Neuro-Ophthalmic Masqueraders

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Disclosure Statement:
Nothing to disclose
Objectives

• Differentiate neuro-ophthalmic disease from orbital disease
  – Visual fields
  – Psychological
  – Retinal disease
Case 1

• 30 year old Hispanic male

• Presenting Symptom:
  – Decreased vision OS x 5 days
Objective

- Pinhole VA: 20/20 OD, 20/100 OS
- Pupils: 5 → 3 mm OD; 5 → 4.5 mm OS
- Slit lamp exam: 2+ cells OS
- Fundus examination: “Unremarkable OU”
What would you do next?

• A. Order an MRI
• B. Order laboratory testing
• C. Follow the patient in 6 weeks
• D. Repeat exam component(s)
Junctional Scotoma
Where is the lesion located?

- A. Retina
- B. Optic nerve
- C. Chiasm
- D. Radiations
- E. Occipital lobe
Retinal Exam
Hemianopia
Binasal Loss
Case 2

- 100-year-old white female
- Seeing 3 young children, well-dressed, cute
- Smaller boy hopping next to 2 girls
- Occasionally sees herself walking with children
- Knows they are not there
- Not afraid of images
- Feels they are there to protect and watch over her
- Hx of end-stage glaucoma, both eyes
- One year ago VA: RT 20/80; LT 20/400
- Today VA: RT hand motion; LT count fingers
• Retinal exam right eye
What does the retinal exam show?

A. Diabetic retinopathy
B. Ocular ischemic syndrome
C. Central retinal vein occlusion
D. Hypertensive retinopathy
The etiology of this patient’s new-onset visual hallucinations is?

A. Alzheimer’s disease
B. Anton’s syndrome
C. Balint’s syndrome
D. Charles Bonnet syndrome
Charles Bonnet Syndrome

- Swiss philosopher described phenomenon in 1760
- 89-year-old grandfather developed complex visual hallucinations after bilateral cataract surgery
- Cognitively intact, visions were “fictions” of brain
- Charles Bonnet experienced similar visual hallucinations in his later life
Charles Bonnet Syndrome

- Mean age of incidence 75 – 80 years old
- Impaired vision – 20/60 or worse in better eye
- Most common ocular pathology – macular degeneration
- Images are recurrent and complex
- Duration: seconds to hours
- Episodes: days to years, variable frequency
- Nonfrightening, patient retains insight
- Reluctance to disclose that they are “seeing things”
Image Content

- People – most common presentation
- Animals, places, simple patterns or colors
- Color most often, can be black and white
- Static, dynamic – repetitive and stereotyped
- Autoscopy – images of themselves
Triggers And Inhibitors

• TRIGGERS
  – Fatigue
  – Stress
  – Low illumination level
  – General sensory reduction
  – Social isolation

• INHIBITORS
  – Closing or opening eyes
  – Blinking
  – Turning on a light
  – Looking for distraction
  – Hitting the hallucination
  – Shouting at the hallucination
Theories

• Release
  – Sensory deprivation leads to release of subconscious perceptions
    – engrams
  – Phantom vision like phantom limb
  – Correct patients vision – stops hallucinations
• Irritative
  – Spontaneous electrical discharge from visual cortex
  – No activity on neuroimaging to substantiate this theory
• Neuromatrix
  – Network of neurons imparts a pattern – neuro-signature
  – Changes in sensory input modulates output of neuromatrix
Treatment

• Correct ocular pathology
• Optical
• Pharmacological
  – Anti-convulsants
  – Neuroleptics
• Keep patients socially involved
Case 3

- 29 yo white female
- BCVA: 20/20 OD, 20/30 OS
- PIP: 10/10 OD; 6/10 with 80% red desat. OS
What tests should be run?

- A. Ocular B-scan
- B. Electroretinography
- C. MRI
- D. Laboratory testing
- E. Lumbar puncture
AIBSE

- Acute idiopathic blind spot enlargement
- Women, 16-50 years
- Photopsias followed by vision loss
AIBSE

- Mild visual acuity loss
- Dyschromatopsia
- RAPD
- Absolute scotoma with steep borders
AIBSE
AIBSE

- Visual acuity improves
- Photopsias resolve
- Visual field loss stable
Case 4

- 32-year-old white female
- 1 day onset acute RT eye vision loss
- Pain with movement for 1 week
- Photophobia for 2 weeks
- Hx of optic neuritis in both eyes
- Hx of relapsing-remitting multiple sclerosis
- Tx: interferon beta-1b 250mcg subQ QOD
• VA: RT count fingers; LT 20/20
• Constricted VF to finger count RT
• Fixed pupil RT, reverse RAPD
• Ishihara: RT 0/12; LT 12/12
• SLE: RT AC deep with 1+ nongranulomatous cell and flare
• Posterior-synechiae 270 degrees RT
• IOP: RT 12; LT 17
• Direct ophthalmoscopy: optic nerve appears normal
What is your diagnosis?

A. Optic neuritis recurrence
B. Uveitis secondary to multiple sclerosis
C. Uveitis induced cystoid macular edema
D. Uveitis secondary to retinal detachment
After pharmacologically breaking the synechiae, the complete fundus could be visualized.
Now, what is your diagnosis?

A. Optic neuritis recurrence
B. Uveitis secondary to multiple sclerosis
C. Uveitis induced cystoid macular edema
D. Uveitis secondary to retinal detachment
Optic Neuritis

- 90% monocular, 10% binocular
- Acute vision loss peaks 1 - 2 weeks
- Typically lasts 6 - 8 weeks
- 92% eye pain with movement
- Afferent pupillary defect
- Any type of visual field defect
- Papillitis 1/3, retrobulbar 2/3
- 30% photopsias
- Dyschromatopsia
  - Ishihara: 88% with defects
  - Farnsworth 100: 94% with defects
Optic Neuritis

• Chronic clinical features
  – 10 years: 20/20 in 74%, 20/25 to 20/40 in 18%, 20/40 to 20/200 in 5%
  – Afferent pupillary defect: 1/4 in 2 years
  – Uhthoffs phenomenon – vision loss with increase in body temperature
  – VEP: 80% delay in amplitude and latency
  – Temporal disc pallor
  – Recovery worse if male
Risk of MS from Optic Neuritis

• Presenting sign in 15% - 20%
• 5 years: 30%; 15 years: 50%
• 74% women, 34% men
• MRI abnormalities: 15 years – 72%
  – Ovoid lesions
  – Periventricular
  – Larger than 3mm
• Recurrent optic neuritis
• Perivenous sheating: 12%
• Oligoclonal bands: 56% - 69%
**Uveitis in Multiple Sclerosis**

- 10x more common than general population
- Uveitis and MS share common genetic risk factors and autoantigens
- Same type of T-cell immune response: lymphocyte infiltration of the retinal venules leads to vasculitis
Uveitis in Multiple Sclerosis

- Anterior: unilateral or bilateral granulomatous
- Posterior: occurs with perivenous sheathing
- Intermediate: vitreal inflammation
  - Snowballs: yellow-white inflammatory aggregates mid-vitreous and inferior retinal periphery
  - Snowbank: inferior pars plana exudates
  - Periphlebitis
  - Macular edema: 12% to 51%
  - Epiretinal membrane: 36%
  - Disc edema: 38.6%
  - Retinal detachment
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