics.^[9] Cardiac myxoma can be a source of emboli to the retina, central nervous system, and elsewhere in the vascular system; however, it often affects middle cerebral arteries and supraclinoid internal carotid artery.^[1] In addition, individuals with myxoma may present with obstructive and constitutional symptoms.^[9,10]

The diagnostic method of choice is transthoracic echocardiography, which has a sensitivity of 95%.^[9] According to echocardiography appearance, myxomas can be classified as smooth or villous types.^[9] Smooth tumors are larger, occur with obstructive symptoms, and benefit from elective surgery, whereas villous myxomas are friable/gelatinous in texture

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Quick Response Code:	Website: www.ijjo.in
	DOI: 10.4103/ijjo.IJO_239_22

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Received: 01-Feb-2022

Revision: 21-Mar-2022

Accepted: 19-May-2022

Published: 30-Jun-2022

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Cite this article as: Balasubramaniam S, Sreenivasan J, Dhawan A, Agarwal A. A case of central retinal artery occlusion due to atrial myxoma with excellent visual recovery. Indian J Ophthalmol 2022;70:2698-700.

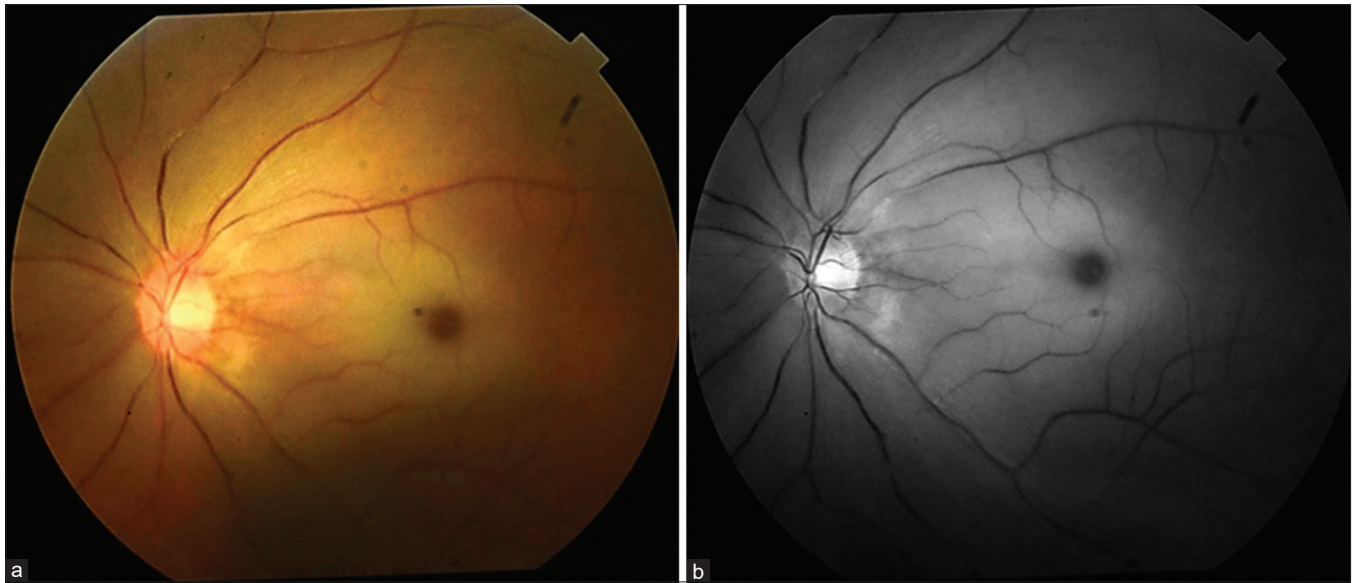


Figure 1: Fundus examination of the left eye (a - color fundus photo and b - red free photo) showed severe attenuation of all the arteries, retinal opacification with cherry-red spot, and cattle trucking of vessels

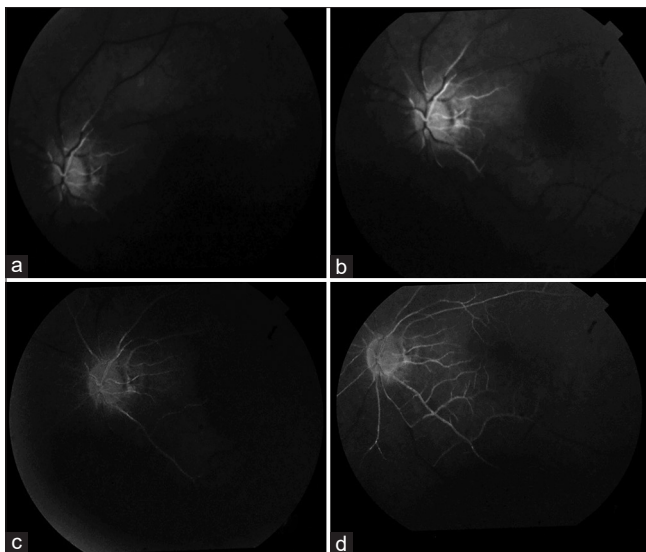


Figure 2: Fundus fluorescein angiography [a and b – early phases, c and d – late phases] of the left eye showed delayed arm-retina time with a delayed filling of arterioles and leading fluorescein sign

with high embolic risk and require urgent surgical treatment.^[9] Surgical resection of cardiac myxomas should be performed early due to the risk of developing systemic embolization.^[10] Surgery is safe and perioperative mortality is low with few recurrences.^[9]

In literature, there have been few reports of CRAO caused by myxomas.^[1-8] Most of the authors reported them at a relatively younger age with many patients having neurological symptoms along with CRAO.^[1,2] The patient reported here was relatively older and there was a history of cerebrovascular accident 6 months back. The occurrence of multiple embolic events should prompt suspicion of myxoma.

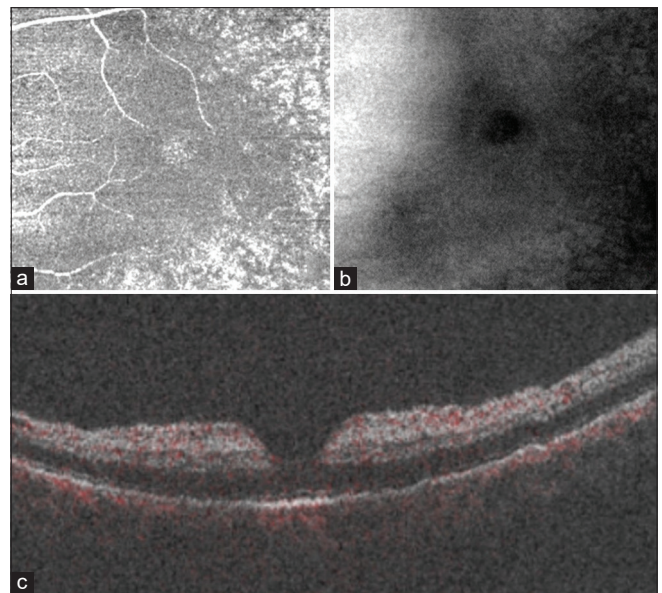


Figure 3: Optical coherence tomography angiography (a - angio scan, b - en face, c - structural OCT) [6 × 6 mm] shows decreased capillary perfusion in superficial I vascular plexus at posterior pole with inner retinal hyper-reflectivity in the structural OCT

In this case, early presentation of the patient within 6 h of the acute event, being single-eyed and timely management of CRAO with gonio massage causing apparent dislodgement of the embolus could be the likely explanation for restoration of posterior retinal circulation and good visual recovery. This was followed by a prompt diagnosis and surgical resection of myxoma. To the best of our knowledge, this is the first case of a CRAO secondary to left atrial myxoma with good visual recovery reported in the literature.

Apart from the immediate efforts to restore circulation in CRAO, which is important for visual recovery, identifying the



Figure 4: Transthoracic echocardiogram showed a large mass of mixed echogenicity measuring 35 × 32 mm, attached to the lower half of the interatrial septum and posterior wall of the left atrium, consistent with the diagnosis of left atrial myxoma (villous type)

source of embolus and its management is equally important to prevent further thromboembolic events. Ophthalmologists should be aware that myxoma may cause multiple embolic problems, such as CRAO and stroke other than cardiac symptoms; once a definitive diagnosis of atrial myxoma is established, the treatment of choice remains surgical excision.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

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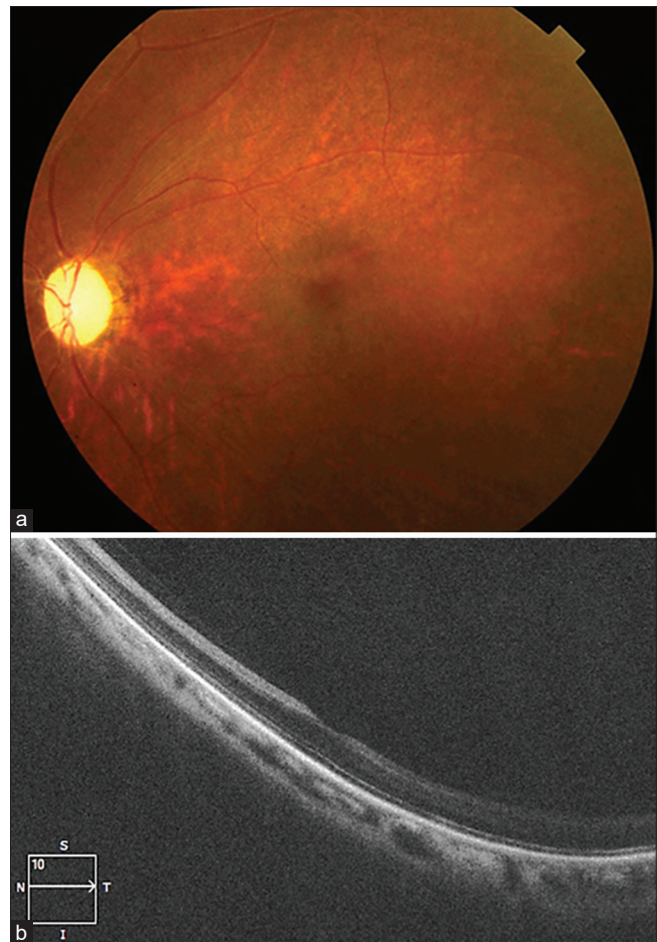


Figure 5: Color fundus photo (a) of the left eye taken at 2 weeks review showed disc pallor with attenuated vessels and resolved retinal opacification. OCT (b) showed macular atrophy