Footnotes and Disclosure:

The authors have no proprietary or commercial interest in any materials discussed in this article.

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Rapid same-day resolution of internuclear ophthalmoplegia in Wernicke encephalopathy following parenteral high dose thiamine

Internuclear ophthalmoplegia (INO) is well known for its association with multiple sclerosis in young patients and ischaemic stroke in older patients. Lesions of the medial longitudinal fasciculus produce the clinical findings of INO. Wernicke encephalopathy (WE) is a metabolic disorder of thiamine deficiency that classically presents with confusion, ataxia, and nystagmus. Diplopia and ophthalmoplegia can also be the presenting or predominant complaint in WE. We report a case of WE presenting with an INO who had same-day resolution of symptoms and signs after high-dose parenteral thiamine. To our knowledge, this is only the second such case of documented unilateral INO with WE to be reported in the English-language literature (Table 1).

A 44-year-old previously healthy male with a history of chronic alcohol abuse presented to the emergency department with acute, painless, binocular diplopia worse on right gaze, oscillopsia, and gait ataxia. He reported consumption of 18 cans of beer the previous night and awoke the next morning with the above symptoms. He denied numbness, tingling, weakness, or confusion. Surgical history was significant for right-hand surgery and tonsillectomy in youth. He had a 4-pack-year smoking history and reported consumption of 30−40 beers per week for years.

On ophthalmologic examination, best corrected visual acuity was 20/20 OU. The pupils measured 6 mm OD and 7 mm OS in the dark and 3 mm OD and 4 mm OS in the light with no relative afferent pupillary defect. Intraocular pressures measured 14 mm Hg OD and 13 mm Hg OS. He had a 2 prism diopter left hypertropia in primary gaze. Motility examination was significant for −2 adduction deficit OS, and a dissociated, horizontal, abducting eye nystagmus OD on gaze to the right consistent with a left INO. There was a gaze-evoked torsional nystagmus in all positions of gaze (Video 1, available online). Doll's head manoeuvre was negative. Computed tomography of the head and orbits and magnetic resonance imaging of the head showed an old left orbital floor fracture. Findings of additional testing, including human immunodeficiency virus, hemoglobin A1c, erythrocyte sedimentation rate, vitamin B12, folate, thyroid function studies, and C-reactive protein, were normal. High-dose intravenous (IV) thiamine at 500 mg TID and magnesium were administered for presumed WE. Later assessment by the neurology service noted fluent speech, normal reflexes, and motor strength. However, tandem gait and finger-to-nose testing showed dysmetria. Same-day follow-up approximately 18 hours after initial examination and treatment demonstrated complete resolution of the adduction deficit, INO, nystagmus, and hypertropia (Video 2, available online).

WE is a clinical diagnosis characterized by acute-onset ophthalmoplegia, ataxia, confusion, and memory disturbance with occasional unexplained hypotension and hypothermia.[1] Typical oculomotor findings in WE include involvement of cranial nerve III, cranial nerve VI, and vestibular nuclei causing conjugate gaze palsies.[2,3] Gaze-evoked and spontaneous upbeat nystagmus can be seen, as can horizontal or vertical ophthalmoplegia.[4] This case demonstrates isolated, unilateral INO as an uncommon presentation of WE, which had been rarely described previously.

In 1987, Atlas et al. noted WE as a cause of INO.[5] Similarly, a 2005 article noted 2 out of 410 patients with INO caused by probable Wernicke syndrome.[6] In a 1990 report of 5 patients with WE, Gallucci et al. noted 1 patient with INO, though no further details were provided.[7] Additionally,

Table 1—Cases of Wernicke encephalopathy with internuclear ophthalmoplegia in the past 30 years

Author (Year)	Case
Kumar et al. (2000)[9]	Unilateral INO with WE in a patient with chronic alcohol misuse
Gallucci et al. (1990)[7]	One out of five patients with WE had INO; laterality unknown
Keane (2005)[6]	Two patients with INO and hydrocephalus with probable WE out of 410 patients with INO; laterality unknown

INO, internuclear ophthalmoplegia; WE, Wernicke encephalopathy.

a case has been reported of bilateral INO with WE.[8] To our knowledge, the only case of unilateral INO due to WE reported in the English language in the past 30 years was in the year 2000 by Kumar et al.[9]

High-dose IV thiamine is needed in individuals with WE, as delay in treatment can lead to permanent neurologic disability. A high plasma-to-CNS concentration gradient is needed for thiamine to cross the blood−brain barrier. Likewise, studies have shown that oral administration of water-soluble thiamine neither elevated thiamine activity in the cerebrospinal fluid nor restored transketolase activity to normal in alcoholics with thiamine deficiency.[10] Additionally, magnesium is an important cofactor for reactions involving thiamine, and thus hypomagnesemia can prevent the adequate use of thiamine.[1,11]

The recommendations for patients diagnosed with WE are high-dose 500 mg thiamine hydrochloride and magnesium parenterally acutely unless contraindicated, as there is minimal side effect with administration. This is in contrast to the lower dose (100 mg) that is typically present in most vitamin supplementation (e.g., "banana bags"). Ordering serum levels of thiamine may be done, but treatment should be started empirically without waiting for the laboratory results as delay in treatment could increase the chances of permanent neurologic damage in WE. High-dose thiamine is continued initially at 500 mg TID for 2−3 days, followed by oral 100−250 mg daily for a minimum of 1 week. Treatment may be continued thereafter in high-risk patients.

Acute unilateral or bilateral ophthalmoplegia (including INO) with or without nystagmus or other neurologic symptoms/signs (e.g., ataxia, nystagmus, confusion) should prompt consideration of the diagnosis of WE. Although excessive chronic alcohol consumption is a common cause of WE, other possible causes, including hyperemesis gravidarum, recent gastric bypass surgery, eating disorders, or malabsorption syndromes, should also be considered. Consider empiric administration of high-dose (500 mg TID) IV thiamine and magnesium in any patient suspected of having WE.

Supplementary materials

Supplementary material associated with this article can be found in the online version at doi:10.1016/j.jcjo.2019.07.007.

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Recovery of stereopsis after strabismus surgery in X-linked ocular albinism

X-linked ocular albinism (XLOA, Nettleship Falls, MIM #300500) is a phenotype of albinism restricted to the eye: iris transillumination, nystagmus, foveal hypoplasia, and excessive decussation of retino-striate fibers at the optic chiasm.[1] Abnormal visual pathway development is reflected in strabismus, absence of stereopsis, nystagmus, and decreased visual acuity.[1] We report the outcomes of 3 patients with clinically diagnosed XLOA who underwent strabismus surgery from 1994 to 2017, two of