9.6.11. Posterior Uveitis (IV): Susac Syndrome

- SICRET syndrome
  - Small Infarctions Of Cochlear, Retinal, and Encephalic Tissue
- epidemiology
  - rare
  - young women
    - ages 16–58 years
- clinical triad
  - encephalopathy
  - hearing loss
  - branch retinal artery occlusions
- ocular presentation
  - ocular findings are highly specific
  - diffuse or localized narrowing of retinal arteries
    - peripheral retinal arteries
    - isolated retinal arteriolar involvement may occur as a very late manifestation

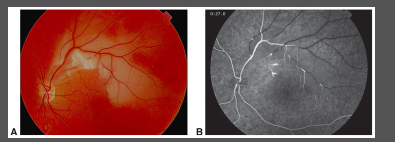

Figure 6-21 (© 2020 American Academy of Ophthalmology)

  - "boxcar" segmentation of the blood column
  - no evidence of embolic material or inflammatory reactions around the vessels
  - no vitreous haze or cells
- Differential diagnosis
  - Behçet disease
  - MS
  - herpetic encephalitis
  - acute disseminated encephalomyelitis
- Lab work-up
  - sensorineural hearing loss
    - audiometry
  - focal nonperfused retinal arterioles with hyperfluorescent walls
    - FA
  - corpus callosum may be involved
  - multifocal supratentorial white matter lesions
    - magnetic resonance imaging (MRI)
- treatment
  - high-dose intravenous corticosteroids
  - anticoagulants
  - IMT
  - +- self-limiting

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