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Case report

Central retinal artery occlusion secondary to Barlow's disease

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ABSTRACT

Purpose: To report a rare case of isolated, unilateral CRAO in a young patient with mitral valve prolapse secondary to Barlow's disease.

Observations: A 29-year-old woman with history of premature ventricular contractions and cardiac ablation presented to the emergency room after sudden onset painless visual loss in her left eye (OS). Her vision was 20/20 in her right eye and hand motion in the left. Fundus exam demonstrated a central retinal artery occlusion (CRAO) OS. Computerized tomography of head and neck were unremarkable. She underwent cerebral angiogram and local intra-arterial thrombolysis. Her vision remained stable post-procedure, with marked APD and stable fundus examination. Her cardiac work-up revealed a left atrial mass with calcified mitral valve, and small atrial septal defect. Rheumatologic, hematologic, and auto-immune work-up were unremarkable. She underwent resection of the mass with repair of mitral valve and ASD closure. Surgical pathology was compatible with diagnosis of Barlow's disease, a cause of mitral valve prolapse. The patient underwent intravitreal injection of anti-VEGF therapy at one month follow-up, with vision stable at hand motion and without neovascularization on subsequent evaluation.

Conclusions: In young patients presenting with CRAO, aggressive work-up for systemic disease or embolic source must be undertaken to avoid future sequelae.

1. Introduction

Central retinal artery occlusion (CRAO) is a well-known ophthalmic emergency, analogous to a stroke of the eye. CRAO is often a harbinger of further cardiovascular morbidity and mortality, and thus merits an emergent medical evaluation beyond ophthalmic care. Though relatively uncommon with an incidence of roughly 1 in every 100,000 people, CRAO from cholesterol or thrombotic emboli has been widely described in relation to carotid, cardiac, and hypercoagulable states.[1] It is exceedingly uncommon in young patients, with few published cases in the literature.

Young patients may be more likely to have systemic or cardiac source of CRAO when compared to adults with carotid atherosclerotic disease.[1,2] In patients with an embolic cardiac source, rapid diagnosis and management must be accomplished to avoid further embolization to cerebral or more distal arterial locations. We report a case of CRAO likely from a calcific embolus in a young woman as the presenting feature of Barlow's disease. The new diagnosis led to prompt surgical intervention for this severe form of mitral valve prolapse and minimization of her risk of future embolic events.

2. Case report

A 29-year-old woman presented to the eye emergency department with sudden onset of painless visual loss in her left eye. Her past medical history was remarkable for severe premature ventricular contractions (PVC), for which she underwent cardiac catheterization and ablation several years prior. She had no significant past ocular history, nor did she report history of auto-immune disorder, deep vein thrombosis (DVT), or coagulopathy. She complained of occasional palpitations, and denied any flashes, floaters, amaurosis fugax, photophobia, temporal headaches, jaw claudication, rashes, or joint pains.

Upon initial examination, her best corrected visual acuity (BCVA) was 20/20 in her right eye (OD), and hand motion (HM) in her left eye (OS). She was found to have a 4 + afferent pupillary defect (APD) OS. Her intraocular pressures were within normal limits. Funduscopic examination was unremarkable OD and revealed cherry-red spot with surrounding retinal pallor, as well as 1 + optic nerve head edema OS. She was diagnosed with central retinal artery occlusion (CRAO) OS (Fig. 1), and emergently transferred to tertiary care inpatient center for advanced imaging and possible intravascular intervention.

After arrival, she was evaluated by the stroke team and taken for

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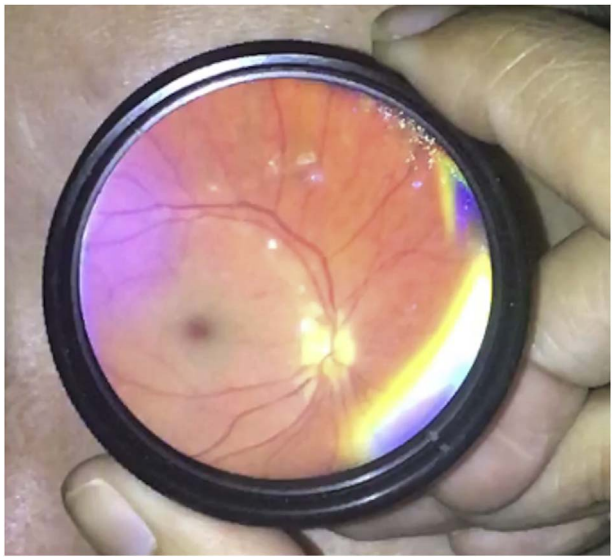

Fig. 1. Fundus photo of left eye at first presentation, performed at bedside using indirect 20D lens and cellular phone. CRAO diagnosed due to obvious retinal pallor and foveal prominence.

emergent CT scan of head and neck as well as CT angiogram. All imaging studies were unremarkable, including bilateral patency of the carotid arteries with 0% observed stenosis. The decision was made to take the patient to interventional radiology for cerebral angiogram, and intra-arterial thrombolysis (IAT) via the left ophthalmic artery. At that point, the patient was ~10 hours from onset of symptoms. Pre- and post-procedure cerebral angiography did not reveal any gross areas of ischemia. The patient tolerated the intervention well and was transferred to the stroke unit post-operatively.

On post-procedure day one her BCVA was stable at 20/20 OD and hand motion OS, with marked APD and stable funduscopic examination. Due to the absence of carotid pathology, she underwent echocardiogram which revealed a calcified mitral valve with pedunculated left atrial mass arising from a mitral valve leaflet (Fig. 2). A small septum secundum atrial septal defect (ASD) was also noted. Upon discussion with the patient's outside cardiologist, the mass appeared to have been a relatively new development which was not seen on catheterization several years prior. The cardiologist made note of a calcified mitral valve at that time, though subsequent work up for hypercalcemia was negative. The patient has no history of rheumatic fever, is from the United States, and has not travelled extensively. Her rheumatologic and hematologic evaluations were both unremarkable. She underwent ultrasonography of bilateral lower extremities which

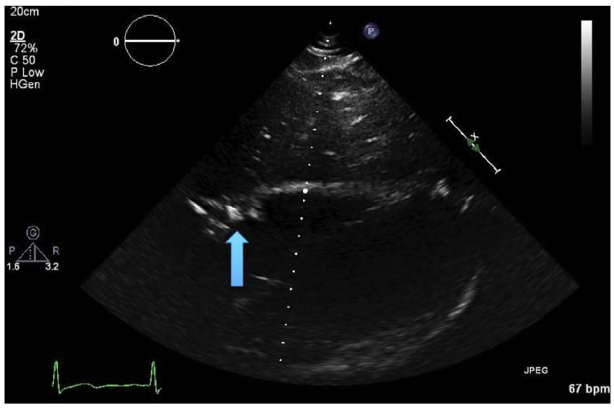

Fig. 2. Echocardiogram demonstrating mobile, pedunculated mass arising from the mitral valve (arrow).

was negative for DVT.

Upon consultation with cardiothoracic surgery, decision was made to take the patient to the operating room for resection of cardiac mass, mitral valve repair, and ASD repair. Intra-operatively, the patient was found to have Barlow's disease - a condition affecting the mitral valve - characterized by excessive myxomatous tissue formation, annular calcification, and severe billowing with prolapse of the mitral valve. The patient tolerated the procedure well, was transferred to the cardiothoracic ICU, recovered well on the cardiac floor, and was discharged home.

At post-op week 1 follow-up (10 days post-CRAO OS), the patient's BVCA remained at 20/20 and HM. An OCT macula was obtained OU which demonstrated increased thickness of the retinal nerve fiber layer (RNFL) OS relative to OD. Funduscopic exam demonstrated resolution of papillitis, with continued RNFL edema. At month 1 follow-up, the patient's vision was stable at HM OS. Repeat OCT macula demonstrated decrease in RNFL thickness OS (Fig. 3). She did not show any signs of neovascularization at 1 month. After discussion of risks and benefits of intravitreal anti-VEGF therapy, the patient underwent intravitreal injection with bevacizumab OS. The patient's vision remained at HM OS at subsequent follow-up.

3. Discussion

CRAO is a vision-threatening condition in which acute ischemia of the retina develops after interruption of blood flow through the central retinal artery (CRA). Typical presentation of a CRAO involves sudden, painless visual loss which is usually unilateral and often occurs in patients with known cardiovascular risk factors such as hypertension and diabetes mellitus.[1] BCVA in patients with CRAO is often 20/400 or worse.[1] In 26–50% of patients, a patent cilioretinal artery has been reported, which, in a fortunate subpopulation, preserves visual acuity to varying degrees depending on its contribution to papillo-macular perfusion.[1,3,4] Without this variation, visual loss is often profound. In a review of 244 cases (260 eyes) it was found that recovery from an average of count fingers vision was best in transient non-arteritic CRAO (83%), and worst in permanent non-arteritic CRAO (22%).[5] This finding suggests that the retina, while the most demanding metabolic tissue in the body, is nevertheless able to withstand short-lived ischemia. Previous animal investigations have demonstrated a retinal ischemic tolerance time of 105 minutes following total occlusion of the CRA.[6]

Most CRAOs are caused by cholesterol emboli (74%), with platelet-thrombin emboli and calcium constituting the remaining proportion.[7] In adults, emboli generally arise secondary to atherosclerosis from diabetes mellitus and hypertension, as well as from malignancy.[1,4,5,8] In younger patients, hypercoagulability such as factor V leiden deficiency, sickle cell disease, or anti-phospholipid syndrome are major causes.[1] Arrhythmias, such as atrial fibrillation, predispose patients to platelet emboli.[9] Cardiac valvular disease such as rheumatic valvular degeneration or inherited abnormalities, such as in our patient, may serve as a nidus for embolus generation.[4,8,9] One retrospective study by Greven et al. found 21 patients younger than 40 years of age with retinal artery occlusions, of which the majority (71%) were branch RAO, and 25% were CRAO.[2] Among these patients, 19% were found to have some form of cardiac valvular disease. Paradoxical embolus, though a rare cause of CRAO, has been reported, and must be investigated with ultrasonography of the lower extremities in patients with patent foramen ovale.[4,10]

In our patient, echocardiography demonstrated a classic triad for Barlow's disease, a condition characterized by myxomatous proliferation of the mitral valve, calcification of the annulus, and mitral valve prolapse.[11] Due to discrete mass on TEE, presumed diagnosis of papillary fibroelastoma was made until surgical specimen were obtained, which demonstrated calcified mass secondary to fibromyxomatous proliferation. Branch retinal artery occlusion from a mitral valve