Oculopalatal myoclonus following a cerebrovascular accident

A sixty five year old African American male develops oculopalatal myoclonus following a cerebrovascular accident. Oculopalatal myoclonus involves a lesion in the Guillain–Mollaret triangle.

I. Case History

- Mr. W presented as a 65 year old African American male
- Chief complaint:

  Binocular double vision that had started since a cerebrovascular accident (CVA) three months earlier.

- No history of eye surgery or trauma and his last eye exam in 2012 was unremarkable
- Medical history
  - hypertension,
  - alcohol dependency with alchololic hepatitis in 11/2011
  - renal insufficiency
  - urinary tract infection in 11/2011
  - sporadic cannabis use
  - vitamin D deficiency
  - CVA(09/2013) resulting in continued left sided weakness, slurred speech, mild dysphagia and diplopia
- Medications
  - Atenolol 100mg, Amlodipine 10mg daily, Bactrim DS bid and Vitamin D

II. Pertinent findings

- Clinical findings at initial presentation for ‘diplopia’
  - VA with habitual glasses
    - OD 20/40+ PH: UNABLE
    - OS 20/50- PH: UNABLE
      - HABITUAL OD: +0.75 +0.50 x 140
      - OS: +1.00 sph +2.00 ADD
  - Pupils equal round and reactive to light
  - Extraocular muscles had full range of motion.
  - Bilateral vertical nystagmus OS>OD
  - Question of mild constriction inferioely, checked with confrontation fields OU
  - Negative Bruit
  - Unable to perform accurate refraction secondary to “letters jumping around” per patient
  - Slit lamp of the anterior chamber was unremarkable other than a narrow anterior chamber
  - Gonioscopy showed an open angle 360 degrees with mild-moderate pigment throughout the angle
  - Amsler grid was negative in both eyes
  - Red Cap 75% OD, and 100% OS
  - Color Testing was normal with Ishihara OU
  - Goldman applanation
    - OD 13
    - OS 14 @ 1600
  - Dilated examination was unremarkable other than:
    - 1-2 + NS with mild anterior cortical changes OU
- Large area of soft juxta-foveal drusen without watzkie sign OS
- Arteriolar attenuation with moderate venous engorgement OU
  - Unable to perform color photos or OCT of the macula because Mr. W was unable to fixate
- An MRI of the brain was ordered but due to physical concerns Mr. W required an open MRI.
  - Impression
    - “No gross acute infarcts seen, although this is low limited technically images. No focal abnormal enhancement noted…diffusion weighted imaging not available for evaluation”.

- 7 month follow up exam
- Mr. W presented still complaining of “diplopia,” reading issues along with decreased peripheral vision, dizziness and falling. Further questioning revealed that the horizontal diplopia did not go away when covering an eye and the patient was actually experiencing an “image jump”.
  - VA with habitual
    - OD 20/30+ PH 20/25
    - OS 20/30 PH Unable
    - HABITUAL OD: +0.75 +0.50 x 140 OS: +1.00 sph with a +2.00 ADD.
  - Pupils equal round and reactive to light
  - Extraocular muscles had full range of motion.
  - Bilateral alternating vertical nystagmus with torsional component OS>>OD
  - Confrontation fields were found to be full to finger count, no inferior constriction noted at this visit
  - Cover testing without correction
    - 5EP 4EP 4LH
    - 6EP 4EP 4LH
    - 5EP Ortho 5LH
  - Evaluation of the palate showed vertical pendular motion

III. Differential Diagnosis

- PRIMARY: Oculopalatal myoclonus
- Whipple's disease
- Labyrinthitis
- Cerebellar ataxia second to ischemia

IV. Diagnosis and discussion

Sea saw nystagmus resulting in oscillopsia with a pendular palate movement is clinically significant for oculopalatal myoclonus and was confirmed by a neuro-ophthalmologist evaluation. Oculopalatal myoclonus is specific to a lesion in the Guillain–Mollaret triangle, which involves a pathway from deep cerebellar nuclei to the contralateral red nuclear, to the inferior olive and back to cerebellum. It is not uncommon for MRI’s of patients with oculopalatal myoclonus to reveal no infarcts.

V. Treatment, management

Our recommendation was to patch the left eye because the left eye experienced greater image jump. Mr. W will return after 4 months time and will consider 2-4 BO prims to reduce image jump. Other treatment options include surgery, and pharmacological intervention. Case reports have suggested that surgical intervention in conjunction with botulism injections have helped reduce oscillopsia with limited success. There is limited evidence to support pharmacological treatment will reduce an acquired pendular nystagmus although there is the potential to dampen the nystagmus with gabapentin or memantine.
VI. Conclusion

It is important for optometrists to look at the palate in patients with pendular nystagmus to rule out oculopalatal myoclonus.

Comments to Reviewers

Supplemental video of nystagmus and palatal tremor are available for presentation. Further literature review is also available for presentation.

Bibliography