A Rare case of Central Retinal Artery Occlusion

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A 58 year old gentleman came with sudden painless loss of vision in right eye of one day duration. He had an episode of amaurosis fugax, few hours before the actual episode. About 12 years back he had a bout of palpitation for which he was evaluated and found to be apparently normal. He was a nonsmoker with no diabetes or hypertension.

On examination his best corrected visual acuity in the right eye was perception of light, with inaccurate projection and left eye was 6/6. Anterior segment examination was normal in both eyes. Pupillary examination showed a relative afferent pupillary defect in the right eye. Fundus examination of the right eye showed pale disc, boxcarring of vessels with cloudy swelling of the posterior pole with cherry red spot at the macula suggestive of Central Retinal Artery Occlusion. Left eye fundus examination was normal (Fig. 1). Red free photograph did not show any emboli (Fig. 2).

On general examination, patient was afebrile, pulse rate 96/min, blood pressure was 130/70 mm Hg. There was no clubbing or pedal edema. Cardiovascular examination revealed pansystolic murmur at the apex with normal S1 and S2. Abdomen was soft with no organomegaly. Central nervous system examination did not reveal any abnormality.

**Investigations**

Haemoglobin 13.20 gm/100ml, PCV 39 %, TC 17000 cells /cu mm, Polymorphs 86 %, Lymphocytes 13 %, Eosinophils 1 %, ESR 78 mm/hr, PT 14.6/12 (INR = 1.22) seconds; Random Glucose 101 mg%, Serum Urea Nitrogen 11 mg %, Serum Potassium 4.3 m Eq/L, Serum Sodium 130 m Eq/ L, Creatinine 1.0 mg dl, Albumin SGOT 47 U/L, SGPT 65U/L, Bilirubin Total 1.17 mg %, Bilirubin Direct 0.35 mg %, Total Proteins 6.8g %, Albumin 3.8 g % Globulin 3.0g %, Alkaline Phosphatase 92 IU/L, Creatine phosphokinase (CPK) 20, Platelet Count 2.19 L/cu mm. Carotid Doppler was normal. Optical coherence tomography showed retinal thickening and increased signals at macula.

Patient was referred for a cardiology evaluation. ECG was normal. Echocardiogram showed mitral regurgitation, flail and ruptured posterior mitral leaf with torn chordae. A linear structure attached to the tip of posterior mitral leaf on underlying torn chordae suggestive of vegetation was noted (Fig. 3). Blood culture showed growth of enterococcus.

Patient was then diagnosed to have enterococcus related infective endocarditis involving the mitral valve. He was started on injection IV crystalline pencillin 5 million units 4th hourly and injection gentamycin 60 mg I.V 8th hourly to be continued for 6 weeks. At 4 weeks follow up CRAO appeared resolving but vision remained the same. His general condition had also improved. Fundus fluoroscein angiography was deferred as physician fitness for the procedure could not be obtained.

**Discussion**

In 1859 Von Graefe first described central retinal artery occlusion (CRAO) as an embolic event to the central retinal artery in a patient with endocarditis. In 1868, Mauthner suggested that spasmodic contractions could lead to retinal artery occlusion. There is a multitude of...
causes of Central Retinal Artery Occlusion, but patients typically present with sudden, severe, and painless loss of vision. Central Retinal Artery Occlusion is found in 1 per 10,000 outpatient visits. Of the patients, 1-2% present with bilateral involvement.

Patients with visualized retinal artery emboli, whether or not obstruction is present, have a 56% mortality rate over 9 years, compared to 27% for an age-matched population without retinal artery emboli. Life expectancy of patients with Central Retinal Artery Occlusion is 5.5 years compared to 15.4 years for an age-matched population without Central Retinal Artery Occlusion. Men are affected slightly more frequently than women. Mean age of presentation is in the sixth decade. Causes of Central Retinal Artery Occlusion vary depending on the age of the patient.

In many instances it is impossible to ascertain the exact pathophysiologic process responsible for a central retinal artery obstruction. The main causes are emboli, intraluminal thrombosis, vasculitis, spasm, hypertensive arterial necrosis, dissecting aneurysm. These causes are intimately related to associated systemic abnormalities like diabetes mellitus (25%) hypertension (66%) and cardiac valvular disease (25%). The emboli are visible within the retinal arterial system in about 20 to 40%
of eyes with Central Retinal Artery Occlusion. An emboli can originate from any part of the arterial system. The most common is cholesterol emboli from atherosclerotic deposits in carotid arteries. The other emboli are fibrin–platelet thrombus and calcific emboli which usually originate from cardiac valves and cause more severe obstruction. Abnormalities in the cardiac valves or circulation as seen in infective endocarditis should be ruled out in every case of central retinal artery occlusion. Embolus from the heart is the most common cause of central retinal artery occlusion in patients younger than 40 years.

Infective endocarditis occurs due to microbial infection of a heart valve, the lining of a cardiac chamber or blood vessel, or a congenital anomaly (example Septal defect). The most common organism is streptococci (Viridans -30-40 % and enterococci -10-15 %). Patients can develop persistent fever, stroke, central retinal artery occlusion, and cardiac failure. Clinically they present with Roth’s spots, subconjunctival haemorrhage, splenomegaly, digital clubbing, hematuria, petechial rash and loss of pulses. In the heart the affected valves develop vegetations composed of organisms, fibrin and platelets, and the vegetation may become large enough to cause obstruction or may break away as emboli. The diagnosis is confirmed by positive blood culture, high erythrocyte sedimentation rate, C-Reactive protein, Echocardiography and is treated with high doses of broad spectrum antibiotics for 4 to 6 weeks.

In this particular patient, central retinal artery occlusion was the only extra cardiac manifestation of infective endocarditis. To have such an isolated presentation is very rare and no such cases have been reported so far (Medline search, Google search 1980-2007). The incidence of such an association has not been reported. The importance of this particular case lies in the fact that the patient has first presented to an ophthalmologist with this life threatening condition. This further emphasizes the fact that every patient with central retinal artery occlusion should have a thorough systemic evaluation and follow up.

References