Neuro-ophthalmic Stroke Syndromes

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Neuro-ophthalmology
Overview

- Ocular and cerebral ischemic syndromes can present with purely visual symptoms
  - Importance of recognition
  - Importance of established referral pathway
Overview

- Cerebrovascular disease
- Monocular syndromes (diagnosis and management)
  - Transient Monocular Vision Loss
  - Central and Branch Retinal Artery Occlusions
- Binocular syndromes (diagnosis and management)
  - Transient Binocular Vision Loss
  - Occipital Ischemic Stroke
Cerebrovascular Disease

- Focal cerebral ischemia
  - 5.2 events/1000 person-years in US

- Causes
  - Atherosclerotic or cardiogenic
  - Carotid circulation
    - Monocular syndromes
  - Posterior circulation
    - 24% of first-time ischemic events
    - Posterior cerebral artery: 5-10%
    - Binocular syndromes
Cerebrovascular Disease

- High morbidity and recurrence rate
  - Visual disability
    - Reading
    - Driving
    - Ambulatory safety
  - Neurologic disability
Blood supply of the visual pathway
Cerebrovascular Disease

Treatment

- Acute treatment
- Why? Early treatment leads to improved acute and long-term outcomes
- Immediate ASA
- Intravenous recombinant tissue plasminogen activator (tPA) associated with improved outcomes
  - Benefit between 3-6 hours of onset
  - Measurable significant defect
    - Not recovering
- Intra-arterial tPA
- Clot retrieval
Cerebrovascular Disease

Treatment

- Secondary prevention
  - Risk of stroke after TIA
    - within one week 5-10%
    - 11% within 90 days
    - Of those, 50% within 2 days
    - Lower with retinal TIA
  - Rate of recurrent stroke
    - 3% at one month
    - 11% at one year
  - Mortality (all cause after first stroke)
    - 7% at one week
    - 14% at one month
    - 27% at one year
Cerebrovascular Disease

Treatment

- Secondary Prevention
  - Acute treatment leads to reduced risk of recurrence
  - Vascular risk factor management
    - Hypertension
    - Hypercholesterolemia
    - Diabetes
    - Smoking
  - Antiplatelet therapy
  - Cardiac disease
    - Atrial fibrillation
  - Carotid endarterectomy
    - >70% stenosis if M/M <6%
      - Within two weeks of event
      - Carotid stenting
Monocular syndromes
Transient Monocular Vision Loss

- “amaurosis fugax”
- 19% of all TIAs
  - 14/100,000 people per year
- Pathophysiology
  - Embolic from the carotid artery
  - Ocular ischemic syndrome
  - Arteritic
  - Migraine aura
Transient Monocular Vision Loss

- Historical differential diagnosis
  - Atheroembolic
    - Sudden onset
    - Altitudinal component
      - Like a curtain
    - Usually just negative symptoms
      - Occasionally flashes, flickers, colors
    - Duration of several minutes (<15)
    - Sudden offset
    - No other symptoms
Transient Monocular Vision Loss

- Historical differential diagnosis
  - Giant cell arteritis
    - Symptoms and findings of giant cell arteritis
      - But not always
  - Migraine aura
    - ?binocular or perceived as monocular
      - Probably no such thing as “retinal migraine”
    - Normal vision and exam
Transient Monocular Vision Loss

- Historical differential diagnosis
  - Vasospastic amaurosis
    - Rare condition
      - May present with expanding blobs of vision loss
    - May occur more frequently in migraineurs
    - Retinal arteriolar spasm visualized during attacks
    - Responds to treatment with calcium-channel blockers
Transient Monocular Vision Loss

- Findings
  - Often none
  - Embolic: Retinal emboli
  - Atherosclerotic
    - Retinal arteriolar narrowing, A/V crossing changes
    - Collapse of retinal arterioles with light pressure
Retinal Emboli

- Hollenhorst plaques, cholesterol emboli
- Carotid or aortic arch atheroembolism
Retinal Emboli

- Platelet-fibrin emboli
  - Suggests carotid or valvular disease
A-V crossing changes

Retinal vasospasm
Transient Monocular Vision Loss

- Immediate testing
  - ESR, CRP to assess for giant cell arteritis
    - For older patients, and/or suspicious symptoms or findings
    - Unless emboli are present
  - Carotid duplex
    - High yield
      - 43% ipsilateral stenosis > 70%
      - 13% ipsilateral occlusion
  - EKG for atrial fibrillation
Transient Monocular Vision Loss

- MRI of brain?
  - Coexistent acute cerebral infarction in 24%
    - On diffusion weighted-imaging
    - 50% higher likelihood of acutely treatable cause
    - Role not clearly established

- Young patients without atherosclerosis
  - Polyarteritis nodosa
  - ANCA vasculitis
  - Takayasu arteritis
  - Thrombophilia
  - Myeloproliferative
Transient Monocular Vision Loss

- **Acute treatment**
  - to reduce risk of retinal arterial occlusion or stroke
  - ASA

- **Subacute treatment**
  - Depends on results of diagnostic testing
  - Carotid endarterectomy or stenting
    - For stenosis > 50%
  - Vascular risk factor/cardiac management
Transient Monocular Vision Loss

- **Management**
  - Who orders the tests?
    - Dependent on local practice and resources
  - Admission?
    - TMVL lower risk for stroke than hemispheric TIA
    - Cost issues
    - Social issues
    - Availability of outpatient investigation within 24 hrs
CRAO/BRAO

- Sudden persistent monocular vision loss
- Exam findings are always present
  - Perhaps only an APD hyperacutely
  - Retinal ischemia
  - Emboli
    - Absence of emboli does not exclude embolic cause
      - May be proximal or already passed
    - Type of embolism not sufficient to make treatment decisions
CRAO/BRAO
Retinal Emboli

- Calcific emboli
  - Originate on heart valves or aortic arch
CRAO/BRAO

- Visual prognosis
  - Likely irreversible vision loss by 1.5 hours
    - By +4 hours no likely chance of recovery
  - Overall
    - 16% transient
    - 22% improve (presenting vision CF or worse)
    - 67% improve with cilioretinal artery sparing
CRAO/BRAO

- **Acute Treatment**
  - **Historical**
    - No evidence of outcome superior to natural history
      - Ocular massage
      - Anterior chamber paracentesis
      - Acetazolamide
  - **Antiplatelet agent**
  - **Thrombolysis**
    - Intraarterial, intravenous
    - Controversial; studies problematic
    - Complications: orbital hemorrhage, stroke
CRAO/BRAO

- Acute Investigations
  - Same as TMVL, to reduce stroke risk

- Subacute Management
  - Same as TMVL, to reduce stroke risk
  - Most common cause carotid atherosclerosis
Binocular Syndromes
Blood supply of the visual pathway
Occipital Lobe Anatomy
Occipital Visual Field Defects
unilateral
Occipital Visual Field Defects

bilateral
Transient Binocular Vision Loss

- Posterior circulation atheroembolism

- Symptoms (atherosclerotic)
  - Abrupt onset hemianopic or total binocular LOV
    - Hemianopic
      - one posterior cerebral artery involved
      - Fetal PCOM
        - P1 segment absent
      - Bilateral hemianopsia
        - Top of the basilar syndrome
Transient Binocular Vision Loss

- Differential diagnosis
  - Cerebral hypoperfusion
    - Usually massive, but may be central
    - Postural hypotension, vasovagal, Stokes-Adams attacks
      - Usually associated symptoms
  - Migraine aura
    - By far most common cause of binocular vision loss
    - Duration, positive symptoms, associated symptoms
      - Occasionally just negative symptoms
        - Multiple stereotypical episodes without sequelae
  - Transient visual obscurations
    - Brief, postural, from papilledema
Transient Binocular Vision Loss

- Investigations
  - Same as for TMVL, except
    - Vascular imaging
      - MR angiography (includes carotid)
      - CT angiography (includes carotid)
      - Duplex ultrasonography
        - Vertebral arteries
        - Emboli monitoring
Transient Binocular Vision Loss

- **Prognosis**
  - 16-fold increase in subsequent stroke over 2-4 years
    - Limited data

- **Management**
  - Same as TMVL
Occipital Stroke

- Posterior cerebral artery syndrome
  - 24% of first-ever cerebral ischemic events
    - 40-54% of these caused by embolism
      - Cardiac 24-33%
      - Artery-to-artery 14-18%
Occipital Stroke

- Symptoms
  - Hemianopic vision loss
  - Asymptomatic
    - Right-sided strokes
      - Right parietal syndrome causes neglect
    - Macular-sparing
      - Visual acuity spared
        - Visual field testing is required
          - Confrontation, tangent screen, formal
  - Bumping into things
  - Changes in visual behavior
  - High morbidity
Occipital Stroke

- Investigations
  - Same as for TBVL

- Treatment
  - Acute
    - ASA
    - Thrombolyis
  - High risk interventions
    - Clot retrieval
    - Vertebral endarterectomy/angioplasty/stenosis rarely performed
Occipital Stroke

- Neurologic Prognosis
  - Patients with posterior circulation stroke may more likely have recurrences or death
  - Particularly in the first seven days
Occipital Stroke

- Visual outcome
  - 10% full recovery,
  - 40% improved
    - Especially lower and peripheral fields
    - Especially in the first month
  - 50% no recovery
    - Occupational, physical, speech therapy
    - Hemianopic prism
    - Read right (for RHH)
    - Vision retraining programs
    - controversial
Thank you!