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Double Trouble: Diplopia and Oscillopsia resulting from Oculopalatal Myoclonus Syndrome

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Abstract

Exploring treatment and management options for a 30-year-old white male symptomatic for diplopia and oscillopsia after suffering from a pontine stroke.

Case History

Chief concern: Follow up with monovision glasses for treatment of diplopia and oscillopsia after suffering from a pontine hemorrhagic CVA causing Oculopalatal Myoclonus Syndrome (OMS). Patient reported that monovision glasses were not effective because he could still see a blurry image moving up and down, and that he experienced more relief with black eye patch.

Personal ocular history:

- (+) Oculopalatal Myoclonus without palate involvement
- (+) Pendular Vertical Dissociated Nystagmus
- (+) Noncomitant strabismus: XT in lateral gazes vs. ET in all other gazes
- (+) Right and left lateral gaze restrictions

Personal medical history:

- Hemorrhagic Pontine Cerebral Vascular Accident 06/2015
- Hypertension
- Medications: Acetaminophen, Amlodipine, **Baclofen 10 mg**, **Gabapentin 900 mg TID**, Hydrochlorothiazide, Lisinopril, Metoprolol.

Clinical Findings

Corrected VA:

- OD (near eye) : 20/160 with Lighthouse Continuous Text card
- OS (distance eye): 20/40+2 with Snellen acuity chart

Distance Only Spectacle Rx:

- OD: -8.00 +2.25 x 095
- OS: -8.00 +2.25 x 085

Monovision Spectacle Rx:

- OD: -5.00 +2.25 x 095 (near eye)
- OS: -8.00 +2.75 x 085 (distance eye)

Cover test with distance correction for OD and OS:

- Distance: roughly 15 PD RE(T) in primary, up and down gazes
- 10 PD XT in right and left gaze.

Pupils: PERRL, (-)APD

EOM: Right and left lateral gaze restriction

OD			OS		
-3	0	-1	-1	0	-3
-3		-1	-1		-3
-2	0	-1	-1	0	-3

Dissociated vertical nystagmus: ocular bob/oscillation in primary gaze, OD: ellipsoid pendular with more horizontal movement, OS: ellipsoid pendular with more vertical movement

Diagnoses

Diplopia, Acquired Dissociated Nystagmus, Lateral Gaze Palsy

Discussion

Pontine CVA:

- Typically caused by a blood clot or hemorrhage in the Pons. Although this may occur secondary to trauma from sudden neck or head movements.
- The Pons is a region of the brainstem which lies between the Midbrain and the Medulla.
- The Pons is responsible for relaying messages between the cerebral hemispheres and the cerebellum.
- Symptoms of a brainstem stroke include: vertigo, dizziness, severe imbalance, double vision, slurred speech or decreased consciousness. Its important to note that most brainstem strokes do not present with the hallmark sign of other strokes: weakness of one side.
- Risk factors include: hypertension, diabetes, cardiovascular disease, and smoking.

Oculopalatal Myoclonus Syndrome (OMS):

- Caused from lesion of the inferior olivary nucleus or its connections.
 - Specifically, it is due to the instability of the projections from the inferior olive to the cerebellar flocculus. This process is thought to be important in the adaptive control of the vestibular ocular reflex (VOR).
 - The hypoglossal nucleus is next to the inferior olivary nucleus which is why the palate becomes involved.
- Develops weeks to months after a brainstem or cerebellar infarction
- Myoclonus is a misleading term because by definition myoclonus is twitching, jerks, or seizures caused by muscle contraction. In OMS the movement of the muscles are typically to-and-fro and synchronized.
- Presentation may vary depending on type of nystagmus and involvement of palate:
 - In a majority of cases the patient will present with predominately vertical pendular oscillations and bilateral palatal myoclonus.
 - If the nystagmus appears to have mixed vertical-torsional movements (sometimes disconjugate) the patient will typically present with unilateral or asymmetrical palatal myoclonus.
 - In some cases the patient will present with eye oscillations with out involvement of palate.
- The main pathological finding of concern is hypertrophy of the inferior olivary nucleus.
 - The inferior olivary nucleus is involved in motor function and directly communicates with the cerebellum.
 - It is mostly involved in the timing and learning of movement and is therefore thought to be involved with blink rate and the vestibular system.
 - Diagnosis of hypertrophy of the inferior olivary nucleus can be made with magnetic resonance imaging.

Treatments for Diplopia:

- Prism: Typically works best for patients with comitant deviations. In this case, the patient tried 15 PD BO Fresnel prism to relieve diplopia in primary gaze. However, while the prism moved the images closer together, the oscillopsia prevented stable fusion.
- Monovision: Works well for patients with large deviations in which prism is not a great optical option. In this case the movement of the blurry image bothered the patient and was more difficult for him to suppress.
- Central Stipple Occlusion: Uses quarter sized clear contact paper over non-dominant eye to blur second image. Over time the clear contact paper can be reduced in size. This allows the patient to still have peripheral awareness, does not create sticky film on glasses, and is cosmetically more acceptable than black pirate patch.

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Discussion Continued

Treatments for Nystagmus:

- Pharmacological Treatment:
 - Although the neuropharmacology of acquired pendular nystagmus is unknown, studies have shown increased acetylcholine esterase activity and cholinergic denervation super sensitivity in patients with OMS and hypertrophy of the inferior olivary nucleus.
 - Trials of anticholinergic agents have been performed including:
 - Barbituates: sedative effects limit use
 - Trihexyphenidyl: only a few patients responded well and the side effects are not well tolerated
 - IV Scopolamine: not practical
 - **Studies showed that GABAergic agents are important in the normal function of the gaze holding mechanism and patients who received a single 600 mg dose of Gabapentin had improved VA, reduction in nystagmus and increased foveation, and it was well tolerated.**
- Optical Treatment:
 - Prism: BO Prism has been effective for those who have less nystagmus when viewing a near target. BI prism works better for those with less nystagmus when viewing distant targets. Prism can also be used to help a patient maintain fixation at null point.
 - High plus glasses and high minus contact lens
 - Try to stabilize image on the retina rather than stabilize eye movement.
 - The spec lens is focusing image at the center of rotation of the eye, when this happens that image is defocused so the minus lens extends the clear image back onto the retina can achieve 90% stabilization
 - Challenges: it disables all eye movements including the VOR and vergence movements so only beneficial if patient is stabilized or monocular, also with the highest power components: CL -58,00 D and Spec +32.00 D get limited field of view
- Other Options:
 - Botulinum Toxin
 - Challenges: Major side effects are ptosis and diplopia. Also there is risk with repeated injection
 - Surgery
 - Typically done for congenital nystagmus
 - Biofeedback

Management

- Patient will continue to seek care with his neuro-ophthalmologist for management of Gabapentin and Baclofen. Maximum dosage of Gabapentin for this patient is 3600 mg.
- In regard to patient's eyeglasses and diplopia, due to previous poor response to prism and monovision, patient was educated about central stipple clear contact paper occlusion.
 - Patient elects to try central stipple occlusion. A high minus CL and high plus spectacle pair of glasses may be an option for this patient in the future, it was not explored at this time due to small change of improving VA past 20/40.

Conclusions

- As the aging population grows treatment and management of diplopia and oscillopsia after stroke will become increasingly important.
- Prism may not be the best option for alleviating diplopia in these patients and other options including: monovision and central stipple occlusion must be considered.

Sources

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