Grand Rounds: Visual Hallucinations and Disturbances

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PACIFIC UNIVERSITY

Statement of Disclosures

No financial disclosures
Cases

“Spot in vision”
“A million dollars!”
“String of lights”
“I’m blind”
“Arm looks really big!”
“Zigzags”

“Spot in vision”
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

BCVAs:   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 of macula and ONH – poor quality, grossly normal
Referred for neuro testing with Dr. Denise Goodwin
Fundus Photos

Visual Field Testing
Macular OCT

ONH OCT
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)
<table>
<thead>
<tr>
<th></th>
<th>AIIBSES</th>
<th>MEWDS</th>
<th>MFC w/P</th>
<th>PIC</th>
<th>POHS</th>
<th>AMN</th>
<th>AZOOR</th>
</tr>
</thead>
<tbody>
<tr>
<td>Age (yrs)</td>
<td>25-40</td>
<td>20-30</td>
<td>30-40</td>
<td>20-30</td>
<td>20-50</td>
<td>20-30</td>
<td>Young</td>
</tr>
<tr>
<td>Sex</td>
<td>F&gt;M</td>
<td>F&gt;M</td>
<td>F&gt;M</td>
<td>F&gt;M</td>
<td>F=M</td>
<td>F&gt;M</td>
<td>F&gt;M</td>
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<tr>
<td>Laterality</td>
<td>Unilateral</td>
<td>Unilateral</td>
<td>Bilateral</td>
<td>Bilateral</td>
<td>Bilateral</td>
<td>Bilateral</td>
<td>Eventually Bilateral</td>
</tr>
<tr>
<td>Lesions</td>
<td>None</td>
<td>Small, soft, gray-white dots; no scarring; macular granularity</td>
<td>White-yellow spots; mixture of old scars and new spots; CME 35%; macular CNV rare</td>
<td>Yellow spots; punched out scars; atrophic scars at macula</td>
<td>“Histo spots”; peripapillary atrophy; possible CNV</td>
<td>Reddish-brown wedge shaped lesions</td>
<td>Initially minimal, eventually chorioretinal degeneration</td>
</tr>
<tr>
<td>Photopsia</td>
<td>Yes, resolves</td>
<td>Yes, resolves</td>
<td>Yes</td>
<td>Yes</td>
<td>No</td>
<td>Yes</td>
<td>Yes</td>
</tr>
<tr>
<td>ERG</td>
<td>Abnormal</td>
<td>Abnormal</td>
<td>Abnormal</td>
<td>Abnormal</td>
<td>Unknown</td>
<td>Abnormal</td>
<td>Abnormal-progressive</td>
</tr>
<tr>
<td>VF defect</td>
<td>Enlarged blind spot-stable, persistent, no improvement</td>
<td>Enlarged blind spot</td>
<td>Paracentral scotoma</td>
<td>Paracentral scotoma</td>
<td>Depends on location of histo spots</td>
<td>Paracentral scotoma (enlarged blind spot rare)</td>
<td>Enlarged blind spot-progressive</td>
</tr>
<tr>
<td>Viral prodrome</td>
<td>No</td>
<td>Yes</td>
<td>No</td>
<td>No</td>
<td>No</td>
<td>Yes</td>
<td>Yes</td>
</tr>
<tr>
<td>Prognosis</td>
<td>Good</td>
<td>Good</td>
<td>Fair to poor</td>
<td>Good</td>
<td>Good</td>
<td>Good</td>
<td>Good</td>
</tr>
<tr>
<td>Notes</td>
<td>Reoccurrence rare; myopic</td>
<td>Reoccurrence rare; myopic</td>
<td>Reoccurs; myopic</td>
<td>Reoccurs; Similar to POHS but no serology or skin test; myopic</td>
<td>No vitreous or aqueous cells</td>
<td>Middle &amp; outer layers affected</td>
<td>Unilateral first, then bilateral; will form chorioretinal degeneration</td>
</tr>
</tbody>
</table>
Management

Monitor
No treatment
Self-resolving

“A million dollars!”
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: 1+ 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:
1. Visual hallucinations: likely Charles Bonnet Syndrome

Plan:
1. 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

https://www.sciencedirect.com/science/article/pii/S0039625702004149?casa_token=QX-E2h8hRZoAAAA:nqapI4BJv3uLoQ9a0LDD-Zzk-pIcyZLZ6zT80H78lovDEvFzHYCI0H8i8wnuc0mfGLydpWCKUQ
Risk factors

Visual impairment
- > 20/60
- Bilateral vision loss >> unilateral vision loss

Other:
- Social isolation
- Shyness
- Cerebrovascular disease
- Fatigue
- Stress
- Suggestibility

Management

Visual Hallucinations: Differential Diagnosis and Treatment

Ryan C. Teegle, B.S., Jason P. Caplan, M.D., and Theodore A. Stern, M.D.

<table>
<thead>
<tr>
<th>Features of Visual Hallucinations</th>
<th>Most Likely Etiologies</th>
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<tbody>
<tr>
<td>Simple patterns, spots, shapes,</td>
<td>Migraine, seizure, tumor</td>
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<tr>
<td>or lines; unilateral distribution;</td>
<td></td>
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<tr>
<td>associated with headache</td>
<td></td>
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<tr>
<td>Macropsia, micropsia, metamorphopsia</td>
<td></td>
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<tr>
<td>Associated with going to, or waking from, sleep</td>
<td></td>
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<tr>
<td>Confabulation of all vision</td>
<td></td>
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<tr>
<td>Frightening content</td>
<td></td>
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<td>Good insight</td>
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https://www.ncbi.nlm.nih.gov/pmc/articles/PMC2600136/
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)

“String of lights”
31 y.o. African American Male

CC: visual disturbance after a car accident 2 years ago
◇ No previous history of visual disturbances
◇ Usually occurred with lights
◇ See “bright string of lights”
◇ Very bothersome when driving – no longer drives
◇ No Hx of MRI

Ocular and Health History
Ocular Hx: black eye from airbag
Medical Hx: PTSD, anxiety
Meds: currently none
Allergies: NKDA
Social: denies alcohol consumption and drug use
Exam findings

BCVAs: OD 20/20 OS: 20/20
Pupils: PERRL (-) APD
EOMs: SAFE OU; denies pain or double vision
CVF: FTFC
  - FDT: OD: Unremarkable OS: Unremarkable
Color test: normal
Corneal sensitivity: normal and symmetric
Facial symmetry: normal and equal

Exam findings

Ant seg:
  - Unremarkable OU
Post seg:
  - ONH: unremarkable OU
  - Macula: unremarkable OU

IOP with GAT: 13mmH OD/OS
Palinopsia

**Characteristics:**

- Usually initiated by bright lights/glare
  - Brightly colored
- Associated with motion
- Associated with VF defect (exception: drug induced cases)
- Exaggerated by moving from light to dark
- Head shaking can help terminate symptoms
- Frequently accompanied by perceptual phenomenon
  - Tough texture on skin
  - Metamorphopsia of stimulus
- Usually transient symptoms (resolves in days to months)
  - Depends on etiology

Etiology

- Systemic disease
  - Classic Benign syndrome
  - Ocular syndrome
  - Cerebral plaque disease
  - Depression
  - Head trauma
  - Human immunodeficiency virus
  - Herpesvirus simplex
  - Korsakoff's syndrome
  - Leber's hereditary optic neuropathy
  - Leukemia
  - Malaria
  - Migraine
  - Multiple sclerosis
  - Narcolepsy
  - Neuroleptic malignant syndrome
  - Octopus-like gladiator
  - Peripheral hyperperfusion
  - Polio disease
  - Post-traumatic stress disorder
  - Migraine
  - Temporal lobe epilepsy
- Prescribed medications
  - Benzodiazepines
  - Mirtazapine
  - Metabolites
  - Mirtazapine
  - Naloxone
  - Paroxetine
  - Topiramate
  - Transderm
  - Zonisamide
- Effect drugs
  - Mirtazapine (mirtazapine)
  - Naloxone (naloxone)
  - Paroxetine (paroxetine)
  - Topiramate (topiramate)
  - Transderm (transderm)
  - Zonisamide (zonisamide)

[Link to related article](https://www.sciencedirect.com/science/article/pii/S1529183910002575?casa_token=UhV6jDOIZTMAAAAA:PeJwRlW8sdPqCaklnefnkS6AhE0J9l17im7aDe7D-n-R8iUrapzKqkPy5wgiyE2xWz8IZAQDxA)
Palinopsia vs Physiological afterimage

**Palinopsia**
- Greek: “again”
- Reoccurence of visual stimulus after stimulus is removed
- Rapidly moving objects leaving trails
- Symptom

**Physiologic afterimage**
- Afterimage perceived after an intense stimulus
- Lasts several seconds after removal of stimulus
- Fades away (passes through color changes)
- Negative of the stimulus

Hallucinations vs palinopsia

**Hallucinations**
- “Perception of an external stimulus without a source in the external world”

**Palinopsia**
- “Visual preservation”
- Stimulus from external world is preserved or repreceived
- Weeks may pass between original stimulus and when it resurfaces
“I’m blind”

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

BCVAs:
- 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. Hysterics – 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, FAF, FA, ICG

https://www.ncbi.nlm.nih.gov/pmc/articles/PMC3808926/
https://www.ncbi.nlm.nih.gov/pmc/articles/PMC3808926/#b1
Tests for malingering in ophthalmology

All Iscan Incegul

Ministry of Health Konya State Hospital of Instruction Eye Clinic, Konya-42090, Turkey
Correspondence to: Armanan Muh. Meram Yeni Yol 36-5 Çekik Apt. Meram Konya 42090, Turkey. allisnca@yahoo.com
Received: 2013-02-19 Accepted: 2013-07-20

Abstract
- Simulation can be defined as malingering, or sometimes functional visual loss (FVL). It manifests as either simulating an ophthalmic disease (positive simulation), or denial of ophthalmic disease (negative simulation). Conscious behavior and compensation or indemnity claims are prominent features of simulation. Since some authors suggest that this is a manifestation INTRODUCTION
Simulation or malingering can be defined as intentionally counterfeiting a disease with benefit instinct like in case of malingering, or misattributing his/her symptoms to another irrelevant clinical entity like in case of exaggerating. If the subject believes that he/she is really ill, then it is called ‘conversion reaction’ or ‘hysteria’. In case of conversion, subject really lives his/her symptoms and can’t control or even know that they are psychogenic in origin. In all cases of real simulation (malingering) or negative simulation there is only one instinct: benefit. It may be monetary or nonmonetary. It would be sometimes escape of military service or work, get reduction of court penalty, get compensation from social security agencies or insurance companies, and get unnecessary free medicines or medical equipments. This is a way of triggering an organic-like fold of

Eye Examination Techniques for Malingering Patients - A Review

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Abstract: Malingering manifest as either imitating an ophthalmic disease, or counterfeiting of ophthalmic disease. In all cases of imitating or denial of ophthalmic disease there is only one reason i.e. Benefit and advantages. Benefit may be financial or nonfinancial. Sometimes it may be the reason for escape of military service or work, depletion of court penalty, tricky way of getting compensation from insurance companies and unnecessary free medicines or medical equipments. Malingerer does everything to cheat ophthalmologist/optometrist. Commonly it is associated with concurrent diagnosis of depression, anxiety, panic attacks, fibromyalgia and psychiatric disorders.

Keywords: Malingering, Simulation, Subjective, Objective, Optokinetik Nystagmus (OKN), Visually Evoked Potential

Patient may assist or shows some misbehavior towards Ophthalmologist/Ophtometrist if we explain about their false behavior. Although at first, it is very difficult for ophthalmologist/optometrist to distinguish whether the patient would be really ill or whether they are malingering.

Listed important points can be very helpful in doubtful cases:
1. We have to Perform examination as a daily routine work smoothly and quickly. Do not let patient to know about your diagnosis or else patient may be very much attentive towards examination.
2. Note down all the complaints and symptoms as well as notice the patients behavior while he enters the ODD, posture, mental profile and reactions.
3. It will be better if you examine the patient alone without

“Arm looks really big!”

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:
- ONH: 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
“Zigzags”

36 y.o. Caucasian female

New patient

“Patient is having a retinal detachment”
36 y.o. Caucasian female

New patient
CC: “I am having a retinal detachment!”
◦ 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
ROS: unremarkable
Ocular Hx: unremarkable
Medical Hx: unremarkable
Meds: none
Allergies: NKDA
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

Usually described as:
- Bright flashing dots or lights
- Blind spots
- Distorted vision
- Temporary vision loss
- Wavy or jagged lines

Pathophysiology:
- Likely caused by spreading cortical depression

http://www.ncbi.nlm.nih.gov/pmc/articles/PMC2660156/
Migraines with aura

Prevalence of migraines: 15-29%
- ~31% of those with migraines experience aura
- 99% of those with aura experience VISUAL symptoms

Last ~15-60mins
Can occur before, during or after the pain occurs

Usually described as:
- Bright flashing dots or lights
- Blind spots
- Distorted vision
- Temporary vision loss
- Wavy or jagged lines

Rule out ocular pathology → refer to PCP

https://www.ncbi.nlm.nih.gov/pmc/articles/PMC2660156/

Summary

Acute, Idiopathic Blind Spot Enlargement Syndrome
Charles Bonnet Syndrome
Palinopsia
Malingering
Alice in Wonderland Syndrome
Migraines with aura
Thank You!
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