Supranuclear ocular motility disorders

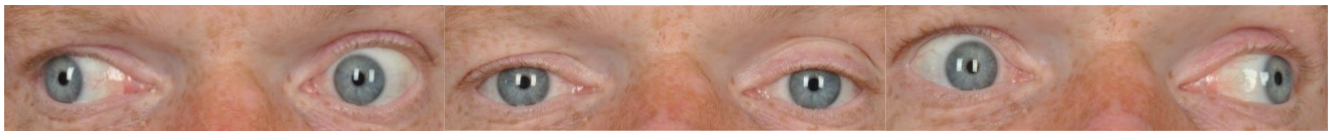

Figure 1: Bilateral INO

Introduction

Complex ocular motility disorders are a diagnostic challenge. These patients come with very complex ocular motility presentations and require a careful and detailed assessment in order to find the correct diagnosis and arrange appropriate investigations.

Supranuclear disorders refer to abnormalities in ocular motility caused by lesions higher than the cranial nerves nuclei. They are usually characterised by preservation of reflex movements such as Bell's phenomena, the vestibular ocular reflex (VOR) and the caloric nystagmus.

Types of ocular motility

1. Saccadic system: fast conjugate movements to refixate on targets
2. Pursuit system: slow conjugate tracking movements to keep target on fovea
3. Vergence system: keeps fixation when target is moving away or towards the observer
4. Vestibular ocular system: removes the effect of head movements on fixation by producing eye movements that are equal and opposite to head movements.

The ocular motility pathways

Conjugate ocular movements are managed in the brain at two levels, higher control and lower control areas. The higher area is responsible for calculation and planning of the eye movement while the lower area is responsible for generating the movement.

1. Saccadic eye movement:

a. Higher control areas are the frontal eye field (FEF), supplementary eye fields (SEF) in the frontal lobe, parietal eye field (PEF) in the parietal lobe and the superior colliculus (SC) in the midbrain. Other important areas include thalamus and basal ganglia.
b. Lower control areas are pontine paramedian reticular formation (PPRF) for horizontal movements and rostral interstitial nucleus of the medial longitudinal fasciculus (riMLF) and interstitial nucleus of Cajal (INC).
c. Sequence of events to generate a voluntary saccade: higher control centres (FEF, SC) send an inhibitory signal to the omnipause neurons in the contralateral PPRF which frees the excitatory burst neurons to stimulate ipsilateral VI cranial nerve nucleus. For the vertical movements, bilateral FEF and SC stimulation is required to riMLF and INC then to the III and IV cranial nerves nuclei.
d. Stimulation of FEF and SC will generate a contralateral conjugate saccadic movement.

2. Pursuit eye movements:

a. Higher control areas are middle temporal area (MT) and middle superior temporal area (MST) as well as FEF.
b. Lower control areas are dorsolateral pontine nuclei (DLPN), cerebellum and vestibular nuclei.

3. Vergence eye movements:

a. Higher control areas are poorly understood.
b. Lower control areas are possibly in the mesencephalic reticular formation.
c. Examples of such disorders include:
   i. Convergence insufficiency:
   ii. Parkinson's disease (PD), progressive supranuclear palsy (PSP) and internuclear ophthalmoplegia (INO)
   iii. Convergency paralysis: Dorsal midbrain syndrome and PSP
   iv. Convergence spasm: usually non organic but could happen with dorsal midbrain syndrome and thalamic infarction
   v. Divergence paralysis: usually pons lesions.

4. Vestibular ocular reflex:

a. Peripheral vestibular apparatus: the three semi-circular canals, the saccule and the utricle.
b. Central vestibular apparatus: the VIII cranial nerve and the brainstem vestibular nuclei.
c. Cerebellar centres.
d. Abnormalities can lead to nystagmus:
   i. A lesion in the vestibular apparatus will lead to a nystagmus with a quick phase away from the lesion
   ii. A lesion in the cerebellar centres will lead to a nystagmus with the quick phase towards the lesion.

Examples of supranuclear disorders

1. Saccadic palsy:

a. This usually happens following damage or hyperactivity in the FEF area in the frontal lobe.
b. The damaging lesion causes a unilateral saccadic palsy to the contralateral side.
c. If bilateral, the patient would have a spasm of fixation and need to use head thrust to refixate.
d. The hyperactivity (frontal adversive seizures) causes a deviation of the eyes to the contralateral side.

2. Pursuit palsy:

a. This happens following damage or hyperactivity in the parieto-occipital lobe.
b. The damaging lesion causes a unilateral pursuit palsy to the ipsilateral direction.
c. The hyperactivity will cause a deviation of the eyes to the ipsilateral side usually associated with visual hallucinations.

3. Unilateral horizontal gaze palsy:

a. A pons lesion affecting the PPRF or the VI cranial nerve nucleus.
b. The lesion causes a unilateral gaze palsy to the ipsilateral direction.

4. One and a half syndrome:

a. A pons lesion affecting the PPFR and the ipsilateral MLF.
b. The lesion causes a unilateral gaze palsy to the ipsilateral direction and adduction deficient to the contralateral direction.

5. Bilateral horizontal gaze palsy:

a. A pons lesions affecting bilateral PPRF or VI nerve nuclei.

6. Dorsal midbrain syndrome:

a. A lesion in the dorsal midbrain such as a tumour in the pineal gland.
b. Saccadic paralysis to elevation but normal pursuit at the early stages but can progress to loss of both movements and loss of downwards movement as well.
c. Convergence retraction nystagmus: convergence and retraction of the globes on attempted elevation.
d. Light near dissociation in pupil responses.

7. Progressive supranuclear palsy (PSP):

a. Vertical saccadic palsy downwards in early stages.
b. Complete vertical saccadic paralysis.
c. Possible complete ophthalmoplegia in late stages.
d. Apraxia of the lid opening (inability to open eyelids).

8. **Parkinson's disease (PD):**
   a. Vertical saccadic palsy upwards but not downwards.
   b. Convergence insufficiency.
9. **Internuclear ophthalmoplegia (INO):**
   a. A lesion in the MLF.
   b. Could be bilateral (Figure 1).
   c. Adduction weakness with abducting nystagmus in the contralateral eye.
   d. Preserved convergence (usually).
   e. A possible skew deviation with the higher eye on the same side of the INO.
10. **Skew deviation:**
   a. Vertical supranuclear strabismus.
   b. Concomitant or incomitant.
   c. Intorsion of the hypertropic eye.
   d. Negative Bielschowsky head tilt test (not always).
   e. Change in the vertical deviation between supine and upright positions.
   f. Ocular title reaction (OTR): skew deviation, ocular torsion and head tilt.
11. **Double elevator palsy:**
   a. Limitation of elevation.
   b. Usually congenital.
12. **Doble depressor palsy:**
   a. Limitation of depression.
   b. Usually congenital.
13. **Vertical one and a half syndrome:**
   a. A lesion in the thalamic mesencephalic junction.
   b. Limitation of elevation bilaterally with limitation of depression in one eye.

Investigations

It is important to investigate these patients appropriately. All patients will need detailed history, medical and ophthalmic examinations as well as orthoptic assessment. Blood tests and neuroimaging is usually necessary in most cases.

Managements

Various options are possible depending on the patient's concern and complaint. Examples would be yoked deviating prisms or Kestenbaum procedure to correct head postures, prisms, or strabismus surgery to help with diplopia and medications or surgery to help with nystagmus.

Conclusion

Supranuclear ocular motility disorders are a diagnostic and management challenge. They need to be fully assessed and investigated prior to discussion about possible management options.

References:

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