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CASE REPORT

Thiamine-deficient optic neuropathy associated with Wernicke's encephalopathy in patients with chronic diarrhea

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The deficiency of thiamine manifesting as Wernicke's encephalopathy (WE) and concurrent optic neuropathy is rare. Herein, we report the case of a 29-year-old patient who suffered from bilateral sudden blindness and a disturbance of consciousness after 2 months of chronic diarrhea and minimal food intake. In addition, bilateral abducens nerve palsy with multidirectional nystagmus and no light perception in both eyes were noted. An ophthalmoscopic examination revealed bilateral disc edema with peripapillary flame-shaped hemorrhages. Although the results of analyzing the composition of cerebrospinal fluid showed that they are within normal limits, magnetic resonance imaging (MRI) revealed bilateral hyperintensity over the mammillary body, dorsal medial thalamus, and periaqueductal gray matter. As we suspected thiamine deficiency-induced WE, a high dose of intravenous thiamine was prescribed. After the administration of thiamine, both visual acuity and visual field rapidly improved with the simultaneous recovery of consciousness. This case indicates that, although rare, thiamine deficiency with WE may still occur in patients with chronic diarrhea in Taiwan. Thiamine deficiency should be considered in the differential diagnosis for patients who encounter sudden visual loss after prolonged periods of poor food intake and poor vitamin supplementation.

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Introduction

Although thiamine deficiency may cause various disorders, such as high output heart failure (wet beriberi), lactic acidosis, and gastrointestinal beriberi, the most important neurological disorder is Wernicke's encephalopathy (WE),[1,2] which is clinically characterized by the classical triad of ocular abnormalities, ataxia, and disturbances of consciousness.[3] All of these conditions carry a high rate of morbidity and mortality when unrecognized.

Ocular manifestations of WE are nystagmus, ophthalmoplegia, and optic neuropathy.[4] However, optic neuropathy is not a common presentation of WE and has only been reported in a few anecdotal case reports.[5–11] In a case series of 245 patients with WE, optic neuropathy was reported to be present in only 2.6% of the patients.[4] To the best of our knowledge, WE with optic neuropathy has never been documented in Taiwan. Herein, we report a case of optic neuropathy presenting as optic disc edema with peripapillary flame-shaped hemorrhages and severe visual impairment in a nonalcoholic male patient with thiamine deficiency and WE.

Case report

A 29-year-old male patient without systemic diseases or associated family history presented with sudden bilateral blindness, ataxia, disturbance of consciousness, and body weight loss (20 kg) after 2 months of chronic diarrhea at a frequency of approximately one to two bowel movements a day and poor food intake. The patient's family reported that he did not use drugs, neither drank alcohol nor smoked tobacco. A few weeks before admission, his food intake decreased and consisted mainly of carbohydrates, because of frequent nausea, vomiting, and intermittent epigastric cramping pains immediately after eating. General weakness had rapidly progressed and he had become more dependent on manual assistance in the last 20 days. He experienced blurred vision (oculus unitas) about 7 days before being admitted in our hospital. As the condition rapidly deteriorated, he ultimately became confined to bed and developed mental changes with total blindness within 3 days. On presentation, he exhibited severe truncal ataxia, a state of total confusion, bilateral abducens nerve palsy (Fig. 1A) with multidirectional nystagmus and no light perception (NLP) in both eyes. The patient showed signs of apathy, inattentiveness, and an indifference to his surroundings. Spontaneous speech was minimal, and provoked speech indicated general disorientation to time, place, and purpose. Results of a physical examination revealed a blood pressure level of 87/64 mmHg, pulse rate of 158 beats/minute, respiratory rate of 20 breaths/minute, and body temperature of 35.6°C. Bilateral disc edema with peripapillary flame-shaped hemorrhages was observed (Fig. 2) after ophthalmoscopic examination, and a slit-lamp examination showed an otherwise unremarkable anterior segment except for an absence of light reflex in both eyes. A computed tomography of the head showed normal findings. An analysis of the cerebrospinal fluid and sepsis workup including cytomegalovirus, syphilis, and human immunodeficiency virus revealed negative results. The intracranial pressure had not increased. Toxin profiles and tumor markers were analyzed, but no abnormal findings were evident. Biological chemicals and blood profile were all within normal limits except for lactic acidosis (lactate: 61 mg/dL; normal range: about 4.5—19.8 mg/dL). Liver functioning was also normal. A subsequent magnetic resonance imaging (MRI) of the brain revealed abnormal hyperintensity over the mammillary body, dorsal medial thalamus, and periaqueductal gray matter in the axial fluid-attenuated inversion recovery (FLAIR) images, with corresponding sites on diffusion-weighted images (DWIs) (Fig. 3). Although the facilities for examining the serum thiamine levels were not available in our hospital, thiamine deficiency was suspected because his nutritional intake had been insufficient for approximately 2 months. Associated WE was suspected based on the clinical history, symptoms, and MRI findings. Therefore, high doses of intravenous thiamine (300 mg/day) were given immediately after the initial evaluation. Visual acuity improved dramatically from NLP to counting fingers after 12 hours. Rapid recovery of

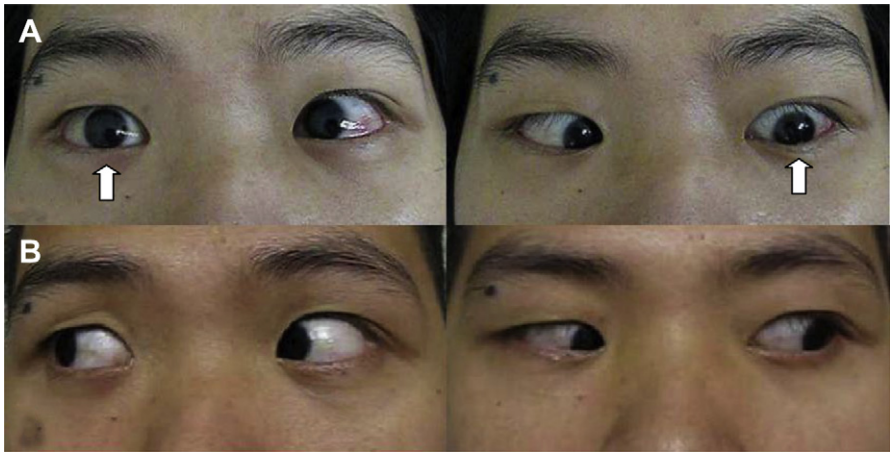

Figure 1 A 29-year-old male with chronic diarrhea and poor nutritional intake presented to our hospital with ataxia, multidirectional nystagmus, and sudden blindness. (A) Bilateral abducens palsy (arrow) was noted at the initial presentation. (B) Dramatic disappearance of bilateral abducens palsy after administering intravenous thiamine supplements for 1 week.