SWOLLEN OPTIC NERVES: NOW WHAT?

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SWELLING VS. PSEUDOSWELLING

Way to differentiate:
1. Are the vessels blurred as they cross the disc margin?
2. Is there SVP?
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   Is there a splitting of retinal layers deep in the retina?

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3.
4.
5.
6. Drusen???
12/2/16

12 yoM
“In for annual eye exam”. No complains, concerns or symptoms

Ocular Hx:
- Longstanding alternating esotropia
- +3.25 with mild astigmatism OU

VA:
- OD – 20/20
- OS – 20/20
A. Pseudoswelling  
B. True swelling

My recommendation:  
- Not overly concerned

Pediatric ophthalmologist:  
- Diagnosis:  
  - Pseudopapilledema
  - Monitor & see back in 4-6 weeks to monitor for stability
SWELLING VS. PSEUDOSWELLING

- Ways to differentiate:
  1. Direct viewing of the ONH
     - Are the vessels blurred as they cross the disc margin?
     - Is there SVP?
  2. OCT
     - rNFL thickness – normal or elevated?
     - Is there a splitting of retinal layers deep in the retina?
  3. Symptoms?
  4. History?
  5. B-scan
     - Drusen???

Pseudotumor Cerebri

- AKA
  - Idiopathic intracranial hypertension

- Elevated intracranial pressure
  - Not caused by tumor, infection, or obstruction of the ventricular system
  - Increased production vs. decreased absorption

- Etiology:
  - Idiopathic (young, obese females)
  - Medications
    - Oral contraceptives, Tetracyclines, too much vitamin A
  - Trauma

Pseudotumor Cerebri

- Symptoms:
  - HA’s (90-98%)
  - Visual disturbances (72%)
    - Transient visual obscurations (TVO’s)
  - Tinnitus (20-60%)
  - N&V (30-40%)
  - Diplopia (20-30%)
  - Blurred vision
  - Abnormal color vision - rare

Pseudotumor Cerebri

- Signs
  - Papilledema – hallmark sign of PTC
    - Increased intracranial pressure -> slowing axonal transport -> accumulation of axonal contents in the NFL -> elevated ONH’s
    - Bilateral disc edema
    - Blurred disc margins
    - Obscuration of blood vessels*
    - Hyperemia of the disc
    - Venous dilation
    - Peripapillary hemorrhages & CWS
    - Paton’s lines
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- **Other signs**
  - Enlarged blind spot
  - 6th nerve palsy
    - Tends to subside as treatment is effective

- **Differential Diagnosis:**
  - Intracranial tumor/mass
  - Intracranial bleed
  - Hydrocephalus
  - Venous sinus thrombosis
  - IIH

- **Diagnosis:**
  - Clean MRI/MRV
  - Lumbar puncture
    - Elevated ICP > 250mmHg in an obese pt
    - > 200mmHg in a non-obese pt
  - Normal CSF composition
  - No other neurological findings
  - Exception -> 6th nerve palsy
  - SVP
    - Yes -> not Pseudotumor
    - No -> ?????

- **Treatment:**
  - Weight Loss*
    - Papilledema resolution with weight loss of 6% of total body weight
  - Diamox (acetazolamide)
    - 500 mg Sequels BID-QID
    - Taper as the sx’s stabilize
  - Lumbar-peritoneal shunt (CSF shunting)
  - Optic nerve sheath fenestration/decompression

- **Non-arteritic Ischemic Optic Neuropathy (NAION)**
  - Lack of perfusion to the ONH or embolic disease that affects the arteries/arterioles that supply the ONH
  - Mean age of onset = 61-66 years old

  - Associated risk factors:
    - HTN, atherosclerosis, DM, nocturnal hypotension, sleep apnea
Non-arteritic Ischemic Optic Neuropathy (NAION)

**SYMPTOMS:**
- Sudden, unilateral, painless loss of vision
- "I woke up and I can’t see out of this one eye"

**SIGNS:**
- Diffuse or segmental disc edema
- Peripapillary flame-shaped hemes
- Retinal arterial attenuation
- (+) APD
- VF defect – often inferior altitudinal

- What does the other eye look like?
  - Small nerve?
  - Small cup?

**DIAGNOSIS:**
- Normal ESR & CRP
- (-) symptoms of GCA

**DIFFERENTIAL DIAGNOSIS:**
- AAION

**TREATMENT:**
- No proven effective treatment
- Options?
  - Aspirin
  - Lower IOP??
  - Intraocular VEGF treatment

**Prognosis:**
- unilateral.....
- guarded.....but it depends on many factors
Non-arteritic Ischemic Optic Neuropathy (NAION)

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Giant Cell Arteritis

**Symptoms:**
- New onset HA
- Jaw claudication
- Scalp tenderness/pain
- Flu-like sx’s/weight loss
- Pain and stiffness in the shoulders, hips, torso
  - Polymyalgia Rheumatica (PMR)
- Sudden, severe, painless vision loss
  - Usually unilateral
  - Diplopia

**Diagnosis:**
- Clinical symptoms
- Prominent temporal artery
- Lack of temporal artery pulsation
- CBC with differential & platelets
  - ESR  males = age/2  females = (age+10)/2
  - CRP
  - Platelets
- Temporal artery biopsy

**Signs:**
- Sudden, severe, painless vision loss
  - (+) APD
  - Pale, swollen optic disc
    - Flame shaped hemes
    - CWS’s
- CRAO
- Ocular ischemic syndrome
- EOM problems

**Treatment:**
- Refer
  - IV and/or oral steroids
  - IV 250 mg i.v. q6h (1g/day) for 3 days and/or
  - Oral 1-2mg/kg/day
  - Baby aspirin

**Prognosis:**
- Extremely poor
**Optic Neuritis**

- Patient is typically < 45 years old
- Females > males

**SYMPTOMS:**
- Acute vision loss - most often unilateral
- Eye pain in/behind the eye (80-90%)
  - worsens with eye movements

**SIGNS:**
- Visible ONH swelling (33%)
- (+) APD
- Color vision abnormalities
  - red cap test
- Brightness reduction
  - brightness comparison test
- Visual field defect - often central
- ONH pallor - 4-12 weeks after onset of symptoms

**DIAGNOSIS:**
- MRI with gadolinium

**TREATMENT:**
- MRI results? Already diagnosed with MS?
  - ONTT (Optic Neuritis Treatment Trial)
    - No oral steroids
    - IV methylprednisolone (1g/day) X 3 days
      - oral steroids (1mg/kg/day) X 10-14 days
      - Taper oral steroids over 4-7 days

**Neuroretinitis**

- Unilateral vision loss in the presence of an optic neuritis and macular star

**Etiology:**
- Idiopathic (25%)
- Cat-scratch disease (60%)
  - Bartonella henselae
- Syphilis, Lyme disease, Sarcoid, Toxo, TB

- Affects all ages, 10-40 year olds most affected

**Symptoms:**
- Painless, usually unilateral visual loss
  - Starts gradual
  - Becomes more severe after about 1 week
- Prior viral-like illness (50%)
**Neuroretinitis**

- **Signs:**
  - Usually unilateral:
    - Papillitis with peripapillary and macular edema
    - Macular star develops as the disc edema resolves
    - Other inflammatory signs (cell & flare, vitreous cells)
  - Parinaud’s oculoglandular syndrome

- **Diagnosis:**
  - Clinical picture
  - History of cat scratch/bit/lick
  - Cat-scratch serology ELISA – very sensitive and specific
  - FTA-ABS, VDRL, Lyme titer, Toxo titer, ACE, ANA

- **Treatment**
  - Usually self-limiting condition in immunocompetent individuals
  - Azithromycin 500 mg p.o. for 1 day, 250 mg/day X 4 days
  - Doxycycline 100 mg p.o. BID
  - Bactrim