A Case of Central Retinal Artery Occlusion with Cilioretinal Artery Sparing

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Abstract: Central Retinal Artery Occlusion (CRAO) is an ophthalmic emergency involving a sudden, painless, dramatic loss of vision. Here we discuss CRAO subtypes, prognosis and current treatment strategies for use by every ophthalmic practitioner.

I. Case History
- Patient demographics: 79 year-old white male with medical history significant for hypertension, atherosclerotic disease and hypercholesterolemia.
- Chief complaint: "Pt's eye is very red, tearing, and pt is seeing around little black hole". Further, more detailed inspection revealed a sudden, painless, loss of vision approximately 30 hours ago in his left eye, awoke with drastically poor vision. He had been rubbing his left eye since the acute loss of vision, subsequently resulting in a red eye.
  Ocular history significant for cataract surgery OD/OS in 2010
-Medications:
  - Zetia 10mg/day
  - Simvastatin 10mg/day
  - Niaspan 1500 mg/day
  - Amlodipine 5mg/day
  - Bystolic 20mg/day
  - Losartan 100mg
  - Levothyroxine 50mg/day
  - Aspirin 81 mg/day

II. Pertinent findings
- Clinical: Visit #1
  - Vas cc OD= 20/30, OS= 20/100.
  - Applanation tensions: 15mmHg, 14mmHg
  - Left APD present
  - Anterior Segment was quiet, aside from mild irritation from rubbing.
  - Dilated examination showed a posterior chamber intraocular lens in each eye, vitreous cavities were clear. Right fundus was intact other than a small hemorrhage temporal to the macula. In the left eye, there was retinal edema posteriorly with a cherry-red spot. There was a small area of cilioretinal sparing which involved the fovea. The right optic nerve showed trace pallor in comparison to the left. There was no embolism noted. Fluorescein angiography of the right eye revealed normal transit in all phases. The left eye revealed delayed transit in all phases except for a small area of perfusion secondary to cilioretinal artery sparing.
  - Underwent digital massage, paracentesis.
  - Laboratory studies ordered: Carotid Duplex study, transesophageal echocardiogram, C-reactive protein and ESR.

- Clinical: Visit #2
  - VAs cc OD= 20/30, OS= CF@2ft.
    - *as measured by a technician, though further inspection by retinal surgeon revealed continued 20/100 vision.
  - Applanation tensions: 14mmHg, 12mmHg
• Left APD still present
• Anterior Segments were quiet, left eye was no longer hyperemic.
• Dilated examination showed a posterior chamber intraocular lens in each eye, vitreous cavities were clear. Right fundus, again, was intact other than a small hemorrhage temporal to the macula. The left eye now appeared intact, with a perfused retina, minimal retinal arterial attenuation, trace disc pallor, no cotton wool spots or embolism.
• Laboratory results:
  o Carotid Duplex revealed calcified plaque bilaterally with intimal wall thickening, but no significant stenosis.
  o Transesophageal Echocardiogram divulged left ventricular hypertrophy, mildly decreased systolic left ventricular function, abnormal diastolic left ventricular relaxation, moderate aortic stenosis.
  o Blood work:
    ▪ Cholesterol, Triglycerides, HDL Cholesterol, LDL Cholesterol, Cardiac risk factor all within normal limits.
    ▪ No ESR, CRP results as of yet.

- Clinical: Visit #3
  • Patient has missed his last appointment, but it is hoped that he will reschedule.

III. Differential diagnosis
  Tier I: CRAO
  Tier IB: AION (Arteritic)
  Tier II: Ophthalmic Artery Occlusion
  Double BRAOs
  Tier III: Berlin’s Edema/Commotio Retinae
  Tier IV: Lipid Storage Disease (eg: Niemann-Pick, Sandhoff’s, Tay-Sach’s).

IV. Diagnosis and discussion
  - Elaborate on the condition:
    • Review anatomy of Central Retinal Artery path from Internal Carotid; review ocular perfusion in regards to retina, ONH.
    • Visual loss from CRAO can be categorized into three subtypes: 1. Permanent CRAO, 2. Permanent CRAO with cilioretinal sparing and, 3. Transient CRAO. (Hayreh 2007)
    • Fundus appearance post CRAO varies depending on the time of presentation, and type of occlusion present. Likewise, pathophysiology and visual prognosis differ in each of the three subtypes of CRAO, underscoring the importance of awareness and appropriate clinical diagnosis.
      o In a CRAO with cilioretinal sparing, early findings (<7days) include: a cherry red spot, retinal opacity, disk edema or pallor, attenuated arteries and veins, boxcarring, and rarely, an embolus. Late findings (>30 days) include possible new appearance of cotton wool spots, sheathed arteries, but no boxcarring and even rarer to find an embolus.
      o Discussion on anatomy/pathophysiology of above-mentioned findings.
    • Risk factors include hypertension, hypercholesterolemia, diabetes, obesity, smoking, heart conditions, to list a few. High prevalence of underlying
systemic disease especially in young patients mandates extensive systemic examination.
  o  CRAO etiology in 'young' patients versus 'old'

• Etiology of the embolus varies based mostly on age of occurrence; appropriate testing in this regard includes a transesophageal echocardiogram (vs. an echocardiogram) for better viewing of a potential patent foramen ovale. Also recommended is a Carotid Duplex.
  o  Blood tests used to rule out possible inflammatory causes include ESR and C-reactive protein.

-Expound on unique features:
  • Cilioretinal artery – only present in approximately 14% of the population (Wills Eye Manual), and 25% of eyes with an acute CRAO have a cilioretinal artery.
  • Compared with permanent CRAO, permanent CRAO with cilioretinal artery spraing was associated with a lower incidence of all macular and optic disc abnormalities (Hayreh 2007).

V. Treatment, management
- Treatment and response to treatment:
  • Digital massage: Research suggests that this method of compression/decompression can give rise to large increases in retinal blood flow through reactive hyperemia, changes in flow of up to 182% have been reported, although the average change is 86%. (Ffytche, 1974)
    o  Methods of performing massage.
  • Paracentesis: Improvement of blood flow by this method of allowing aqueous escape does not amount to more than 20%. (Ffytche, 1974).
    o  Method of performing paracentesis.
  • Thrombolysis of embolus: All studies up to this point have been retrospective and uncontrolled. Most recent attempt at a controlled clinical trial called the Multicenter Study of the European Assessment Group for Lysis in the Eye (EAGLE) trial was terminated in 2008 due to 'adverse effects'. (Clinical Trials.gov)
  • Other: hyperbaric chamber, hyperventilation into paper bag, vasodilators, surgical removal of clot, topical glaucoma medication, systemic diuretics. (Biousse, 2007)

VI. Conclusion
- Fundus findings for the various subtypes of CRAO can differ, which should be kept in mind when evaluating them; awareness of multiple types of CRAO will enhance appropriate diagnosis and application of intervention, as well as prognosis.
  • Intervention comparison: effectivity of digital massage vs. paracentesis; echocardiogram vs. transesophageal echocardiogram.

VII. Bibliography:


