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# Cogan's anterior internuclear ophthalmoplegia in young Ethiopian: A case report and review of literature

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**KEYWORDS**

Medial longitudinal fasciculus lesion;
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**Abstract** *Introduction:* Lesions in medial longitudinal fasciculus (MLF) produce internuclear ophthalmoplegia (INO) with characteristic horizontal gaze abnormality. Here, the author reports a rare clinical spectrum of internuclear ophthalmoplegia called Cogan's anterior internuclear ophthalmoplegia in young Ethiopian suspected to have multiple sclerosis. To the best of the author's knowledge, this is the first report in non-white population near the equator.
*Case presentation:* A 19-year-old female Ethiopian patient presented with bilateral internuclear ophthalmoplegia. Attempted convergence was abnormal. The patient regained normal ocular mobility after six months of follow-up.
*Conclusion:* Isolated reversible internuclear ophthalmoplegia in a previously healthy young patient could be an initial manifestation of a serious illness like multiple sclerosis. Though the incidence of multiple sclerosis in non-white population near the equator is low, high index of suspicion and close follow up are warranted.


## 1. Introduction

Supranuclear ocular motor disorders can be caused by brainstem, cerebellar or cerebral hemisphere lesions. Lesions in medial longitudinal fasciculus (MLF), a fiber tract that rises from the abducens nucleus in the pons to the contralateral oculomotor nucleus in the midbrain, produce internuclear ophthalmoplegia (INO) with characteristic horizontal gaze abnormality in the absence of third nerve palsy.[1–4]

The gaze abnormality could be a total or partial failure of adducting one eye in lateral gaze and is associated with a monocular abducting nystagmus in the contralateral eye.[1,4] Other associated findings in a patient with INO include vertical gaze-evoked nystagmus, impaired vestibular and pursuit vertical movement, impaired convergence, skew deviation, paralysis of horizontal gaze to one side, and involvement of other cranial nerves.[1–4]

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Any brain-stem disorder or lesion can interrupt the medial longitudinal fasciculus (MLF) within the dorsomedial pontine or midbrain tegmentum and result in impaired horizontal eye movement.[1,2] The most common causes of INO are multiple sclerosis (MS) and brainstem infarction. Other causes include head trauma, infection, brainstem and fourth ventricular tumors, hydrocephalus, Arnold-Chiari malformation and systemic lupus erythematosus.[1–5]

To the best of the author's knowledge, there are no or limited case reports on the rare clinical spectrum of internuclear ophthalmoplegia called Cogan's anterior internuclear ophthalmoplegia in Ethiopia and/or African region. This might be due to geographical variation in the incidence of MS disease, one of the main causes of INO, which seems higher as the distance from the equator in both hemispheres is greater and with less incidence in non-white population compared to white people.[6] This paper, thus, reports a case of transient bilateral INO with loss of convergence in the young black female Ethiopian patient and discusses its epidemiological variations.

## 2. Case presentation

A 19-year-old female Ethiopian presented with one week history of abnormal bilateral horizontal eye movements associated with double vision, staggering gait and occasional vertigo. She noted transient sudden worsening of vision during exercise at school and discontinued her education because of these disabling symptoms. She also reported an on-and-off headache and burning sensation on the lower extremities for about a year. The patient was healthy prior to this problem and she did not have any known medical illness/prodromal illness, vaccination, history of head trauma or drug intake. The patient did not have any previous episodes of transient neurological symptoms and no family history of known neurological diseases. On examination, her uncorrected visual acuity was 6/9 in each eye. Pupils were isocoric and normally reactive with no relative afferent pupillary defect. Intraocular pressure, color vision, and confrontation visual field of both eyes were normal. In primary position, the patient had an exotropia in the left eye (Fig. 1), which increased on both right and left horizontal gaze but not on upward or downward gaze.

Ophthalmic examination revealed horizontal diplopia during lateral gaze in both eyes. The patient had an adduction deficit in the right eye and ataxic abducting nystagmus in the left eye on left gaze (Fig. 2A). She also had an adduction deficit in the left eye and ataxic abducting nystagmus in the right eye on right gaze (Fig. 2B). The patient had a vertical gaze-evoked nystagmus and impaired vertical pursuit. The patient was also unable to converge. Ocular motility on up gaze, down gaze, and at primary position was unremarkable. The anterior and posterior segment examinations were normal in both eyes.

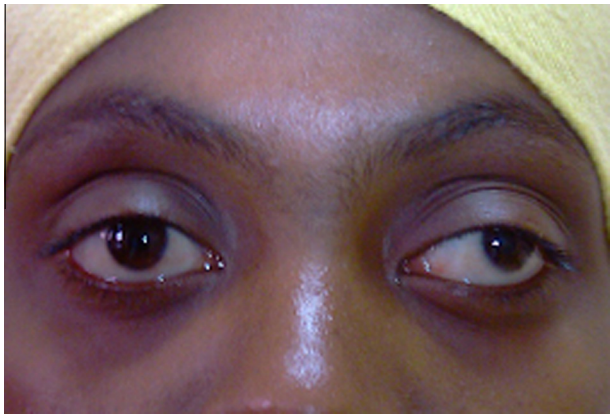

**Figure 1** Extropia of the left eye in the primary position.

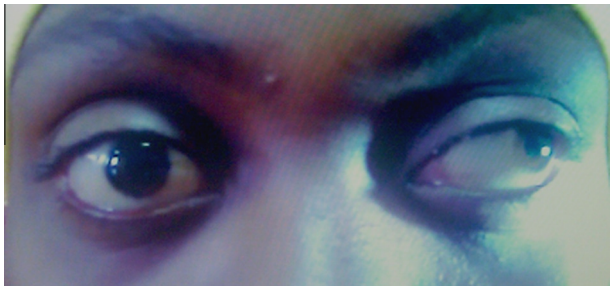

**Figure 2A** Adduction deficit in the right eye on left gaze.

**Figure 2B** Adduction deficit in the left eye on right gaze.

Neurological motor examination revealed up-going plantar reflex in the right lower leg and deep tendon reflexes were grade 2. Other neurological exams (sensory and coordination) were normal.

Serology test for syphilis and HIV was negative. Skull X-rays and ultrasonography of both eyes were also normal. The diagnosis of transient Cogan's anterior internuclear ophthalmoplegia (INO) was made and multiple sclerosis (MS) was suspected; and the diagnosis and other relevant information were communicated to the patient. The patient was referred for magnetic resonance imaging (MRI) but could not afford. On follow-up at 2 months, the patient's ocular movement and gait showed improvement and her vision acuity was 6/6 in both eyes. At six months, adduction deficit and nystagmus had improved (Fig. 3A and 3B) but there was still mild residual convergence insufficiency and gaze-holding nystagmus. The patient was given a short course of oral steroid therapy and is having follow up at the neurology and ophthalmic clinic.

## 3. Discussion

Studies estimate that around 75% of people with multiple sclerosis (MS) experience double vision in the course of the disease and this double vision can result from internuclear ophthalmoplegia, sixth nerve palsy or combinations of other cranial nerve

**Figure 3A** Improved adduction deficit in the right eye on left gaze.

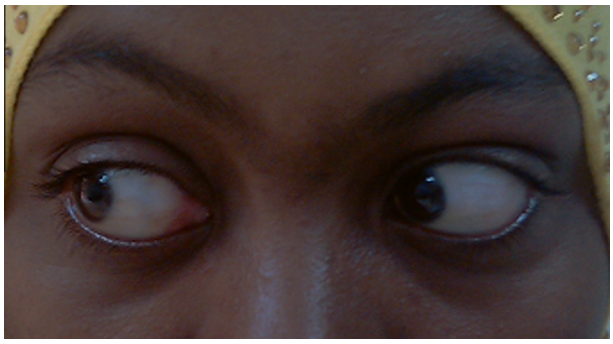

**Figure 3B** Improved adduction deficit in the left eye on right gaze.

palsies.[7] Internuclear ophthalmoplegia (INO) is seen in about 22% of MS patients[9] and can be the first neurological finding.

INO could be unilateral or bilateral; partial or complete; anterior or posterior depending on the age of the patient, the anatomic site and the cause of MLF lesions. When patients with INO are able to converge, despite the absence of voluntary adduction, a caudal lesion with the preservation of the medial rectus subnuclei of the oculomotor nuclear complex can be assumed, and this, a more common and recognized phenomenon, is called Cogan's posterior INO. This patient was, however, unable to converge and this is indicative of a mesencephalic lesion, which results in Cogan's anterior INO, as convergence is mediated by nuclei in midbrain.[1,2,9] Besides bilateral INO and absent convergence, the patient had exotropia in the primary position and hence the diagnosis of reversible wall-eyed bilateral INO (WEBINO) syndrome was also made.

The young age of the patient and bilateral involvement in the present case were consistent with the clinical diagnosis of multiple sclerosis. Isolated bilateral INO in young patients is often considered as a clinical pathognomonic sign of MS.[10,11] This was the case in this 19-year-old Ethiopian patient as MS is the most likely cause of a bilateral internuclear ophthalmoplegia in a young adult while brainstem infarction is the commonest cause of a unilateral internuclear ophthalmoplegia in middle aged and elderly adults.[1,2,8]

Keane JR described INO as 'arguably the most discrete localizing sign in medicine' because of its crucial clinical value in predicting MS or stroke, based on the laterality of INO (bilateral or unilateral) and the age of the patient (young or old).[12] Bolaños et al. analyzed bilateral INO cases according to age and etiology and found that eight of nine patients with multiple sclerosis were younger than 45 years, in comparison with one of six cases of vascular cause.[13] The Uhthoff phenomenon reported by the patient is also supportive of the diagnosis of MS in the present case. Diagnostic imaging was not available in the study setting and hence the patient did not have brain scan. Although magnetic resonance imaging (MRI), the method of choice for diagnostic imaging of MLF lesions, frequently shows a lesion in the MLF in patients with INO, there are many exceptions[1,4] and is not always possible to detect the responsible lesions.[14]

The gender (female sex) of the patient is also another supportive risk factor for the consideration of MS in the present case as MS affects more women than men.[6,8]

Bilateral INO can occur as an isolated clinical entity or with other ocular motor and visual sensory manifestations of MS like optic neuritis, nystagmus, and ocular motor nerve palsies.[1,7,10] Other neurological and psychological features may precede, occur concurrently with, or follow the development of bilateral INO and disturbances of visual sensory function. A positive Babinski reflex is one of the common neurological signs that occur with bilateral INO and this was seen in this young Ethiopian patient. Smith and Cogan found that 45% of patients with bilateral INO had a positive Babinski's reflex revealing upper motor neuron lesions.[10] Other symptoms of MS, including paresthesia of the extremities, weakness of the limbs and face, vertigo, ataxia, micturition disturbance, fatigue and depression, may occur with bilateral INO.[10,11] The present patient had staggering gait, vertigo, paresthesia and burning sensation in the lower extremities, which were remitted with a short course of oral steroid therapy.

The horizontal gaze abnormality in the present case showed improvement within 3 months and completely resolved in 6 months. This is in agreement with Bolaños et al. who found similar result and further demonstrated that the etiology of INO and the MRI findings are key factors which affect the recovery or resolution rates. INO caused by infection, demyelination, and traumatic processes showed complete recovery in 66.6%, 61.9% and 60.0%, respectively while INO caused by stroke and tumors showed bad to no recovery.[13] Since the patient did not have history of head trauma and known medical illness or infectious process, the rapid and complete recovery of the horizontal gaze abnormality further supports the diagnosis of MS in the present case.

Myasthenia gravis can produce a motility pattern that can mimic a true INO and hence results in a pseudo-INO.[1,2] However, the patient did not have any other symptoms and signs of myasthenia gravis ruling out myasthenia gravis in the differential diagnosis of this case.

## 4. Conclusion

The occurrence of bilateral INO in young patients is highly suggestive of MS. Bilateral INO and other neuro-ophthalmic features are important clinical findings that may help establish a diagnosis of MS.

Though the incidence of MS in non-white population near the equator is low, eye care workers and other health care professionals should have high index of suspicion and be well versed with ophthalmic manifestations of MS as these manifestations could be the presenting sign of MS. Effective management of varied manifestations and/or complications

of MS requires an interdisciplinary team approach. The general practitioners/primary care physician, neurologist, ophthalmologist/neuro-ophthalmologist, and other health professionals should all be involved in the management and follow up of patients with INO and MS.

## Conflict of interest

The author declares that he has no competing interests.

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