Iranian Journal of Neurology

Clinical Note

Ir J neurol 2012; 11(4): 159-161

Wernicke's encephalopathy in a non-alcoholic Patient: Difficulties of early diagnosis and treatment

Omid Hesami[1], Nahid Beladimoghaddam[1], Farhad Assarzadegan[1], Nasim Kazemi[1]

[1] Emam Hossein Hospital, Department of Neurology, Shahid Beheshti University of Medical Sciences, Tehran, Iran

Keywords
Wernicke's Encephalopathy, MRI, Total Parenteral Nutrition, Non-Alcoholic, Iran

Introduction

Wernicke's encephalopathy (WE) is characterized by nystagmus, ophthalmoplegia, ataxic gait, and mental change resulting from thiamine deficiency.[1]

WE is a well-known complication of thiamine deficiency due to excessive alcohol consumption. Symptomatic thiamine deficiency in nonalcoholic patients is a less recognized and often misdiagnosed condition. The diagnosis of WE is mostly made clinically; nevertheless, magnetic resonance imaging (MRI) has been recognized as a useful adjunct in diagnosis. WE is a treatable disorder. If treated in the acute stage, many of the neurologic complications can be reversed.[1-3] In the current essay, we report a patient with Crohn's disease who developed WE as a result of thiamine deficiency caused by malnutrition and lack of multivitamin infusion (MVI) in total parenteral nutrition.

A 20 year-old non-alcoholic man was admitted to the Gastrointestinal Ward of Emam Hossein Hospital due to severe vomiting, diarrhea and general weakness since the previous 2 weeks. He had suffered a similar episode 15 months earlier and Crohn's disease had been suggested as a possible cause. Since then, he had had 40 kg weight loss. On admission, he was put on TPN. After one week, neurologic consultation was done for progressive confusion and weakness. Neurologic examination revealed signs of encephalopathy. He had apathy and impaired memory. Bidirectional horizontal nystagmus and right lateral rectus palsy were noticed. Muscle strength exam was difficult to interpret because of severe allodynia in limbs and confusion, but he was able to move his limbs against gravity. Tendon reflexes were absent and plantar stimulation produced no movement. The patient was unable to stand and ataxia could not be assessed reliably due to impaired consciousness.

Given the clinical history of dietary deprivation together with the neurological signs, a diagnosis of WE was strongly suspected and intravenous thiamine was prescribed. Intravenous thiamine is not available in the Iranian pharmaceutical market; thus, high dose intravenous B complex (30 ml per day containing 300 mg thiamine) was administered.

The patient's brain MRI revealed symmetrical high signals in periventricular regions and mammillary bodies most evident in fluid-attenuation inversion recover (FLAIR) sequence.

Two days later, the patient went into cardiac arrest. Intubation was done and cardiopulmonary resuscitation was started immediately. Sinus rhythm was restored and the patient was admitted to the intensive care unit (ICU).

Echocardiography showed 60% ejection fraction and mild pericardial effusion. No other abnormality was seen. Considering the side effects of long-term

Email: ijnl@tums.ac.ir

Corresponding Author: Nasim Kazemi, MD
E mail: kazemi90@yahoo.com

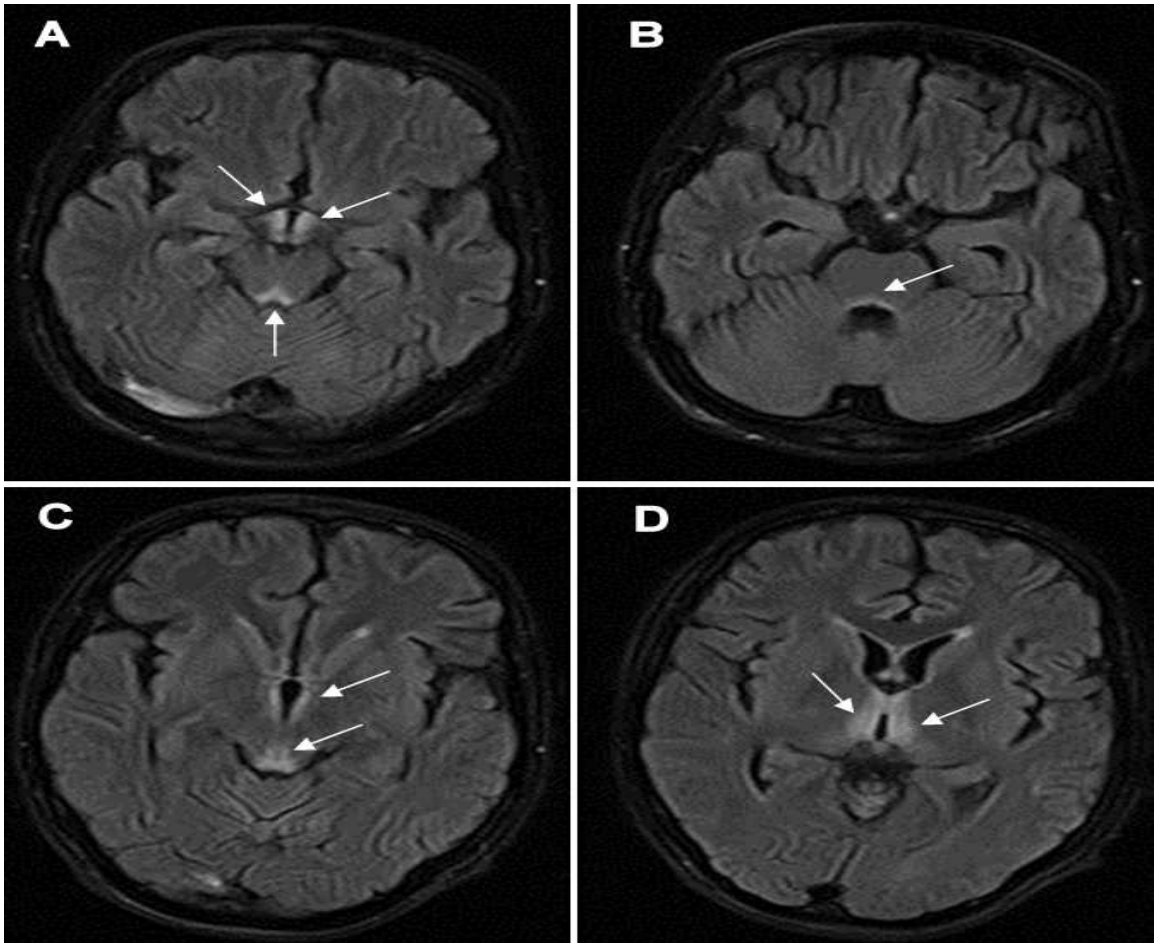

Figure1. Brain MRI on FLAIR in a nonalcoholic young man with WE due to malnutrition. Hypersignal lesions are observed in; A: tectalplate and mammillary bodies; B,C: periventricular regions; and D: medial thalamus

high dose IV administration of B complex (due to its high content of preservatives and associated risks for patient's cardiac status), and the limitations of IM injections in a bedridden patient (although in the first three days following ICU admission the patient received Neurobion injection TDS containing 100 mg thiamine), there was no other choice but oral administration despite its malabsorption. Therefore, IV and IM administration was replaced by oral consumption of high dose vitamin B1 (300 mg tablets three times a day). After one week he was awake but unconscious and unresponsive, although nystagmus and gaze palsy had disappeared. There was no limb movement. After two months, the patient was transferred to the Gastrointestinal Ward. No further improvement was observed and the patient passed away due to cardiac arrest.

WE is characterized by ocular signs, ataxia and mental change. These symptoms may occur in various combinations; thus, WE should be considered as a possible diagnosis when the classic triad is absent. The most common presenting symptom is the altered consciousness. The ocular abnormalities include nystagmus, paralysis of abducens and the conjugate gaze. Ataxia usually manifests as gait instability.[1] Beriberi is the systemic counterpart of thiamine deficiency and often manifests as cardiac involvement and painful peripheral neuropathy. Pericardial effusion and cardiac arrest in this young patient without any previous history of cardiac disease could be attributed to the systemic manifestations of thiamine deficiency.

WE is most commonly associated with heavy alcohol consumption; however, it has been reported in non-alcoholic patients with low dietary thiamine intake. In a healthy individual, thiamine reserves are estimated to be exhausted after only 2-3 weeks of dietary deprivation.[1] In addition, recent evidence shows that many patients under total parenteral nutrition (TPN) develop this syndrome after a few weeks due to the absence of adequate thiamine supplementation (MVI).[2-5] In our patient, nutritional deficiency was the consequence of ongoing vomiting and diarrhea, and TPN with inadequate thiamine supplementation. If not recognized, WE can progress to Korsakoff's syndrome and may be even fatal. On

the other hand, immediate thiamine administration can reverse the symptoms.

MRI has been recognized as a useful adjunct for WE diagnosis and reveals the typical lesions in up to 80% of suspected cases. A typical MRI finding in WE is the symmetrical signal alteration in the periventricular regions of the brain, including medial thalamus, periaqueductal grey matter, tectal plate and mammillary bodies. FLAIR imaging is the ideal MRI sequence for identifying the lesions.[3,4] MRI study in this patient revealed all these typical changes.

A good response to treatment with thiamine supplementation confirms the diagnosis. In this patient, brain hypoxic–ischemic damage due to cardiac arrest interrupted the natural course of the disease. Thus, response to treatment could not be seen completely considering the additional complications.

This patient represents the nonalcoholic patient population at risk of thiamine deficiency. In this case report, we emphasized on the low clinical threshold for initiation of thiamine administration in non-alcoholic patients with mental changes and highlighted the predisposing factors for dietary thiamine deficiency. The characteristic MRI changes and thiamine assays may help in confirming the diagnosis.

Recently, there has been a nationwide lack of multivitamin infusion (MVI) and intravenous thiamin in Iran. Many patients are receiving TPN without MVI. Because of the shortage of MVI and the importance of appropriate vitamin supplementation during TPN, we recommend that intravenous thiamine be available in each emergency unit and medical ward as a routine medication. There are no formal dose-ranging placebo-controlled studies on the use of B-complex vitamins in the treatment of WE. Thiamine 500 mg IV three times daily for 2 to 3 days and 250 mg IV daily for the next 3 to 5 days given as an infusion followed by 100 mg orally three times daily is in accordance with the evidence-based guidelines.[6]

References

1. Sechi G, Serra A. Wernicke's encephalopathy: new clinical settings and recent advances in diagnosis and management. Lancet Neurol. 2007; 6(5): 442-55.
2. Mattioli S, Miglioli M, Montagna P, et al. Wernicke's encephalopathy during total parenteral nutrition: observation in one case. JPEN J Parenter Enteral Nutr. 1988; 12(6): 626-7.
3. Zuccoli G, Santa CD, Bertolini M, et al. MR imaging findings in 56 patients with Wernicke encephalopathy: nonalcoholics may differ from alcoholics. AJNR Am J Neuroradiol. 2009; 30(1): 171-6.
4. Davies SB, Joshua FF, Zagami AS. Wernicke's encephalopathy in a non-alcoholic patient with a normal blood thiamine level. Med J Aust. 2011; 194(9): 483-4.
5. Francini-Pesenti F, Brocadello F, Manara R, et al. Wernicke's syndrome during parenteral feeding: not an unusual complication. Nutrition. 2009; 25(2): 142-6.
6. Thomson AD, Guerrini I, Marshall EJ. The evolution and treatment of Korsakoff's syndrome: out of sight, out of mind? Neuropsychol Rev. 2012; 22(2): 81-92.