SWOLLEN OPTIC NERVES: NOW WHAT?

Nate Lighthizer, O.D.

Disclosures

Aerie Pharmaceuticals

Nova Ocular

Biotissue

Novartis

Diopsys

Optovue Quantel

Ellex

Reichert

EyePromise

■ RevolutionEHR

Ivantis Lumenis

Sight Sciences

Maculogix

Shire

Nidek

■ Sun Pharma

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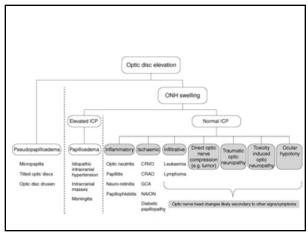
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Expected Learning Objectives

- □ To enable the ON to increase their comfort level in managing swollen optic nerves
- At end of session, attendees should be able to:
 - To become familiar with the key signs to help differentiate pseudoswelling of the ONH with true swelling of the ONH.
 - To become more familiar and update the signs, symptoms, differential diagnosis and treatment for optic neuritis and MS.

 - To become more familiar and update the signs, symptoms, differential diagnosis and treatment for non-arteritic anterior ischemic optic neuropathy

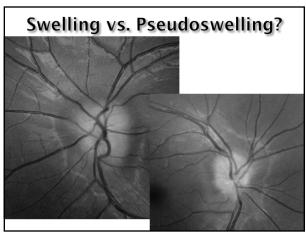
 To become more familiar and update the signs, symptoms, differential diagnosis and treatment for arteritic ischemic optic neuropathy
 - To become more familiar and update the signs, symptoms, differential diagnosis and treatment for papilledema/pseudotumor
 - To become more familiar and update the signs, symptoms, differential diagnosis and treatment for neuroretinitis.



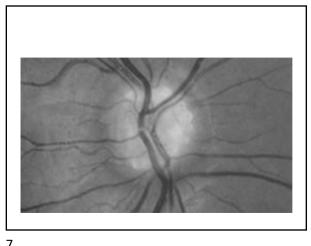
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SWELLING VS. PSEUDOSWELLING

- Ways to differentiate:
 - 1. Direct viewing of the ONH
 - Are the vessels blurred as they cross the disc margin?
 - Is there SVP?

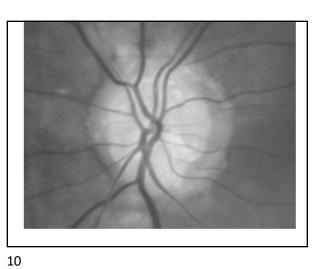


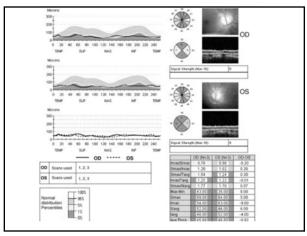
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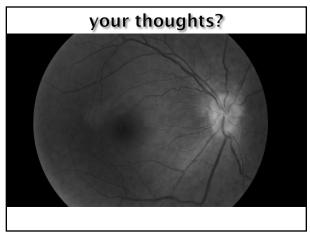


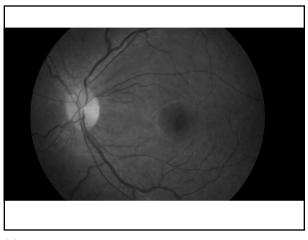


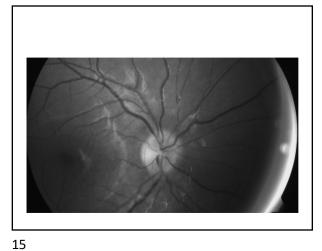












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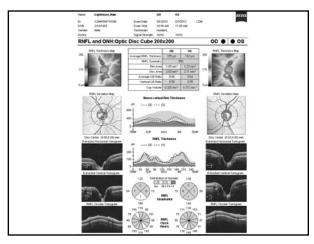




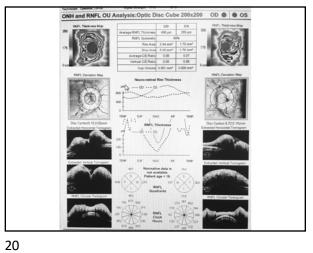
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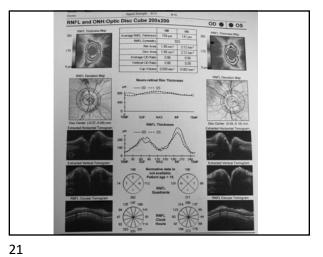
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 - ${\scriptstyle 1.} \quad \text{Direct viewing of the ONH} \\$
 - Are the vessels blurred as they cross the disc margin?
 - Is there SVP?
 - 2. OCT
 - rNFL thickness normal or elevated or thin?
 - $\ ^{\square}$ $\ ^{\square}$ Is there a splitting of retinal layers deep in the retina?



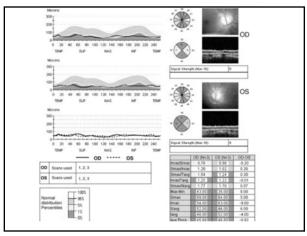
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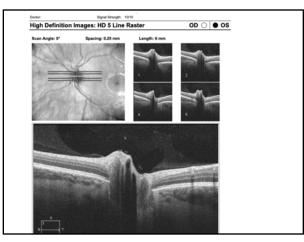




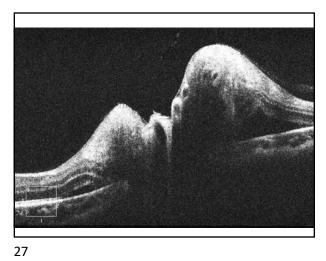


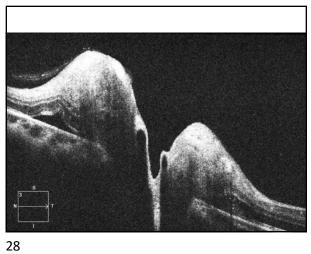


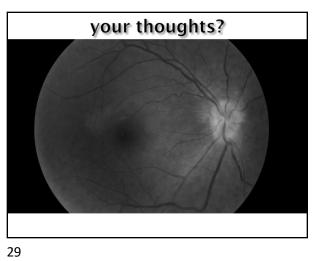




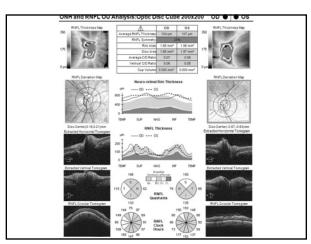


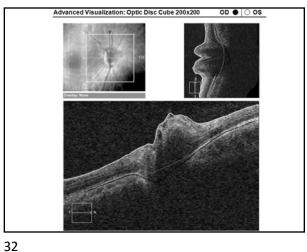


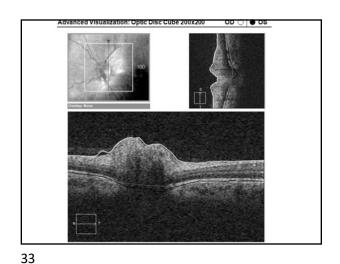








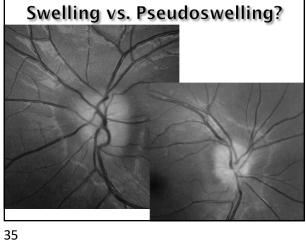




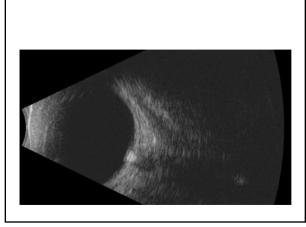
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- $\mbox{\ensuremath{}^{\circ}}\mbox{\ensuremath{}}$
- 3. Symptoms?
- 4. History?
- 5. B-scan
 - Drusen???



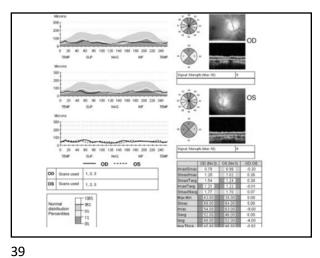
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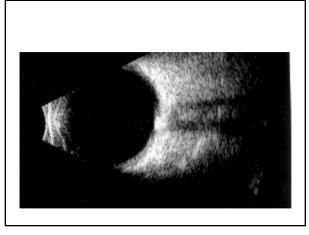


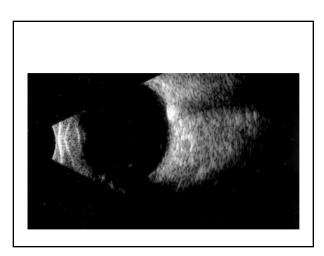
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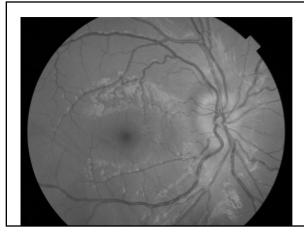




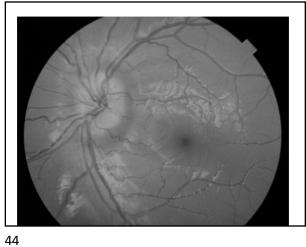
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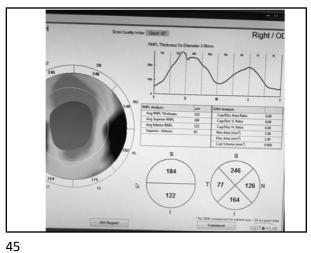
True swelling vs. Psuedoswelling case????

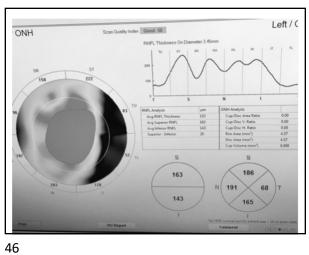
- 12 yoM
- ☐ "In for annual eye exam". No complains, concerns or symptoms
- Ocular Hx:
 - Longstanding alternating esotropia+3.25 with mild astigmatism OU
- VA:
 - OD 20/20OS 20/20

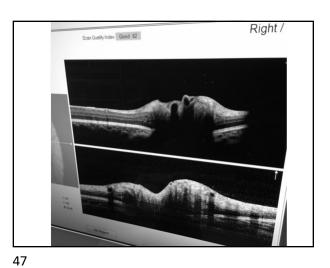


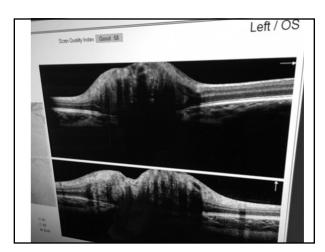
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What do you think? Pseudoswelling vs true swelling?

A. Pseudoswelling

True swelling

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True swelling vs. Psuedoswelling case????

- My recommendation:
 - see a pediatric or neuro-ophthalmologist for a second opinion
 - Not overly concerned
- Pediatric ophthalmologist:
 - Diagnosis:
 - Pseudopapilledema
 - $\mbox{\ensuremath{}^{\circ}}$ Monitor & see back in 4-6 weeks to monitor for stability

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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
 - ^o 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

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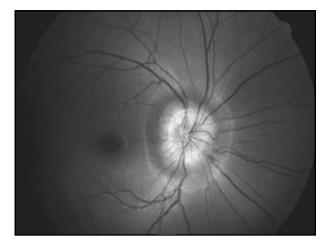
Abnormal color vision - rare

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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



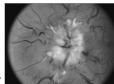




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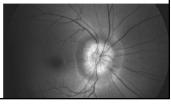
Pseudotumor Cerebri

- Other signs
 - Enlarged blind spot
 - 6th nerve palsy
 - Tends to subside as treatment is effective



Pseudotumor Cerebri

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



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Pseudotumor Cerebri

Diagnosis:

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- Clean MRI/MRV
- Lumbar puncture
 - Elevated ICP > 250mmH₂0 in an obese pt > 200mm H_20 in a non-obese pt
 - Normal CSF composition
- No other neurological findings Exception -> 6th nerve palsy
- □ Yes -> not Pseudotumor
- □ No -> ?????



Pseudotumor Cerebri

- Treatment:
 - Weight Loss*
 - $\ ^{\square}$ 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)**

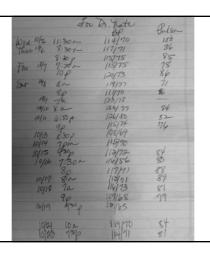
■ SYMPTOMS:

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

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Non-arteritic Ischemic Optic Neuropathy (NAION)

SIGNS:

- Diffuse or segmental disc edema
- Peripapillary flame-shaped hemes
- Retinal arterial attentuation
- (+) APD
- VF defect often inferior altitudinal
- What does the other eye look like?
 - Small nerve?
 - □ Small cup?

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Non-arteritic Ischemic Optic **Neuropathy (NAION)**

■ DIAGNOSIS:

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

AAION

Non-arteritic Ischemic Optic **Neuropathy (NAION)**

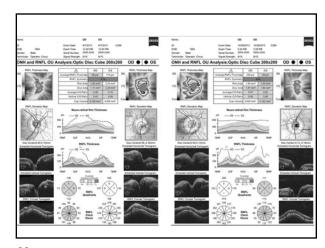
■ TREATMENT:

- No proven effective treatment
- Options?
- Aspirin
- Lower IOP??
- □ Intraocular VEGF treatment



- Prognosis:
 - unilateral.....
 - guarded.....but it depends on many factors

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Non-arteritic Ischemic Optic Neuropathy (NAION)

■ TREATMENT:

- No proven effective treatment
- Options?
 - Aspirin
 - Lower IOP??
 - □ Intraocular VEGF treatment
- Prognosis:
 - unilateral.....
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Giant Cell Arteritis

- Chronic inflammatory disorder affecting the medium-large sized cranial blood vessels
- Inflammatory mediators cause:
 - proliferation, thickening, and fibrosis of vessel walls
 inflammatory occlusion
- Risk factors:
 - Age

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- Females
- Scandinavian
- Accounts for 6% of ischemic optic neuropathy cases

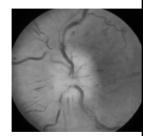
Giant Cell Arteritis

- 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

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Giant Cell Arteritis

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



Giant Cell Arteritis

- 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



Giant Cell Arteritis

- Treatment:
 - Refer
 - IV and/or oral steroids
 - ¹ 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

■ SYMPTOMS:

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

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Optic Neuritis

■ 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

Optic Neuritis

■ DIAGNOSIS:

MRI with gadolinium

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Optic Neuritis

■ TREATMENT:

- MRI results? Already diagnosed with MS?
- ONTT (Optic Neuritis Treatment Trial)
 - $\mbox{\ensuremath{\square}}$ No oral steroids
 - $^{\mbox{\tiny o}}$ IV methylprednisolone (1g/day) X 3 days
 - oral steroids (1mg/kg/day) X 10-14 days
 - Taper oral steroids over 4-7 days

Optic Neuritis

■ TREATMENT:

- MRI results? Already diagnosed with MS?
- Controlled High-Risk Subjects Avonex MS Prevention Study (CHAMPS)
 - $^{\rm o}$ IV methylprednisolone (1g/day) X 3 days
 - Avonex (interferon beta-1a)

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%)

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Neuroretinitis

- 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

Neuroretinitis

- Signs:
 - Usually unilateral:
 - Papillitis with peripapillary and macular edema
 - $\mbox{\ensuremath{}^{\circ}}$ Macular star develops as the disc edema resolves
 - $^{\tt o}$ Other inflammatory signs (ce<u>ll & flare, vitreous cells)</u>
 - Parinaud's oculoglandular syndrome

