GRAND ROUNDS:
VISUAL HALLUCINATIONS AND
DISTURBANCES

Amiee Ho, O.D.
Pacific University

24 Y.O. HISPANIC MALE

- CC: see circular “spot” in vision
- Only in left eye
- Doesn’t really notice it when both eyes are open
- Noticed 5 days ago while playing soccer
- Looks like an afterimage from looking at a light
- When eyes are closed, it appears as greenish white light
- Constant, stable
- Denies trauma
- “I cannot see your face, only your hair”

OCULAR AND HEALTH HISTORY

- ROS: unremarkable
- Ocular Hx: unremarkable
- Medical Hx: unremarkable
- Meds: none
- Allergies: NKDA
EXAM FINDINGS

- BCVA: OD 20/20 OS 20/20
- Pupils: PERRL (-) APD
- EOMs: SAFE OU; denies pain or double vision
- CVF: FTFW
- FDT: OD: Unremarkable OS: enlarged blindspot

EXAM FINDINGS

- Ant seg:
  - Unremarkable OU
- Post seg:
  - ONH: unremarkable OU
  - Macula: unremarkable OU
- Fundus photos
  - OCT and macula ONH – poor quality, grossly normal
  - Referred for neuro testing

FUNDUS PHOTOS

VISUAL FIELD TESTING
MACULAR OCT

ONH OCT

MULTIFOCAL ERG

DIFFERENTIALS

- Optic neuritis
- Migraine with aura
- Multiple evanescent white dot syndrome
- Acute zonal occult outer retinopathy
- Acute idiopathic blind spot enlargement syndrome
ACUTE IDIOPATHIC BLIND SPOT ENLARGEMENT SYNDROME (AIBSES)

- Fletcher 1988
- 7 patients (25-39 y.o.)
- Sudden onset of scintillations and unilateral blind spot enlargement
- Normal fundi
- Normal VA
- Normal color vision
- Normal pupillary responses

BACKGROUND

- The details:
- Young, adult, healthy, FEMALE
- Unilateral blind spot enlargement
- Photopsia

- Included under AZOOR complex?
- Multiple evanescent white-dot syndrome (MEWDS)
- Multifocal choroiditis (MFC)
- Punctate inner choroidopathy (PIC)
- Presumed ocular histoplasmosis syndrome (POHS)
- Acute macular neuroretinopathy (AMN)
- Acute zonal occult outer retinopathy (AZOOR)
MANAGEMENT

- Monitor
- No treatment
- Self-resolving

BY

89 Y.O. CAUCASIAN MALE

- CC: "I want to share a story with you. I went to the bank and I thought I saw a million dollars on the ground and tried to bend over and pick it up but realized it was not really there."
- Reports often seeing people or images that are not really there
- Images and people never talk to him
- He is fully aware images and people are not real
- He is not afraid or bothered by the images or people

OCULAR HISTORY

- Legal blindness
- H/O narrow angle glaucoma OU
- s/p bleb/LPI OU
- Tx Timolol qAM OS
- Dry AMD AREDs category III
- Tx Ocuvite with lutein (light smoker; quit ~13yrs ago)
- Hazy corneas 2/2 surgery OU
- Pseudophakia OU
- Dry eye syndrome OU
- RE and presbyopia
EXAM FINDINGS

- BCVA: OD: 20/400 OS: 20/200
  - Pt described seeing flowers, smiley faces, and little animals
- EOM: SAFE OU
- CVF: FTFC OD, OS
- Pupils:
  - OD: non-reactive surgical pupil, peaked toward 11:00
  - OS: + reactivity
  - OU: (-) APD

EXAM FINDINGS

- Slit Lamp
  - Cornea: 2+ diffuse corneal haze OD>OS; Descemet's folds OD=OS; 2-3 dense areas of stromal opacity OS <1/2mm in diameter
- IOP: OD: 10 OS: 16 @ 8:30am
- DFE:
  - Media: hazy OU (2/2 corneal haze)
  - C/D ratio:
    - OD: 0.55; extensive PPA 360
    - OS: 0.55
  - Macula:
    - OD: ring-like circular pigment mottling parafoveally
    - OS: GA nasal to macula

ASSESSMENT/PLAN

Assessment:
I. Visual hallucinations: likely Charles Bonnet Syndrome

Plan:
I. Ordered CT without contrast of orbits to r/o visual pathway or other central lesion as cause for visual hallucinations

Results: THERE ARE NO INTRAORBITAL MASSES OR OTHER VISIBLE ABNORMALITIES AND THE VISUALIZED PORTIONS OF THE OPTIC PATHWAYS ARE UNREMARKABLE IN APPEARANCE.
BACKGROUND

• Charles Bonnet Syndrome is the occurrence of visual hallucinations without having psychosis or dementia (with intact cerebral function)
  • Often associated with vision loss
  • Often elderly patients

• Charles Bonnet (1720-1793)
  • Renown Swiss naturalist philosopher and biologist
  • 1769 first to describe hallucinatory experiences of his grandfather Charles Lullin (89 yrs old)

FEATURES OF HALLUCINATIONS

• Content
  • Clear hallucinations vs. blurred real objects
  • Person, faces (regular or distorted), animals, figures, shapes, earlier stages of themselves
  • Black/white or color
  • Simple or complex hallucinations
  • Movement

• Time Course: Lasts for seconds, mins, hours
• Triggers: sensory reduction, fatigue, stress, low or bright illumination, closing eyes
• Relieving factors:
  • Disappear spontaneously, closing eyes, executing ocular saccades, looking directly at image, approaching image, conversing with image
• Patient reactions:
  • Depends on hallucinations: indifference, curiosity, irritation, or terror
  • Fear of being considered psychiatrically unstable

RISK FACTORS

• Visual impairment
  • > 20/60
  • Bilateral vision loss >> unilateral vision loss
• Other:
  • Social isolation
  • Shyness
  • Cerebrovascular disease
  • Fatigue
  • Stress
  • Suggestability

MANAGEMENT

• Examine: ophthalmic, neurological, neuropsychological
• Reassurance and counseling
• Maximizing visual function – spectacles, visual aids, surgery
• Social and Environmental factors – social isolation and sensory deprivation
• Psychological and Pharmacotherapy – hypnosis, relaxing training, anticonvulsants, antipsychotic (not universally effective)
53 Y.O. HISPANIC MALE

- POV: Service connection
- CC: "I'm blind, I can't see anything"
  - "I've lost my vision from my time in the service"
  - "You don't need to examine me, I won't be able to see anything"
- Patient was able to independently ambulate from waiting area to exam room by following me
- Ocular and Medical Hx: Unremarkable

EXAM FINDINGS

- BCVA:
  - OD: CF @ 1 ft. PH NI
  - OS: CF @ 2 ft. PH NI
- Pupils: unable, too light sensitive, refused to open eyes
- EOMs: unable to follow target
- CVF: severe constriction
- Color testing: unable

EXAM FINDINGS

- Slit lamp:
  - All structures grossly normal
  - Patient too light sensitive and refused to open eyes
- IOP: soft and equal to touch OU
- DFE:
  - All structures grossly normal
  - Patient too light sensitive and refused to open eyes
WHAT'S NEXT?

- Patient is not cooperative
- Inconclusive or poor exam findings
- Patient is losing patience

MORE TESTING

- OKN drum ~20/400
- Modified tangent visual field

MALINGERING

- Three types:
  - 1. Intentional simulation
  - 2. Hysteric – innocent but open to autosuggestions
  - 3. Exaggerating symptoms
- Motivation: Benefit
- Prevalence?

MALINGERING OR NOT?

- Rely more on OBJECTIVE testing
- Be observant
  - Eye contact
  - Ambulation
  - Hand shaking
- Pupils
- OKN drum
- VF testing at two distances
- OCT
- VEP, ERG, FA, FA, ICG

https://www.ncbi.nlm.nih.gov/pmc/articles/PMC3808926/
PE

8 Y.O. CAUCASIAN FEMALE

CC:
- “Sometimes my arm or leg looks really big”
- “My room can also grow very long and the door is so far away that I feel like I can’t get out of my room”
- “Time sometimes can feel like it is going so slow, it makes me really want to sleep”
- “It really scares me”

OCULAR AND HEALTH HISTORY
- Ocular Hx: unremarkable
- Medical Hx:
  - Neurological: Alice in Wonderland Syndrome
- Family Medical Hx:
  - Alice in Wonderland Syndrome: (+) mother, (+) grandmother
- Meds: none
- Allergies: NKDA

EXAM FINDINGS
- BCVAs: OD 20/20  OS: 20/20
- Pupils: PERRL (-) APD
- EOMs: SAFE OU
- CVF: FTFC
- CT:
  - Distance: Ortho
  - Near: Ortho
EXAM FINDINGS

- Ant seg:
  - Unremarkable OU

- Post seg:
  - O/NH unremarkable OU
  - Macula unremarkable OU

ALICE IN WONDERLAND SYNDROME

- Named after the classic novel, “Alice’s Adventures in Wonderland” by Lewis Carroll

- Definition: distorted perceptions of time and space, vision, hearing, and somesthetic sensations

  - Distortions:
    - Visual (75%): micropsia, macropsia, teleopsia, pelopsia
    - Somesthetic (10%): macrosomatognosia, microsomatognosia
    - Altered time perceptions, auditory distortions, extrapersonal misperceptions

ALICE IN WONDERLAND SYNDROME

- Rare condition

- Incidence unknown (underreporting? no diagnostic criteria)

- Most commonly reported in children

- Episodes can last ~20-30mins

- Children:
  - Epstein-Barr virus infections
  - Tends to disappear after a few years

- Adults:
  - Migraine (occurring in approximately ~15%)
  - Some cases linger into adulthood

MANAGEMENT

- Monitor
- No treatment
- Comfort
- Reassurance
- Migraine
- Self-resolving
MK

36 Y.O. CAUCASIAN FEMALE

- New patient
- CC: seeing flashes of zigzag patterns in vision
  - Started in the left eye
  - Went across vision, lost central vision for a bit
  - Then went over to right eye
  - Lasted probably an hour
  - Vision is still blurry and not the same
  - No longer seeing zigzag patterns now
  - Had a headache during this episode
- Patient was VERY nervous
  - Mom has had h/o retinal detachment

OCULAR AND HEALTH HISTORY

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EXAM FINDINGS

- Entering VAs: OD 20/20 OS: 20/20
- Pupils: PERRL (-) APD
- EOMs: SAFE OU; denies pain or double vision
- CVF: FTFC
- Anterior seg: unremarkable
- IOP: 16/17
- Posterior seg: unremarkable, no holes, tears or breaks 360
MIGRAINES WITH AURA

- Occurs in ~15-20% of migraine sufferers
- Can occur before, during or after the pain occurs
- Last ~15-60mins
- Usually described as:
  - Bright flashing dots or lights
  - Blurred spots
  - Distorted vision
  - Temporary vision loss
  - Wavy or jagged lines
- Rule out ocular pathology → refer to PCP