Management of Central Retinal Artery Occlusion in Young Adults as a Sight Threatening Disease: a case series

ABSTRACT

Introduction :

Central Retinal Artery Occlusion (CRAO) is an ocular emergency that causes sudden visual loss. Common systemic risk factors for retinal arterial occlusive disease include hypertension, diabetes, lipid disorders, and cardiac and systemic atherosclerotic disease. In young patients with CRAO, etiologic factors are more often obscure. Central retinal artery occlusion continues to be a challenging disease entity to treat. Spontaneous resolution can occur up to 22% of patients and has been reported to occur up to 3 days after initial onset.

Purpose :

To report management of CRAO in young adults as a sight threatening disease.

Case :

First patient, female 25 years old, presented with suddenly blurred vision in her right eye (RE) since 1 day ago, with best corrected visual acuity (BCVA) was 1/300 on the right eye (RE) and left eye (LE) was 0,63. Had migraine and thypoid fever history. Optical Coherence Tomography (OCT) shows macular edema.

Second patient, female 33 years old, presented with suddenly blurred vision in her left eye (LE) since 3 day ago, with best corrected visual acuity (BCVA) was 0,32 on the right eye (RE) and left eye (LE) was 2/60. Had migraine and hypertension. Optical Coherence Tomography (OCT) shows within normal limit.

Both patients showed color fundus photograph that there were cherry red spot and posterior pole retinal opacity/ whitening, Optical Coherence Tomography Angiography (OCT A) shows decreased vascular perfusion in the superficial and deep capillary plexuses. Both patients have been managed with ocular massage, paracentesis, and medical management. Outcome of both patients have visual improvement after follow up of 1 month.

Conclusion :

Simple therapeutic approach should be performed, such as reducing IOP by ocular massage, administrating IOP lowering medications, and performing anterior chamber paracentesis. Spontaneous resolution can occur up to 22% of patients and has been reported to occur up to 3 days after initial onset. Further studies are needed for evaluate management of CRAO in young adults as a sight threatening disease.

I. Introduction

Central Retinal Artery Occlusion (CRAO) is an ocular emergency that causes sudden visual loss. The estimated incidence of CRAO is reported to be roughly 1 in 10,000 cases at tertiary referral centers, being even lower for the general population, at approximately 8.5 cases per 100,000. Similar to other vascular disorder, CRAOs are largely seen in older adults, but cases in children and young adults have been reported. Common systemic risk factors for retinal

arterial occlusive disease include hypertension, diabetes, lipid disorders, and cardiac and systemic atherosclerotic disease. In young patients with CRAO, etiologic factors are more often obscure. Brown et al reported, 30% of patients less than 30 years old who had CRAO had a history of migraine headaches and autoimmune disease (antiphospolipid antibody syndrome, sickle cell syndrome, and systemic lupus erythematosus/SLE).[1-4]

The hallmark of arterial occlusive disease of the retina on clinical examination is ischemic retinal whitening, and on fluorescein angiography is delayed perfusion of the affected vessel. Central retinal artery occlusion continues to be a challenging disease entity to treat. Simple therapeutic approach should be performed, such as reducing Intra Ocular Pressure (IOP) by ocular massage, administrating IOP lowering medications, and performing anterior chamber paracentesis. Spontaneous resolution can occur up to 22% of patients and has been reported to occur up to 3 days after initial onset. However, less than 10% of patients report meaningful visual recovery. Rarely do patients have complete spontaneous recovery.[1-3]

II. Case Report

2.1 Case one

Ms. B, 25 years old, went to vitreoretinal unit in RSMC on 11 June 2017 with chief complain of suddenly blurred vision in her RE since 1 day, high minus lens history + (OD S-10.00, OS S-13.00), migraine +, thypoid fever history +. She denied floater, photopsia, pain, red eye, trauma, diabetes, hypertension, cardiac, cholesterol, smoke, and alcohol.

Vital sign was within normal limits. Best corrected visual acuity (BCVA) was 1/300 in her RE and 1/60 in her LE. The visual acuity with her glasses was 1/300 in her RE and 0,63 in her LE with Snellen chart. Intraocular pressure measurement in RE and LE was within normal limit. Anterior segment examination was within normal limit. Posterior segment examination using dilated indirect funduscopy showed macular edema, cherry red spot, latice, and myopic fundus in her RE while in her LE showed latice and myopic fundus.

Laboratory examination of hemogram with erythrocyte sedimentation rate (ESR), renal function tests, vasculitis screening profile (Anti ds-DNA antibody, antihistone antibody, ANA, c-ANCA, p-ANCA), homocysteine levels, antiphospolipid antibody (Ab), coagulation profile (protrombin time, APTT, bleeding time, clotting time, protein C and S levels), haemostasis physiology (D dimer), immunoserology (anti cardiolipin IgG and IgM, anti HIV), lipid profile, echocardiography, carotid Doppler, electrocardiogram, and CT/MRI brain were within normal limits. Color fundus photography examination of RE shows that there were cherry red spot and posterior pole retinal opacity/whitening, while LE shows within normal limit (Figure 2.1.1). OCT examination of RE shows that there was macular edema (Figure 2.1.2). OCT-A examination of RE shows that there was decreased vascular perfusion in the superficial and deep capillary plexuses (Figure 2.1.3).

The patient was diagnosed with CRAO OD + myopia gravior ODS+ myopic fundus ODS + peripheral degeneration ODS. She was managed with ocular massage OD + paracentesis OD. Postoperative therapy with timolol maleat 0.5% ed 2xOD + ofloxacin ed 6xOD. Postoperative follow-up of 1 day and 1 week found that visual acuity was 1/300 in her RE. Postoperative follow-up of 1 month found that visual acuity was 1/60 in her RE.

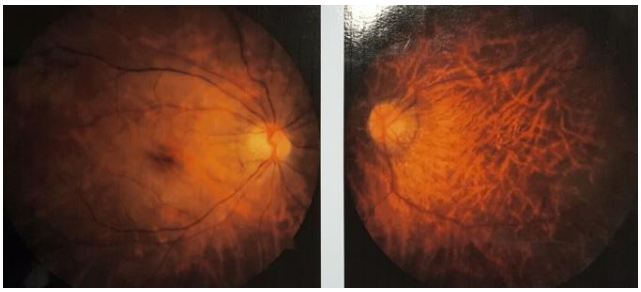

Figure 2.1.1 Color fundus photography RE shows cherry red spot and posterior pole retinal opacity/whitening, LE shows within normal limit.

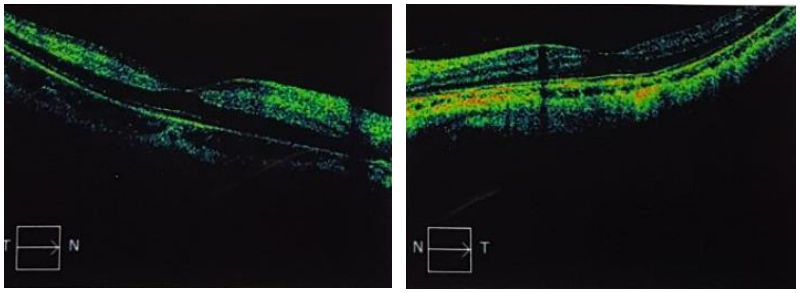

Figure 2.1.2 OCT RE shows macular edema, LE shows within normal limit

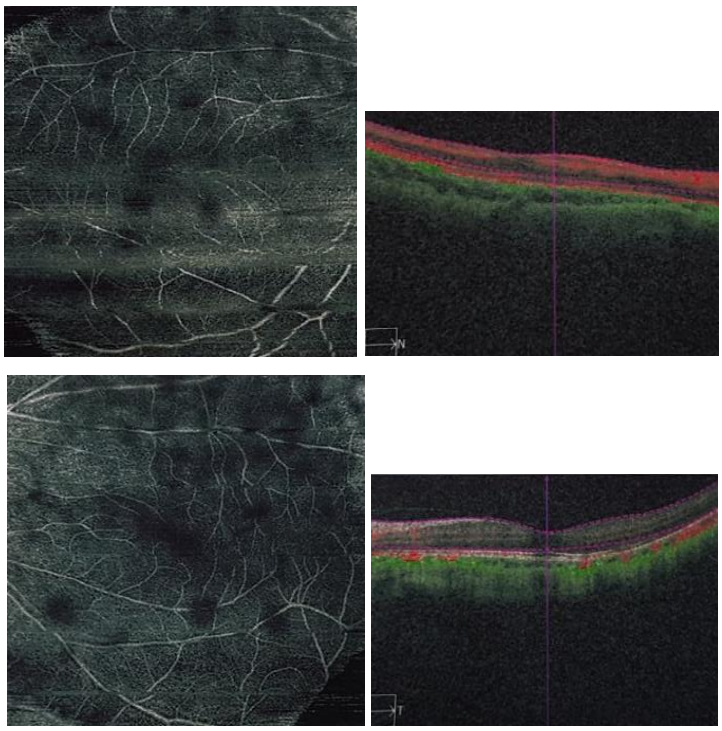

Figure 2.1.3 OCT A RE shows decreased vascular perfusion in the superficial and deep capillary plexuses, LE shows within normal limit

2.2 Case two

Mrs. P, 33 years old, went to vitreoretinal unit in RSMC on 12 June 2017 with chief complain of suddenly blurred vision in her LE since 3 day, migraine +. She had hypertension and well treated with amlodipine 5 mg and captopril 25 mg. She denied floater, photopsia, pain, red eye, trauma, diabetes, cardiac, cholesterol, smoke, and alcohol.

Vital sign was within normal limits. Best corrected visual acuity (BCVA) was 0,32 in her RE with Snellen chart and 2/60 in her LE. Intraocular pressure measurement in RE and LE was within normal limit. Anterior segment

examination was within normal limit. Posterior segment examination using dilated indirect funduscopy in her RE was within normal limit while in her LE showed cherry red spot.

Laboratory examination of hematology, erythrocyte sedimentation rate (ESR), renal function tests, coagulation profile (protrombin time, APTT, bleeding time, clotting time, protein C and S levels), lipid profile, and electrocardiogram were within normal limits. Color fundus photography examination of LE shows that there were cherry red spot and posterior pole retinal opacity/whitening (Figure 2.2.1). OCT examination shows within normal limit (Figure 2.2.2). OCT-A examination of LE shows that there was decreased vascular perfusion in the superficial and deep capillary plexuses (Figure 2.2.3).

The patient was diagnosed with CRAO OS + hypertension. She was managed with ocular massage OS + paracentesis OS. Postoperative therapy with timolol maleat 0.5% ed 2xOS + ofloxacin ed 6xOS. Postoperative follow-up of 1 day and 1 week found that visual acuity was 0,1 in her LE. Postoperative follow-up of 1 month found that visual acuity was 0,2 in her LE.

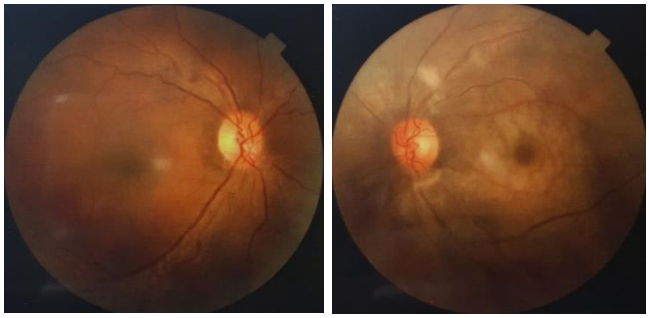

Figure 2.2.1 Color fundus photography RE shows within normal limit, LE shows cherry red spot and posterior pole retinal opacity/whitening

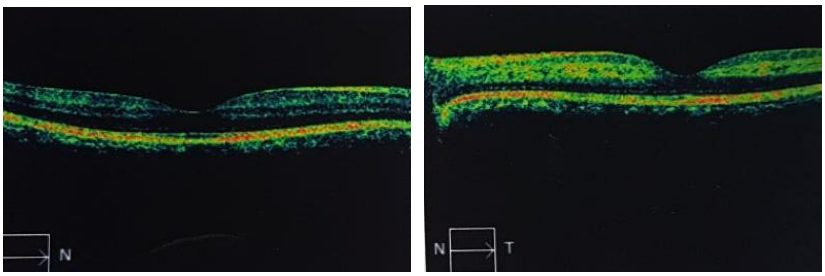

Figure 2.2.2 OCT RE and LE shows within normal limit

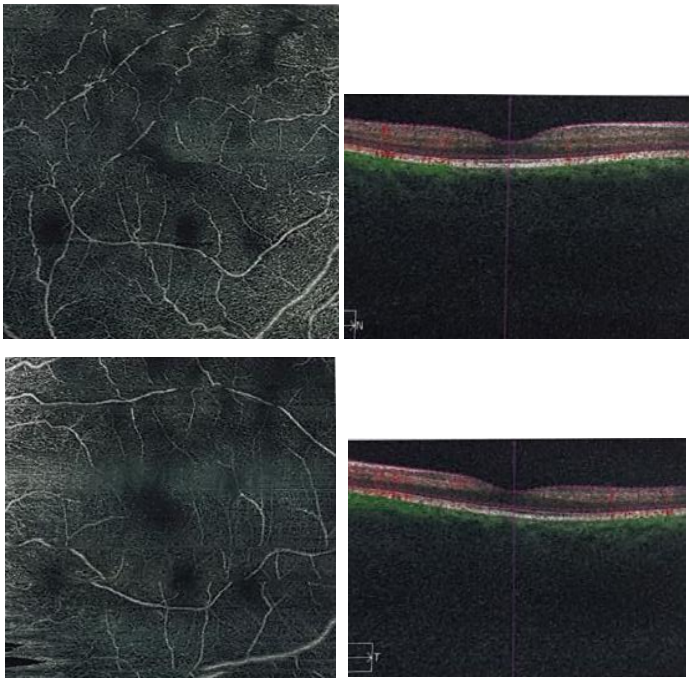

Figure 2.2.3 OCT A RE shows within normal limit, LE shows decreased vascular perfusion in the superficial and deep capillary plexuses

III. DISCUSSION

Central Retinal Artery Occlusion is an ocular emergency that causes sudden visual loss. Visual acuity of less than 20/400 occurs in more than 90% of eyes at the time of presentation. Similar to other vascular disorder, CRAOs are largely seen in older adults, but cases in children and young adults have been reported. Brown et al estimated that less than 1 in 50,000 outpatient visits to an ophthalmologist will be a patient less than age 30 with retinal arterial occlusive disease. Brown et al, showed a nearly equal number of males and females involved while Greven et al, showed a female predominance. No predilection for one eye over the other has been reported, however 1-2 % of cases may manifest bilateral. Common systemic risk factors for retinal arterial occlusive disease include hypertension, diabetes, lipid disorders, and cardiac and systemic atherosclerotic disease. In young patients with CRAO, etiologic factors are more often obscure. Brown et al reported, 30% of patients less than 30 years old who had CRAO had a history of migraine headaches and autoimmune disease (antiphospolipid

antibody syndrome, sickle cell syndrome, and systemic lupus erythematosus/SLE).[1-4]

A study by Suvajach et al, showed that ocular arterial and venous occlusion were common in patients with antiphospolipid antibody syndrome and they suggest that all young patients with retinal artery occlusion should be investigated for the same. Greven et al (4,8%) were found to have renal disorder leading to CRAO. Cardiac causes for the occurrence of CRAO have been documented. Cardiac abnormalities were found in six (18,7%) patients in Dhanashree et al study. A study by Au et al has shown presence of CRAO in 4,5% patients of SLE. Smoking and alcoholism is considered as risk factors in the development of emboli causing CRAO. In a study by Hayreh et al, in patients who smoked, a high prevalence of CRAO was seen. Sickle cell disease is a major cause of thromboembolic events in young patients. It is imperative to do a hemoglobin electrophoresis in all young patients with CRAO, 11% patients in the study by Brown et al had sickle cell disease. A study by Greven et al and Dhanashree et al have shown that hypertension is a major risk factor leading to vascular occlusion.[3-4]

Both patients came with a complaint of suddenly blurred vision of one their eyes. The first patient visual acuity was 1/300 in her RE and the second patient was 2/60 in her LE. They are both of young adult age, i.e., 25 and 33 years old. The first patient there was a migraine and thypoid fever history. Second patient there were hypertension and migraine diseases. Both patients have result of laboratorium examination within normal limits.

Schmidt and associated categorized CRAO as incomplete, subtotal, or total CRAO on the basis of the degree of vision loss, extent retinal edema, and delay in arterial blood flow. Hayreh investigated fundus changes in CRAOs in a large retrospective review in 2007 of 248 eyes of 240 patients. In the acute phase, his group noted a cherry red spot (90%), posterior pole retinal opacity or whitening (58%), box carring of retinal arteries and veins (19% and 20% respectively, retinal arteries attenuation (32%), optic disc edema (22%), and optic nerve pallor (39%). The retinal findings were

predominantly located in the posterior pole with a normal appearing periphery. Central retinal artery occlusion shows a distinct pattern on OCT. In acute stage, OCT shows increased reflectivity and thickness of the inner retina and a corresponding decrease of reflectivity in outer layer of the retina and Retinal Pigment Epithelium (RPE) layer. Optical Coherence Tomography Angiography is a method of visualizing the movement of red blood cells by analizing the changes in the intensity and/or phase signal that arise from repeated B Scans performed in the same location. Coherence Tomography Angiography can help characterize and improve the accuracy of microvascular changes in retinal vascular disease such as retinal venous occlusion and retinal arterial occlusion. Coherence Tomography Angiography can detect many of the key feature of retinal arterial occlusion, including areas of impaired capillary perfusion, vascular shunting, and some types of intraretinal edema.[2,5-8]

Color fundus photography of the two patients showed cherry red spot and posterior pole retinal opacity/whitening. First patient, OCT examination showed macular edema, while second patient showed within normal limit. OCT-A of both patients showed decreased vascular perfusion in the superficial and deep capillary plexuses.

Management of patients with CRAO can be considered in three aspects; management of the acute occlusive event in an attempt to restore visual function, workup of the patient looking for potential systemic conditions requiring treatment, and management of the remote complications and sequelae of the arterial occlusive event. The goal of treatment for acute CRAO has been the restoration of circulation before retinal ischemia has advanced irreversibly to autolysis and necrosis. Typically treatment is either conservative or invasive. Simple therapeutic approach should be performed, such as reducing Intra Ocular Pressure (IOP) by ocular massage, administrating IOP lowering medications, and performing anterior chamber paracentesis. Ocular massage can be used in an attempt to relieve the obstruction or break up the embolus so it moves distally to restrore some

blood flow to the retina. Anterior chamber paracentesis is the removal of fluid from the anterior chamber, the area just anterior to iris and lens, and immediately posterior to the cornea. Anterior chamber paracentesis is a fairly quick, simple, and safe procedure with important diagnostic and therapeutic roles. Central retinal artery occlusion, usually from an atherosclerotic embolic event, is another potential cause of visual loss that may benefit from an anterior chamber paracentesis in combination with digital ocular massage and medical management. Neubauer et al (2000) reported the results of conservative treatment of CRAO with ocular massage, paracentesis, and medical management, 15% showed distinct improvement. Magargal et al (1977) showed 25 % significant improvement with paracentesis. Rumelt et al (1999) showed 7,2% significant improvement with ocular massage and medical management. Significant improvement in visual acuity as a sustained improvement of three or more Snellen chart gradation and a final visual acuity 20/100 or better on follow up examination 1 month or more after treatment. Spontaneous resolution can occur up to 22% of patients and has been reported to occur up to 3 days after initial onset. However, less than 10% of patients report meaningful visual recovery. Rarely do patients have complete spontaneous recovery.[1,2,9,10]

Both patients were managed with a 30 minute ocular massage, paracentesis, and medical management with timolol maleate ed 2xOD/OS. Both patients were followed up 1 day, 1 week, and 1 month postoperative. First patient, postoperative follow up of 1 day and 1 week found that visual acuity was 1/300 in her RE, and 1 month follow up found an improvement visual acuity was 1/60 in her RE. Second patient, postoperative follow up of 1 day and 1 week found that visual acuity was 0,1 in her LE, and 1 month follow up found an improvement visual acuity was 0,2 in her LE.

Prognosis is dependent on several factors stage of occlusion, number of occluded retinal branches, occluding material (ie cholesterol, platelet fibrin, calcific emboli), duration of occlusion, and patient age. Transient or partial obstruction of the CRAO may lead to recovery of some or all visual function

in minutes or hour, but permanen visual loss is usually a prominent feature of complete arterial occlusion. The prognosis remained poor when total occlusion of the central retinal artery existed or if, in addition to the CRAO, a distinct hypoperfusion of the choroidal vessels was present, as shown by fluorescein angiography. By definition visual loss which resolves fully within 24 hours is a transient ischemic attack of the retinal circulation. There is evidence, however, that more than 90 minutes of ischemia will result in permanent retinal damage and concomitant visual loss. Clinically, the patient's CRAO may not be complete, in which case recovery of partial vision may occur hours or days after the onset of symptoms. The prognosis for visual recovery varies with the site of occlusion, being worse with more proximal occlusion such as those in CRAO and better with the distal occlusion. In CRAO, 70% of eyes have a final vision of 20/400 or worse.[2,11,12]

The prognosis of both patients is dubia. Duration of CRAO in first patient is 24 hours, while second patient is 3 days. They are both of young adult age, i.e, 25 and 33 years old. First patient had migraine and thypoid fever history, while second patient had migraine and hypertension. Laboratory examination of both patients showed within normal limit. Both patients have not been able to determine the stage of occlusion because it has not been examined fluorescein angiography.

IV. CONCLUSSION

Central Retinal Artery Occlusion is an ocular emergency that causes sudden visual loss. Visual acuity of less than 20/400 occurs in more than 90% of eyes at the time of presentation. Brown et al reported, 30% of patients less than 30 years old who had CRAO had a history of migraine headaches and autoimmune disease (antiphospolipid antibody syndrome, sickle cell syndrome, and systemic lupus erythematosus/SLE). Simple therapeutic approach should be performed, such as reducing IOP by ocular massage, administrating IOP lowering medications, and performing anterior

chamber paracentesis. Spontaneous resolution can occur up to 22% of patients and has been reported to occur up to 3 days after initial onset. The prognosis for visual recovery varies with the site of occlusion, being worse with more proximal occlusion such as those in CRAO and better with the distal occlusion. Further studies are needed for evaluate management of CRAO in young adults as a sight threatening disease.

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