Retinal Artery Occlusive Disorders in the Young

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Disclosures

• None
Glimpse of each
Sudden superior field loss
Several emboli
Multiple emboli
Sudden superior field loss in a 40 year old woman
22 year old woman

Courtesy: Dr. William Mieler
24 year old male
Retinitis adjacent to the BRAO
14 year old male

Courtesy: Dr. Rick Spaide
• Young adults
• Multiple BRAO’s
• Simultaneous or recurrent
• Unilateral or bilateral
• Photopsias
Susac’s Syndrome: Ocular Symptoms

- Sudden visual field loss
- Photopsias
- Recurrent field losses
- Hearing loss
- Balance problems
- Neuropsychiatric symptoms
- Focal neurological deficits
Susac Syndrome: Ocular Symptoms

• Field loss caused by recurrent episodes of branch retinal artery occlusions

• Field loss resolves partly when some of the watershed zone recovers

• Photopsias - irregular shimmering geometric lines or shapes of light just preceding a new scotoma and confined to the area of the scotoma
Susac syndrome

- Mean age of onset 46 years (14-60 years)
- Both sexes equally affected, though some neurology literature suggests more women
- Recurrent episodes of BRAO extending sometimes to over 10 years
Idiopathic Recurrent Branch Retinal Artery Occlusion (Susac’s)

- Can occur in the mid portions or bifurcation of arterioles
- Retinal opacification due to decreased axoplasmic flow
Idiopathic Recurrent Branch Retinal Artery Occlusion (Susac’s)

- No visible emboli
- Periarterial plaques likely from extravasations of lipid through the damaged endothelium into the vessel wall
Idiopathic Recurrent Branch Retinal Artery Occlusion: Fl Angiography

Fusiform staining of the vessel wall
Idiopathic Recurrent Branch Retinal Artery Occlusion: FA

- Fusiform staining of the vessel wall
Often in areas remote from the occlusion
Instruct the photographer to scan un-involved areas
Susac: Long term appearance

- Narrowed & sheathed arterioles
- Optic atrophy
- Occasionally: New vessels on the disc, retina or iris
Retinal Non perfusion
Recurrence 3 years later

Fusiform staining
Recurrence - new area 3 years later
Visual field defects: Patchy
Peri corpus callosum lesions
Often the vessel wall staining is in the unobstructed arteriole.
Susac’s Syndrome - Etiopathogenesis

• **What is known –**
  – Affects retinal, CNS and cochlear vessels
  – No circulating antibodies found as yet
  – Responds variably to immunosuppression

• **Possibilities –**
  – Peculiar antigenicity sharing of the retinal, CNS and cochlear blood vessel wall
  – Immunologic response
Susac’s syndrome: Treatment

- **Systemic Steroids**: Initial high dose and maintenance on low dose
- **Immunosuppressive agents**: Mycophenolate mofetil, methotrexate, others
- **Immunomodulator**: Rituximab
- **IV Immunoglobulin**: monthly initially, followed by bimonthly
Embolic BRAO: heart valve

Large, calcific
Single or multiple

Color of embolus: not always accurate in correlating to source/etiology
Lodge at bifurcation
Arterio-arterial anastamosis occasionally
Cardiac origin

- Echocardiogram including transesophageal echo
- Clinical evaluation
Several immediately post procedure
Choroidal and retinal infarcts
Amalric’s triangle
During Onyx embolism of a cranial aneurysm

- Manipulation of the hardware in the carotid bulb
- Released cholesterol plaques
- Not the onyx
Pigment change: choroidal inv.
15 year old male
Autofluorescence imaging
Collar bone fracture

- Persistent foramen ovale
- 29% of general population have a persistent foramen ovale
- Paradoxical embolus
44 year old woman
CT angiogram
Birefringence of Talc

lung biopsy
Talc embolism

- IV drug user
- Patent foramen ovale
- Talc emboli
Talc Retinopathy: Usual
Giant cells & Foreign bodies
Lung (lung Tx for COPD)
Several Bilateral Emboli
Intralesional triamcinolone for a hemangioma on the face

- VA dropped to LP in both eyes
- Paracentesis done
- Vision recovered to Count fingers soon after and eventually to 20/60 in each eye
20/60 OU
16 year old male

history of left epistaxis
received 32 mg of triamcinolone acetate into the left nasal septum
Pruning of the Retinal arterioles
Patchy Choroidal obstruction
2 years Follow up: 20/80 with central scotoma
Corticosteroid Embolization of the Retina and Choroid

• reported after intralesional injections of capillary hemangiomas, chalazion, and various locations of the head and neck
Mechanism

- intrallesional pressure greater than diastolic
- retrograde arterial flow into the ophthalmic artery and downstream into the choroidal and retinal circulation
Protein S deficiency
African American woman
Sickle cell occlusion
Treatment: Exchange transfusion - Recovered
Cat Scratch Disease

Foci of retinitis and branch retinal artery occlusion
Bacillary Angioma (? VEGF)
Cat Scratch Disease

**Multifocal retinitis & papillitis**

- Children and young adults
- Acute visual loss, usually unilateral
- Antecedent illness
- Multiple white retinal lesions
- Branch retinal artery occlusion
Cat Scratch Disease

Predilection for branch retinal artery occlusion
Cat Scratch Disease

Predilection for capillary proliferation in foci of retinitis & optic neuritis
Cat Scratch Disease

Predilection for capillary proliferation in foci of retinitis & optic neuritis

FA: pseudoangiomaticous lesions
Retinitis adjacent to the BRAO
Toxoplasma retinitis
Pre papillary loop
Summary: BRAO

• Several causes
• Look out for the company it keeps
• Remember 29% have persistent foramen ovale – paradoxical embolism
• Specific angiographic appearance
• Target management based on the mechanism
• Occasionally unusual mechanisms