Central retinal artery occlusion in pediatric patients secondary to anomalous retinal vasculature

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Abstract

Background

The incidence of central retinal artery occlusion (CRAO) in persons under the age of 30 has been estimated at less than 1 in 50,000.

Cranial lesions in children are exceedingly rare and are seen in the setting of systemic conditions that predispose to embolic disease. These include trauma, coagulopathies, collagen vascular disease, fibromuscular dysplasia, neoplastic disease, and cardiac disease1.

Congenital prelaminar bifurcation of the central retinal artery (CRA) is a unique variant rarely described in the literature. These hemi-trunks are at much greater risk for acute occlusion and may predispose patients with other underlying risk factors to permanent vision loss.

Methods

Results

Clinical records of five children with underlying anomalous retinal vasculature were identified and retrospectively reviewed for this case series. A complete literature review was performed to further investigate this rare pathology.

The patients in this study presented symptptomatically between 11 and 16 years old. Two patients had multiple episodes of transient vision loss, and two patients had permanent vision loss. One patient remained asymptomatic. There was no predilection to sex. Basic patient demographics, prior medical history, and clinical presentation are shown in Table 1. All patients showed anomalous vessels on fundus exam. Four patients had bilateral anatomic changes and one had unilateral changes. All symptomatic patients had extensive workup including hematologic, cardiac, vascular, and neurologic. These patients all had some form of hypercoagulable predisposition to occlusion. All patients, symptomatic and asymptomatic, were treated with aqueous suppressants and antiplatelet or anticoagulation therapy once patients showed anomalous vessels on fundus exam. Four patients had bilateral anatomic occlusion and disc edema in a child. LE, Shields JA, Goldberg RE, Walsh PN. Retinal arterial obstruction in children and young adults. Ophthalmology 1981;88:18

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Table 1. Basic patient demographics, prior medical history, and clinical presentation.

<table>
<thead>
<tr>
<th>Case</th>
<th>Age at onset</th>
<th>Sex</th>
<th>Visual symptoms</th>
<th>Medical history</th>
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<tr>
<td>1</td>
<td>15</td>
<td>Male</td>
<td>Transient line in vision, blurred inferior field</td>
<td>None</td>
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<tr>
<td>2</td>
<td>11</td>
<td>Male</td>
<td>Transient line in vision, blurred inferior field</td>
<td>None</td>
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<tr>
<td>3</td>
<td>13</td>
<td>Male</td>
<td>Transient line in vision, blurred inferior field</td>
<td>None</td>
</tr>
<tr>
<td>4</td>
<td>16</td>
<td>Female</td>
<td>Transient line in vision, blurred inferior field</td>
<td>None</td>
</tr>
<tr>
<td>5</td>
<td>11</td>
<td>Male</td>
<td>Transient line in vision, blurred inferior field</td>
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</table>

Discussion

This is an exceedingly rare disease process and, to our knowledge, is the largest series reported in the literature. It has been proposed that with early bifurcation of the central retinal artery, the hemi-trunks are at much greater risk for acute occlusion. This is a newly used imaging and ocular imaging technique and should be considered as a means of safe, noninvasive, objective, and quantitative measurement of retinal vasculature. Three patients were evaluated by the same examiner (LJ) and one by another examiner (MR). Figure 11 shows color doppler ultrasound of the normal central retinal artery bifurcation.

Extensive multidisciplinary workup including hematologic, cardiac, vascular, and neurologic was performed to identify and treat all risk factors. All of the symptomatic patients in this series had some form of hypercoagulable predisposition found on workup. Three patients had history of migraine or new onset severe headache symptoms. These results coincide with the limited amount of data currently present in the literature. A case series by Breen describes coagulation abnormalities and history of migraines as the highest risk factors for central retinal artery occlusion in young adults and children. Other systemic risk factors included trauma, sickle cell hemoglobinopathies, cardiac disorders, blood disorders, and use of anticoagulants, contraceptives pills, pregnancy, systemic lupus erythematosus, and intravenous drug use. The goal of treatment was prevention of future occlusions. All patients were treated with antiplatelet, anticoagulants, and topically suppressants to improve ocular perfusion pressure. Ocular perfusion pressure is currently being investigated for its contribution to glaucomatous damage, but its role in artery occlusion has not been firmly established. The prognosis can be variable and is dependent on other factors including timing and etiology of the thromboembolic event and recanalization of the retinal vasculature.

References


