1) **Clinical Case History**
   a) 55 year old female
   b) moderate hyperopic astigmat
   c) 1+ Nuclear Sclerotic lens changes
   d) Fuchs Dystrophy
      i) Epithelial microcysts
      ii) Edema
      iii) Corneal guttata
         (1) Lesions between the corneal epithelium and Descemet’s membrane
         (2) Exccrescences abnormal elaborations of basement membrane and fibrillar collagen
         (3) Collagen droplet accumulations

2) **Treatment**
   a) Laser Cataract removal
      i) Insertion of toric IOL
   b) DSAEK for Fuch’s
      i) Removal of Descemet’s membrane and dysfunctional endothelium

3) **Complications**
   a) Miscalculation of IOL power resulting in 2.25 hyperopia and 5.00 of astigmatism
      i) Removal suture to relax cornea and Limbal Relaxing Incisions
         (1) Review of corneal topography
   b) Development of CME
      i) FA reviewed
      ii) Inject Ozurdex
         (1) Slow release dexamethasone capsule in posterior chamber
         (2) Resolution of CME documented by OCT’s
   c) Residual hyperopia
      i) Resolved by topography guided LASIK
d) Summary
   i) Cataract removal and DSAEK resulting in 2.25 hyperopia and 5.00 cylinder. Astigmatism resolved by relaxing sutures followed by LRI’s. CME complication resolved by Ozurdex injection. Residual hyperopia resolved by hyperopic LASIK. All conditions demonstrated by corneal photos, OCT’s, FA’s and corneal topographies.
Grand Round Case Report

Title: Charles Bonnet Syndrome secondary to Pleomorphic Xanthoastrocytoma (PXA) Status Post a Right Parieto-Occipital PXA Lesionectomy

Abstract:

Charles Bonnet Syndrome is a clinical condition characterized by complex visual hallucinations. It is more typical in the elderly with impaired vision and ocular pathology but can occur in patients that have undergone lesionectomies.

I. Case History

Patient demographics: 28 year old Caucasian female

Chief complaint: The patient reported fuzziness to her left side. Additionally, she had reported that after her neuro-surgery in March 2011, she had new visual symptoms and described seeing "vivid people and buildings" when they were not really present. She described these images much different that the visual aura she perceived prior to seizing. Additionally, she described typical visual aura with white spots in her vision lasting 2-10 minutes before her seizure occurred.

Ocular history: Unremarkable, normal ocular health and no visual field defects noted at neuro-ophthalmology appointment 2 months earlier

Medical history:

1. Localization-related (focal) (partial) epilepsy and epileptic syndromes with complex partial seizures, with intractable epilepsy
2. Pleomorphic Xanthoastrocytoma (parieto-occipital)
3. S/P Right Parieto-Occipital PXA Lesionectomy
Medications: Sumatriptan 50mg PRN, Norethindrone 0.35mg QAM, Topiramate 100mg BID, Levetiracetam 1000mg BID

Other salient information: No auditory symptoms with visual imagery

II. Pertinent findings

Clinical:

Visual acuity OD 20/20

OS 20/20

Pupils: Round and reactive to light, no afferent pupillary defect

Intraocular pressures OD 12 mmHg

OS 12 mmHg

EOMs: Smooth and full

CT: Orthophoria

Slit Lamp Examination

Lids/lashes: clear OU

Conjunctiva: clear OU

Cornea:

OD: clear

OS: clear

Anterior chamber: clear OU

Iris: clear OU

Lens: clear OU

Dilated fundus examination:

C/D ratio: (no edema or pallor OU)
OD: 0.2
OS: 0.2
Macula: clear OU
Vitreous: clear OU
Vessels: clear OU
Peripheral retina: clear OU, no intraocular metastasis noted

Humphrey visual field 24-2: inferior left quadrantopsia

Radiologic findings (No pre-operative MRI readings available)

There is a small right temporoparietal craniotomy defect with an underlying parenchymal defect involving gray and white matter of the lateral right occipital lobe. Overall impressions include postoperative changes in the lateral right occipital lobe

Differential diagnosis

Primary: Inferior left quadrantopsia, Charles Bonnet Syndrome
DDx: Visual aura with Epilepsy

III. Discussion

PXA is a rare type of astrocytic tumor that accounts for less than 1% of all primary brain tumors (1) (2). The majority of cases occur in patients less than 30 years of age (3).
The supratentorial region of the brain is the location of 98% of these tumors, however they have been reported in the cerebellum, spinal cord, and retina as well (1) (4). Of the supratentorial region in the brain, the temporal lobe is more frequently affected (39%), followed by the frontal lobe (19%), parietal lobe (14%), occipital lobe (9%) (2) (5). The most common presenting symptom of this tumor is seizure which occurs in over 70% of patients, however, other symptoms include headache, nausea, dizziness, and visual disturbance (1) (6). The most common treatment is maximum, careful surgical resection, called a lesionectomy, as was performed on the patient in this case study (2) (7).

While a lesionectomy is able to most often accomplish the goal of reduction or the complete cessation of seizures and also decreasing the risk possible future malignant conversion of the tumor in most patients; there are risks. These risks include visual field defects, cognitive defects, stroke (1-2%), and death (0.1%) (8). The risk of visual field defect is low at approximately 8-10 percent (9).

Charles Bonnet Syndrome is complex visual hallucination that is most often reported in elderly patients with blindness or severe visual impairment suffering from degenerative eye conditions. There have been a handful of cases described where visual images occur in a younger patient that has experienced a visual field defect as a consequence of their lesionectomy that involve the optic tract (10).

Charles Bonnet Syndrome has essential features that must be present for diagnosis including elaborate, vivid images, insight that these images are not real, and absence of psychiatric disorders (11). All of these criteria were met by the patient in the case study. There have also been arguments that visual hallucinations in these post surgical epilepsy patients are epileptogenic in nature, however, the description of this visual phenomena differs (12). In contrast, visual perceptions with a seizure are described as brief, disconnected, simple (such as flashing lights, stars, a blinking square), and associated with other symptoms such as vocalizations, vegetative or motor phenomena, and loss of consciousness (13). The patient in this case study had experienced visual aura (a simple flashing white light) prior to her seizures, but described her current visual hallucinations as much more vivid and colorful.

IV. Treatment and Management

As in the treatment of Charles Bonnet Syndrome in patients who are more elderly with advanced eye conditions; therapeutic options are limited. Reassurance to the patient that these visual hallucinations have been reported in those that have undergone surgical brain resection and with visual field defects and that it is not a sign of mental illness is critical. The use of antipsychotic drugs remains debatable. Most cases of Charles Bonnet Syndrome will resolve spontaneously without intervention (12).
V. Conclusions

This case shows the importance that eye care practitioners understand that Charles Bonnet Syndrome can occur in a patient population other than the elderly and those with advanced eye condition. In this case, the patient’s visual acuity was normal; however because of her visual field defect and lesionectomy, she experienced visual hallucinations consistent with Charles Bonnet. The patient was relieved about her diagnosis and that her visual hallucinations were not a sign of impending mental illness. She remains seizure free and is able to function well despite her visual field defect.

Bibliography


Abstract: A 35-year-old Hispanic female presents with complaints of photopsias. Examination reveals a retinochoriditis and foveal granularity in the right eye (OD).

I. Case History

- 35 year old Hispanic female reported to the urgent care service with a chief complaint of flickering lights in her right eye that started two weeks ago. Her symptoms were improving. The patient did not notice any curtain vision or floaters. Patient also mentioned stationary spots in her central vision.
- Primary ocular history: Patient wore soft contact lenses for myopia
- Family ocular history: unremarkable
- Primary medical history: had flu like symptoms 2 weeks ago that resolved without treatment
- Medications and Allergies: unremarkable

II. Pertinent findings

Clinical Testing:

- Best corrected distance acuity through a -3.50 sphere prescription OU was 20/20- OD, 20/20 OS.
- Amsler grid testing showed paracentral scotomas OD. Amsler grid OS showed no metamorphopsia or scotomas. Pupil testing was equally round and reactive to light with no afferent pupillary defect.
- Confrontation fields showed paracentral scotomas OD with full to finger counting OS.
- Extraocular muscle testing showed a full range of motion OU.
- Slit lamp examination was unremarkable OU
- Goldmann applanation tonometry revealed intra-ocular pressures of 10mmHg OU.
- Dilated fundus examination showed a clear vitreous OU; the optic nerve was flat, sharp, and showed good color OU; the cup to disc ratio was estimated at 0.15/0.15 OU; the macula showed a granular orange yellow appearance OD, but was flat with no hemorrhages or exudates; the OD macula also showed areas of brown granularity temporally, at the level of the RPE; the OS macula was flat, with no hemorrhages,
exudates or pigmentary changes; the retinal vessels were normal with an arterial-venous ratio of 2/3 OU

- Peripheral retinal examination in the right eye showed numerous round white spots in the deep retina that started in the mid-periphery and extended to the equator in the superior quadrants; the retina was flat, with no holes or breaks OU.

**Other testing:**

- Spectral domain optical coherence tomography (SD-OCT) 5-line raster scan of the macula showed disruption of the photoreceptor inner segment-outer segment junction subfoveally as well retinal pigment epithelium disruption OD; the 5 line raster scan OS was unremarkable
- Fluorescein angiography of the right eye revealed no early staining of any lesions OD or OS; late findings included a mild staining of the optic disc in the right eye; all findings in the OS were unremarkable.
- Fundus auto fluorescence OD revealed small hyper auto fluorescent areas temporal to the fovea.

**III. Differential diagnosis**

Differential diagnoses considered include:

1. Acute Posterior Multifocal Placoid Pigment Epitheliopathy (AMPPE)
2. Birdshot Retinochoroidopathy
3. Multifocal Choroiditis (MFC)
4. Multiple Evanescent White Dot Syndrome (MEWDS)

1. AMPEE
   a. Appears in young adults usually after a viral illness.
   b. Occurs equally in men and women, presents bilaterally.
   c. Signs and symptoms include blurred vision, scotomas, and photopsias.
   d. There may be a mild vitreous reaction.
   e. Retinal lesions are creamy yellow in nature.

2. Birdshot
   a. Presents in middle age adults.
   b. Occurs more in men compared to women and presents bilaterally.
   c. Signs and symptoms include blurred vision, floaters, and photopsias.
   d. Moderate vitreous reaction.
   e. The retinal lesions present as multiple ill-defined cream colored spots scattered throughout the posterior pole.
3. Multifocal choroiditis
   a. Occurs in young myopic adults.
   b. Occurs more frequently in women than men and presents bilaterally.
   c. Signs and symptoms include blurred vision, floaters, scotomas, and photopsias.
   d. Moderate vitreous reaction.
   e. Multiple small, round, yellow gray lesions similar to histo spots are seen in the macular area.

4. MEWDS
   a. Occurs in young adults, more so in females.
   b. Patients also show a degree of myopia.
   c. Condition is unilateral. Signs and symptoms include blurred vision, scotomas, and photopsias.
   d. There may a mild vitreous reaction.
   e. Multiple small white dots are seen with an orange granularity of the fovea.

IV. Diagnosis and discussion

• MEWDS is rare disease classified under the white dot syndromes. First described by Jampol et al\textsuperscript{1} in 1984 as a rare, sudden onset, unilateral retinitis with the predominant finding being white dots in the retina.
• Predilection for young myopic women more so than men.\textsuperscript{1-5}
• Etiologies include a link to the hepatitis A and B virus, human papilloma virus HPV\textsuperscript{6-8}
• Condition has been linked to the HLA-B51 gene as some patients may test positive for the gene, however, many patients do not possess this gene at all\textsuperscript{9}.
• Viral cause has also been associated as 33-50% of patients present after a recent case of flu like symptoms; however, no specific virus has ever been isolated\textsuperscript{1-7}.
• Retinal findings include flat, gray white lesions at the level of the RPE\textsuperscript{3-5}. There are usually no visible signs of the dots after they resolve, but they may leave a granular appearance in some cases\textsuperscript{10}.
• Granular appearance was evident in the patient, as areas of granularity were seen temporal to the macula.
• One finding specific to MEWDS is an orange yellow granularity of the fovea. This finding is thought to explain the blur and scotomas that patients notice. Rare findings associated with MEWDS include a retinal vasculitis, vitritis, and disc edema\textsuperscript{1-9}.
• Fluorescein angiography, SD-OCT, FAF, and ERG studies have shown that MEWDS can be isolated to the RPE and photoreceptors.\textsuperscript{1,2,11-13}
• Uniqueness of condition:
  o Rare and difficult to diagnose due to similarity to other white dot syndromes
Early examination/timing is essential in diagnosis as condition resolves without treatment.
Symptoms of photopsias are usually correlated to vitreo-retinal conditions not white dot syndromes.

V. Treatment, management

- MEWDS is a self-resolving condition and does not require any treatment, only observation.\(^{14}\)
- Majority of cases will regain their acuity in three to ten weeks.
- The white dots will resolve but may leave a brown granular appearance.
- Photopsias and scotomas resolve with time.
- Steroids have been attempted in hopes of improving signs and symptoms. Researchers used pulse oral steroids in one patient who showed improved findings within three days.
  - Researchers in the study however cautioned this approach as side effects of large amounts of pulse steroids include damage to the circulatory system.
  - Oral steroids have also been attempted, however it took several weeks for patients to improve therefore it was determined that this treatment would not be considered effective when compared to the natural course of MEWDS \(^{15}\).

VI. Conclusion

- Diagnosis of MEWDS can be challenging as many of the white dot syndromes present in a very similar manner.
- With a comprehensive history, exam findings, and ancillary testing, the white dot syndromes can be differentiated from each other.
- Clinicians should thoroughly educate their patients about the natural course of the disease as the symptoms can be very frustrating to patients.
- Symptoms of photopsias are similar to posterior vitreous detachments and retinal detachments. Clinicians should also be aware to include MEWDS in their differential diagnosis when a patient presents with symptoms of photopsia.
References


