I. Case History

- Patient demographics: 65 year old white female
- Chief complaint:
  - Difficulty reading and looking upwards with associated dizziness; in order to look superiorly patient compensates by performing an upwards head thrust lifting her chin to move eyes upwards.
- Ocular history:
  - Hypermetropia and Presbyopia OU
  - Wears separate single vision distance and near spectacles
- Medical history:
  - Aortic valve replacement 20 years ago with a subsequent stroke and bleeding in her frontal lobes
  - Dysarthria following stroke
- Other salient information:
  - Longstanding complaint yet patient has never sought care

II. Pertinent findings

- Clinical:
  - VA:
    - OD 20/20, OS 20/20
  - Distance Phoria:
    - Horizontal: orthophoria
    - Vertical isophoria
  - Near Phoria:
    - Horizontal: 14 prism diopters exophoria
    - Vertical: isophoria
  - Pursuits:
    - Grade +1 (Southern California Grading System)
  - Saccades:
    - Volitional and reflex absent in all directions
  - Near point of convergence:
    - 12”/16” + diplopia
  - Vergence ranges:
    - Near Base In: x/8/6
    - Near Base Out: x/6/4
    - Distance Base In: x/4/2
    - Distance Base Out: x/6/2
  - Stereopsis:
    - Random Dot Stereogram: 250 seconds of arc
    - Wirt Circles: 25 seconds of arc
  - Optokinetic Nystagmus:
    - Absent reflex saccade in both directions
- Radiology studies:
  - History of MRI

III. Differential diagnosis
Primary/Leading: Selective saccadic palsy
• Acquired ocular motor apraxia
• Progressive supranuclear palsy
• Congenital ocular motor apraxia

IV. Diagnosis and discussion
• Selective saccadic palsy is characterized by impaired volitional and reflex saccades. The patient’s smooth pursuit ability, vergence, and vestibulo-ocular eye movements are preserved.
  o Horizontal and vertical saccadic palsy is a common finding in patients with neurological deficits following cardiac surgery. (Kim, E., et al., 2014).
  o If saccadic function exists, it tends to be hypometric and slowed; vertical saccades are usually more impaired than horizontal saccades. (Solomon, D., et al., 2008)
• Imaging studies for diagnosis:
  o Many published cases on selective saccadic palsy following cardiac surgery report no lesions or abnormalities of the brainstem on magnetic resonance imaging (Eggers, S. et al., 2015).
• Proposed causes of selective saccadic palsy following cardiac surgery:
  o The pathophysiology remains unclear.
  o Previous studies hypothesized selective saccadic palsy results from either damage along the pathway from the frontal eye fields to the brainstem or damage to the brainstem, specifically the excitatory burst neurons or omnipause neurons. (Yee, R., Purvin V., 2007).
  o Damage affecting the cerebral hemispheres or superior colliculus could also cause this condition.
  o In comparison, recent studies have postulated the role of perineuronal nets contributing to absence of horizontal and vertical saccades, rather than direct damage to premotor excitatory burst neurons or omnipause neurons (Eggers, S. et al., 2015)
  o Potential contributing factors during cardiac surgery: ischemia, hypotension, intraoperative hypothermia, hyperviscosity, and microembolism (Yee, R., Purvin V., 2007).
• Associated symptoms to selective saccadic palsy after aortic surgery:
  o Dysarthria, dysphagia, and gait disturbance (Kim, E., et al., 2014).

V. Treatment and management
• There is no standard of care for the treatment of selective saccadic palsy at this time.
• Patients try to adapt by blinking to shift their gaze or performing head thrusts.
• Recommend vision therapy to improve vergence ranges and accuracy of pursuit eye movements; improve head thrusts to re-fixate targets
• A case report proposed rehabilitation technique: opaque glasses with vertical slits
VI. Conclusion

Selective saccadic palsy is a relatively unrecognized visual disability as the patients present with co-morbidities such as dysarthria. The pathogenesis still remains unknown, however one recent post-mortem immunoassay study identified damage and fragmentation to perineuronal nets surrounding excitatory burst neurons and omnipause neurons as a likely cause. (Eggers, S. et al., 2015). Unfortunately for most patients, improvement in saccadic function has only been found during the acute time period following the cardiac surgery. (Solomon, D., et al., 2008)

References


