Photo Essay

Cystoid Macular Edema with Idiopathic Acute Central Retinal Artery Occlusion in a Healthy Child

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A 5-year-old girl presented to the emergency department with a 6-day history of sudden painless vision loss in the right eye without any history of prior trauma, pain, infection, malignancy, or other known systemic diseases. Her best corrected visual acuity was no light perception (NLP) in the right eye and 20/20 in the left eye. Ophthalmic examination of the left eye was unremarkable [Figures 1b and 2b]. The right eye had relative afferent pupillary defect. Fundus examination revealed optic disc pallor, intraretinal edema at the posterior pole with a cherry-red spot, arteriolar attenuation suggestive of central retinal artery occlusion (CRAO), and cystic changes over the macula along with resolving sub-internal limiting membrane (ILM) hemorrhage temporal to the macula [Figure 1a]. Spectral-domain optical coherence tomography (Cirrus HD OCT, Carl Zeiss Meditec, Dublin, CA, USA) of the right eye showed cystoid hyporeflective spaces involving the inner retinal layers at the fovea, with increased hyperreflectivity in the inner retina secondary to the ischemic changes [Figure 2a]. Full-field electroretinography revealed delayed and reduced amplitude of scotopic and photopic responses in the right eye [Figure 3]. Systemic investigations, including a complete hemogram with erythrocyte sedimentation rate, platelet count, lipid profile, prothrombin/activated partial thromboplastin time, serum anti-thrombin-3 level, serum homocysteine level, and serum antinuclear, anticardiolipin, and antineutrophilic antibodies, were normal. Mantoux test result was negative. Cardiovascular workup, including 2D echocardiography, carotid doppler, magnetic resonance angiography, and magnetic resonance imaging of the brain and orbit, were unremarkable. At

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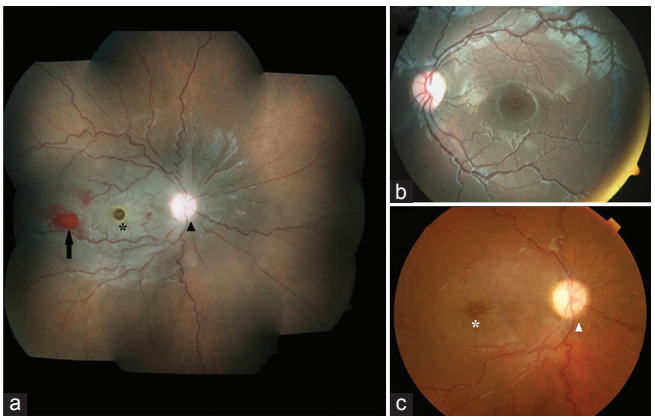

Figure 1. (a) Fundus photograph of the right eye showing a pale disc (black triangle), cherry-red spot with a cystic macula (black asterisk), posterior pole retinal edema suggestive of central retinal artery occlusion along with sub-internal limiting membrane hemorrhage temporally (black arrow), and generalized arteriolar attenuation. (b) Left eye fundus is unremarkable. (c) Right eye fundus after 1 month showing a pale disc (white triangle) and foveal thinning (white asterisk).

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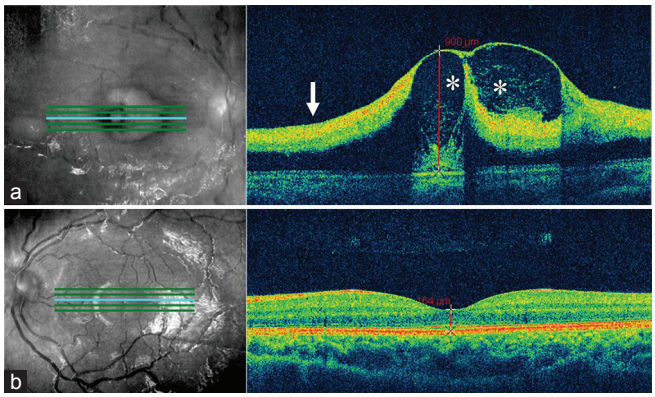

Figure 2. (a) Spectral-domain optical coherence tomographic (SD-OCT) raster scan of the right macula showing cystoid changes of the fovea (white asterisk) and inner retinal layer hyperreflectivity (white arrow) due to ischemic damage, which produces hyporeflectivity of the outer layers because of shadowing. (b) SD-OCT raster scan of the left macula.

the 1-month follow-up, fundus of the right-eye showed persistent optic disc pallor, arteriolar attenuation, and posterior pole retinal edema [Figure 1c].

DISCUSSION

The incidence of retinal arterial obstruction in patients aged <30 years is <1 in 50,000.[1] Pediatric CRAO has a poor visual outcome.[2] Detailed systemic evaluation is necessary in pediatric patients to identify the underlying causes such as hyperhomocystenemia, anti-phospholipid antibodies, polycytemia and protein C and S deficiencies. The test for serum protein C and S levels should be considered as part of the panel of tests for factors causing vessel occlusion in young patients. Protein S serves as a cofactor with another plasma protein, protein C, to inhibit the clotting cascade at the levels of factors V and VIII. The deficiency of protein S can cause venous

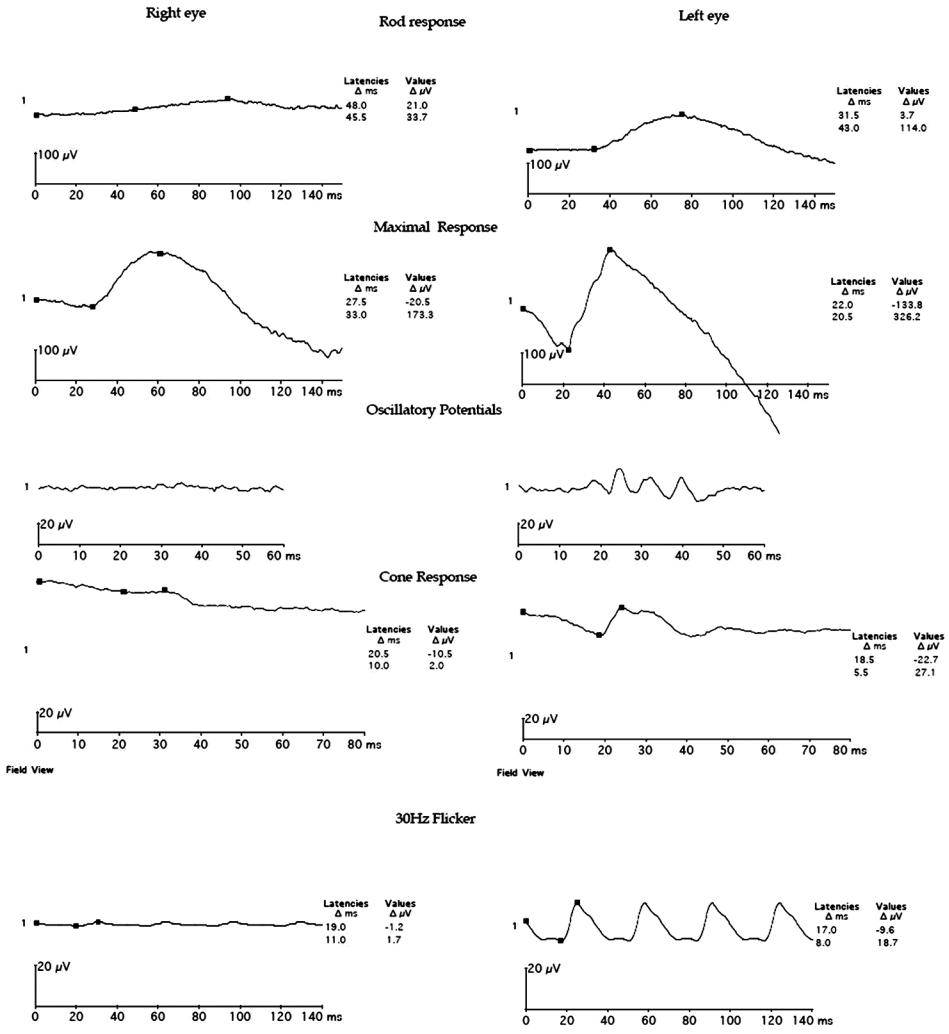

Figure 3. Full-field electroretinographic recordings of both the eyes showing delayed and reduced amplitude of scotopic and photopic responses in the right eye and normal scotopic and photopic responses in the left eye.

or arterial thrombosis in young people. To date, two reports have documented pediatric idiopathic CRAO that was assumed to be secondary to underlying inflammatory components.[2,3] Cases of cystoid macular edema (CME) with a cherry-red spot in acute CRAO without impaired choroidal circulation have also been reported in elderly patients.[4] In non-arteritic CRAO, the initial presenting vision could be NLP in 7% of the eyes without impaired choroidal circulation.[5] CRAO causes acute ischemic insult to the inner retinal layers, resulting in diffuse posterior pole intraretinal edema. Presence of CME suggests that CRAO could affect outer retinal layers along with the superficial and deep capillary plexus. Presence of subretinal fluid and outer retinal thickening in CRAO may be secondary to acute choroidal ischemia accompanying CRAO, which correlates with the choroidal thinning observed in OCT of eyes with total CRAO.[6] Since choroidal vessels supply outer retinal layers, including photoreceptors, acute choroidal iscehmia may contribute to permanent vision loss in CRAO. The presence of coexisting retinal and sub-ILM hemorrhages and tortuous veins with posterior pole retinal edema is presumably due to combined venous and arterial occlusion in this case. Hence, in cases with CRAO and CME, one should consider the possibility of combined CRAO/CRVO. Fluorescein angiography was not performed because of the unwillingness of the patient's parents for an invasive test. This is a study limitation; however, the main clinical features are clearly those of CRAO.

To conclude, we report a rare occurrence of cystoid macular edema associated with central retinal artery occlusion in pediatric patient. Since incidence is rare in this age group, one should always emphasize on evaluating with appropriate investigations to rule out underlying systemic etiologies.

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Conflicts of Interest

There are no conflicts of interest.

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