Case Report

# Combined Central Retinal Artery Occlusion and Presumed Ischemic Optic Neuropathy in Atrial Septal Defect: A Case Report

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## Abstract

A female presented with acute visual loss in the right eye. Fundus examination showed whitening retinal edema, disc swelling, and a cherry red spot in the right eye. Findings were suggestive of central retinal artery occlusion and ischemic optic neuropathy. Echocardiography revealed an atrial septal defect, which was successfully closed.

**Keywords:** Central retinal artery occlusion, Ischemic optic neuropathy, Atrial septal defect

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## Introduction

Central Retinal Artery Occlusion (CRAO) is an acute ischemic retina that is a form of cerebral stroke. Initial visual acuity ranges from counting fingers to light perception in 74-90% of cases.[1] Typical findings in CRAO include whitening retinal edema, a cherry-red spot, arteriolar attenuation, and normal optic disc. Retinal emboli can be found in up to 40 % of patients.[2] Mild optic disc swelling may be seen in 22% of patients.[3] Ischemic optic neuropathy (ION) is an acute ischemic optic nerve that also is a form of cerebral stroke due to the optic nerve being a white matter tract. Anterior ischemic optic neuropathy (AION) is characterized by concurrent optic disc swelling and requires further evaluation and establishment of the arteritic versus non-arteritic etiology.[4] The association of optic disc swelling with acute CRAO indicates a rare combination of ION and inner retinal ischemia, suggesting a vascular effect on posterior ciliary arteries that supply the optic disc.[5]

An atrial septal defect (ASD) is characterized by a failure to close the communication between the right and left atria. ASD accounts for approximately 25% of congenital heart defects in children and allows a left-to-right blood flow shunt to occur through a defect.[6] The authors report a rare case of combined CRAO and presumed ION from an atrial septal defect.

### Case Report

An 18-year-old female presented with acute painless visual loss in the right eye for 5 days. She went to the tertiary care center and she received initial treatment by ocular massage, anterior chamber tapping, oral carbonic anhydrase inhibitor, and topical beta-blockers before being referred to Thammasat University Hospital. Her vital sign; blood pressure was 100/60 mm Hg, her pulse rate was 64 bpm, and her respiratory rate was 20/min. Initial visual acuity was light perception (PL) in the right eye and 20/30 in the left eye. The intraocular pressure was 10 mm Hg in the right eye and 14 mmHg in the left eye. Slit-lamp examinations revealed a normal anterior segment with relative afferent pupillary defect (RAPD) positive grade II in her right eye. Fundus examination showed diffuse whitening retinal edema, a cherry red spot, arterial narrowing without visible retinal emboli in the right eye, and normal fundus in the left eye. The right optic nerve head had a hyperemic and swollen optic disc without peripapillary flame-shaped hemorrhages (Figure 1). Optical coherence tomography (OCT) macula showed an increased reflectivity and thickness of the inner retina in the right eye. While waiting for laboratory tests and results, her right visual acuity was worsened to no light perception (NPL) at a 2-week follow-up visit. Findings were suggestive of combined ischemic retinal and choroidal circulations. A possible diagnosis of central retinal artery occlusion and presumed ischemic optic neuropathy was made. Electroretinography (ERG) testing is not available at that time. Fundus fluorescein angiography (FFA) and indocyanine green angiography (ICGA) did not perform in the acute stage due to a malfunctioning device. FFA was performed at the 2-month follow-up visit and showed no delayed choroidal filling and no delayed arteriovenous (AV) transit time in the right eye.

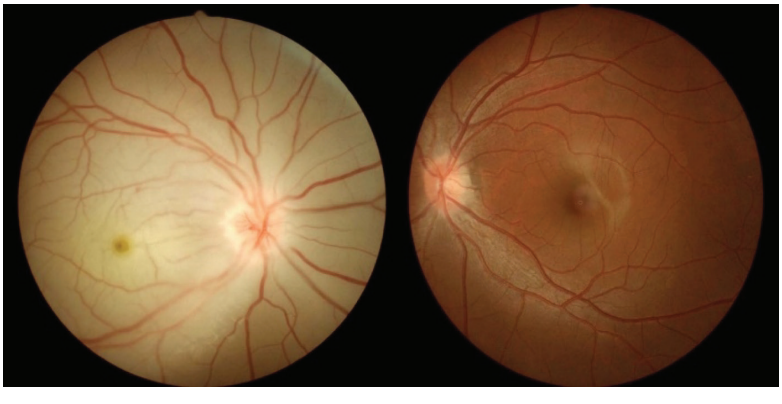

**Figure 1** Fundus examination shows diffuse whitening retinal edema, optic disc swelling, cherry red spot, arterial narrowing without visible retinal emboli in the right eye, and normal fundus in the left eye.

The following laboratory analyses were unremarkable: complete blood count, fasting blood sugar, lipid profiles, coagulogram, erythrocyte sedimentation rate (ESR), C-reactive protein (CRP), rheumatoid factor (RF), anti-double-stranded DNA (anti-dsDNA) and antinuclear antibody (ANA). Electrocardiography (ECG) revealed sinus rhythm with a left atrial enlargement (P-mitrale). The patient was referred to a cardiologist for evaluation of the cardiac embolic source. Transthoracic echocardiography revealed a large ASD with a diameter of 25 mm with a significant left-to-right shunt. Computed tomography angiography (CTA) of the thoracic aorta revealed a large ASD measuring about 35.3 × 38.7 mm with a left-to-right shunt (Figure 2). We did not perform carotid doppler ultrasonography for evaluating atherosclerosis of the carotid artery. However, magnetic resonance imaging (MRI)/MR angiography (MRA) brain including the internal carotid artery revealed no arterial stenosis, no cerebral infarction, and no abnormal vasculatures. The patient received oral antiplatelet therapy and was referred to a cardiothoracic surgeon for surgical intervention. This defect was successfully closed, however, the visual prognosis remained poor due to severe retinal ischemia and optic disc atrophy.

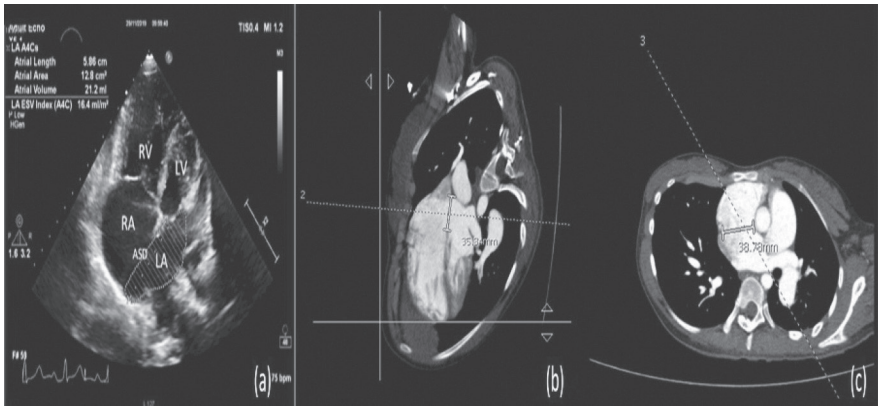


**Figure 2** (a) Transthoracic echocardiography shows a large ASD, (b) CTA of the thoracic aorta sagittal view, and (c) axial view shows ASD size of 35.3 × 38.7 mm.

ASD = atrial septal defect; LA = left atrium; LV= left ventricle; RA = right atrium; RV = right ventricle

## Discussion

The absence of light perception usually indicates either ophthalmic artery occlusion or combined retinal and choroidal artery occlusion. Ophthalmic artery occlusion usually has initial visual acuity of NPL and absence of cherry red spot as the choroidal perfusion is occluded. FFA in the acute CRAO is often found with a variable delay in arterial filling and masking of background choroidal fluorescence by retinal edema. The present study has a limitation because the patient performed FFA in the late stage. Moreover, the patient did not perform ERG which might be helpful to establish the diagnosis. A diminished a-wave is present in CRAO, but diminished a-wave and b-wave are present in ophthalmic artery occlusion. The ischemic optic neuropathy is only presumptive given there were no relevant timely FFA, ICGA, or electrophysiology findings to support the assumption which was made based on the NPL vision and concurrent optic disc swelling in this case. Our patient might be combined CRAO with presumed ischemic optic neuropathy because initial visual acuity was PL and progressed to NPL later.

Purgert et al. first reported a case that presented with segmental optic disc swelling and a whitened superior retina from simultaneous AION and hemi-retinal artery occlusion during hemodialysis.[7] They considered the possibility that multifactorial risk factors may predispose a rare combination of vascular insults. In the present study, we investigated according to the guideline of CRAO management. We ruled out giant cell arteritis (GCA) through laboratory tests such as CBC, ESR, and CRP because GCA can cause arteritic CRAO and arteritic AION. We also sent carotid imaging

to rule out carotid artery disease. Detailed cardiac evaluation should be required in patients in whom the carotid disease has been ruled out, especially in a young patient as in this case. Gupta et al. reported a rare case of CRAO from ASD.[8] They considered the possibility that intracardiac shunts through the defect may predispose the embolic source of CRAO. In patients with an uncorrected left-to-right shunt, a reverse shunt can occur due to vascular modeling as increased pulmonary pressure and resistance. In certain circumstances, the Valsalva maneuver can generate increased intrathoracic pressure as increased right atrial pressure, the left-to-right shunt will become a right-to-left shunt, resulting in transient reverse shunt into the systemic circulation. Paradoxical embolism is an uncommon cause of acute arterial occlusion, but it can be an embolic source in up to 14% of patients with ASD.[9] Paradoxical emboli due to a transient right-to-left shunt is a possible mechanism of acute atrial occlusion in this case. The patient did not improve despite receiving the initial ocular management including ocular massage, anterior chamber paracentesis, and medications to reduce IOP. Visual recovery may be irreversible if occlusion persists longer than 240 minutes.[10]

In summary, the present study demonstrated a patient with an undiagnosed heart defect who had an ocular stroke, which is a form of cerebral stroke. Detailed cardiac and carotid workups and complete laboratory tests should be investigated to rule out risk factors related to morbidity and mortality in all patients, especially young patients as in our case.

## Acknowledgments

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Conflict of interest. All authors report no conflicts of interest relevant to this article.

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