Internuclear Ophthalmoplegia, a Stroke of Double Vision

Abstract:
A 73 year old male presents with a sudden onset of diplopia, with an adduction deficit OS and end gaze nystagmus OD. Signs and MRI lead to diagnosis of internuclear ophthalmoplegia.

I. Case History:
   - Patient Demographics
     73 Year old white male

   - Chief complaint
     Sudden onset of binocular double vision for less than one week, noticed more at a distance rather than near.

   - Ocular & Medical History
     Ocular History:
       Nuclear Sclerotic Cataract OU
       Presbyopia
     Medical History:
       COPD
       Hypercholesterolemia
       Hypertension
       Raynaud’s Disease
       Diverticulosis
       Heart Murmur
       Carcinoma of Prostate
       Gastroenteritis and colitis

II. Pertinent Findings
   - Clinical
     Initial Visit with new onset diplopia
     Pupils: PERRL(-) APD
     EOMs: Full/ Smooth without restrictions OU
Near Point of Convergence: To The Nose
Cover Test (distance): 3 exophoria and 6 left hyperphoria
Von Graefe Phoria:
  - Distance: 4 prism diopters BI, 6 prism diopters BD OS
  - Near: 6 prism diopters BI, 9 prism diopters BD OS
Red Lens Test (9 gazes): more hyper and exo deviation toward right gaze
Visual Field SITA Fast 30-2
  - OD: clear, no defects
  - OS: clear, no defects
Trial of 4BD OS over current glasses, patient reports single vision and good comfort after 30 minutes of trial
Palpebral Aperture:
  - OD: 8mm
  - OS: 10mm
SLE/Tonometry: Unremarkable OU
DFE: Unremarkable OU
Secondary Visit (five days later):
  - EOMs: endpoint nystagmus noted OD on right gaze, and mild adduction deficit OS. Full and smooth movements in all other gazes
  - Cover test @ distance with 4 BD Fresnel prism OS
    - Primary gaze: 5 exophoria
    - Right gaze: 20 exophoria
    - Left gaze: ortho
    - Upgaze: 4 exophoria
    - Downgaze: ortho

-Physical
Patient denies any amaurosis, weakness, pain or paresthesias of either side, dizziness, eye pain, trauma, nausea, unsteady gait, or slurred speech.

-Radiology studies
MRI with/without contrast with attention on brainstem and MLF was ordered. MRI showed deep white matter ischemic changes and evidence of pontine lesion on scan.

-Carotid Duplex
Bilateral irregular surface on the carotids, with mild occlusive disease bilaterally, right greater than left, both <70%

III. Differential diagnosis
  - Myasthenia Gravis, intermittent decompensation of an existing phoria, cranial nerve palsies, thyroid eye disease, cavernous sinus fissure syndrome, internuclear ophthalmoplegia, vertebrobasilar artery insufficiency, neoplastic involvement

IV. Diagnosis and discussion
Internuclear ophthalmoplegia (INO) is characterized by horizontal gaze impairment, with weak adduction of the affected eye on the side of the lesion and nystagmus on abduction of the contralateral eye. The lesion causing INO occurs along the medial longitudinal fasciculus (MLF) located in the brainstem by blocking the connection between contralateral cranial nerve six nucleus and the ipsilateral cranial nerve three nucleus. The MFL is responsible for conjugate horizontal eye movement, by connecting the 6th nucleus, adjacent horizontal gaze
center (paramedian pontine reticular formation) and the contralateral 3rd cranial nerve nucleus. A differential characteristic of INO versus cranial nerve palsy can be that convergence remains intact with INO. However, this is not always the case as convergence can be absent in a mesencephalic lesion (an anterior lesion) and tends to be intact in posterior lesions.

Internuclear ophthalmoplegia was first described in 1903. Studies have shown a diverse pathological history with demyelination and stroke being the most common. Demyelinating diseases such as multiple sclerosis, tends to cause INO in a younger adult or adolescent and is more often bilateral. Where as a stroke tends to be the culprit in older patients and are more often unilateral(1,2). Other cause of INO can be trauma, brainstem mass lesion, neurosyphilis, Lyme disease, drug intoxication (tricyclic antidepressants), hydrocephalus, nutritional, metabolic disorders, CNS cryptococcosis, tuberculosis granuloma, and pyoderma gangrenosum.

Ischemic lesions tend to affect the MLF in a unilateral manner because of the segmental distribution involvement of the paramedial branches of the basilar artery(1). A patient may appear to have an INO, but may in fact be pseudo-INO usually caused by myasthenia gravis (4).

Symptoms may range from vague complaints of blurry vision or double vision (3). Many patients will not report horizontal diplopia but in fact will report vertical diplopia. This can be due to an associated skew deviation and a difficulty tracking fast moving objects because of the mismatch in the velocity of saccadic eye movements. A skew deviation is an acquired vertical misalignment of the eyes due to an asymmetrical disruption of supranuclear input from the otolithic organs such as the utricle & saccule of the inner ear. Skew deviations are an important exception to the rule that supranuclear lesions do not produce double vision(7).

V. Treatment, management

Treatment depends on the underlying cause. If acute stroke is the diagnosis, the patient should be admitted to the hospital for neurological evaluation and observation. For other pathologies patients should be managed by a physician familiar with the underlying disease. MRI can be a valuable diagnostic tool in determining the underlying culprit, but a lesion will not always be detected. The long-term outcome of INO has not been well studied and most of the data available appears to be related to multiple sclerosis (1). Bolanos states that the resolution time of an INO depends on the etiology and MRI findings, not age, gender or type of INO. Most patients with an infectious etiology (66.6%), demyelinating (61.9%), and traumatic (60.0%) showed complete recovery. Patients with cerebrovascular disorders had a less favorable recovery; INO persisted in (62.5%). However, when the INO is an isolated predominant symptom of brainstem infarction the resolution of INO was excellent, (100%) (5). Kim found that when INO was an isolated brainstem infarct it disappeared in all patients usually within one month. Kim also found that the duration of the INO tended to be longer in patients that had other neurological signs such as paresthesia, mild dysarthria, gait ataxia, or facial palsy(5). This was true for this patient. Fresnel prisms were prescribed at the second visit to help the patient cope with vertical diplopia but were no longer needed by his three month follow up exam.
VI. Conclusion

Internuclear ophthalmoplegia is a condition that gives the precise location of a lesion. Given its possible causes, recognition and diagnosis is very important and can be life saving. As mentioned above, patients will not come in with straightforward complaints, and the clinical findings may be so slight they are easy to overlook or miss at times. It is important to note that in some cases ocular motility may appear to be full, but muscle weakness can be detected by observing slower saccadic eye movements in the involved eye compared with the contralateral eye.