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CASE REPORT – OPEN ACCESS

Wernicke's Encephalopathy: A Forgotten Disease

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ABSTRACT

Wernicke's encephalopathy (WE) is an acute neurological condition characterized by ataxia, confusion, ocular findings, and impairment of consciousness due to thiamine deficiency. Although alcoholism is the most common reason, WE cases resulting from prolonged total parenteral nutrition (TPN) without multivitamin complex have been reported. Here we present a dramatic improvement in symptoms with high-dose thiamine in a patient who developed WE due to TPN after gastrointestinal surgery.

Keywords: Thiamin, Wernicke's encephalopathy, total parenteral nutrition, micro nutrient, ICU

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INTRODUCTION

Wernicke's encephalopathy (WE) is a syndrome caused by thiamine deficiency. It is characterized by ocular findings, cerebellar dysfunction (ataxia), and confusion. Thiamine (vitamin B1) is a water-soluble vitamin absorbed from the duodenum and jejunum, which acts as a coenzyme in glucose metabolism and neuronal activity (1). In developed countries, thiamine deficiency is typically associated with alcoholism because alcohol interacts with thiamine intake and use. However, WE can also be seen in nonalcoholic conditions, such as diuretic use, prolonged starvation, gastrointestinal disorders, excessive vomiting due to hyperemesis gravidarum, and increased nutritional requirement. The WE prevalence increases due to lack of thiamine use in total parenteral nutrition (TPN) (2, 3). The TPN is indicated in short bowel syndrome, gastrointestinal fistula, intestinal obstruction, and prolonged intestinal resting. Recently, the TPN incidence has increased in intensive care units (ICU) and clinics. Thus, these patients have a high risk of WE, but there is still not enough awareness about this syndrome.

In our report, we present a dramatic improvement of symptoms with high-dose thiamine treatment in a patient who received TPN after gastrointestinal surgery and developed WE.

CASE REPORT

A 31-year-old man with toxic megacolon surgically treated with intestinal resection 18 years before, which resulted in short bowel syndrome, was followed for the Hirschsprung disease, and he had no history of alcohol abuse. The patient applied to hospital with jaundice, and a gallbladder stone was detected; thus, he was admitted to hospital for cholecystectomy. During surgery, a complication occurred due to intestinal injury that caused fistula of the small intestine. Oral intake was discontinued due to fistula, and the patient received TPN for 1 month. But TPN solutions did not contain thiamine supplement, and he did not receive any intravenous thiamine. During the treatment and follow-up with TPN, the patient was transferred to ICU due to worsening clinical symptoms.

In ICU, the patient presented with impaired consciousness, metabolic acidosis, ataxia, nystagmus, diplopia, and hypotension. Septic shock was initially diagnosed, and an antibiotic, fluid replacement, and vasopressor therapy were started immediately. The arterial blood gas was demonstrating metabolic acidosis: pH, 7.11; pCO_2, 12; pO_2, 136; lactate, 13; and HCO_3, 7. However, no improvement was observed during follow-up. On cranial magnetic resonance imaging obtained during the ICU follow-up, a hyper-intensity surrounding the aqueduct was detected on axial FLAIR sequences, while bilateral, symmetrical hyper-intensity was detected at dorsomedial of thalamus on DWI images, but no increased signal intensity was detected at the same localization on ADC images (Fig. 1a). In addition, symmetrical, bilateral hyper-intensity was detected in the subthalamic nucleus localization at dorsomedial of thalamus on axial T2-weighted and FLAIR images (Fig. 1b), while a symmetrical, bilateral hyper-intensity (arrows) was observed in the subthalamic nucleus localization at dorsomedial of thalamus on coronal T2-weighted images (Fig. 1c). The clinical and radiological findings were considered to be in agreement with WE, and thiamine (3×100 mg, IM) was added to treatment. There was no necessity to apply a vasopressor after 24 hours, and arte-

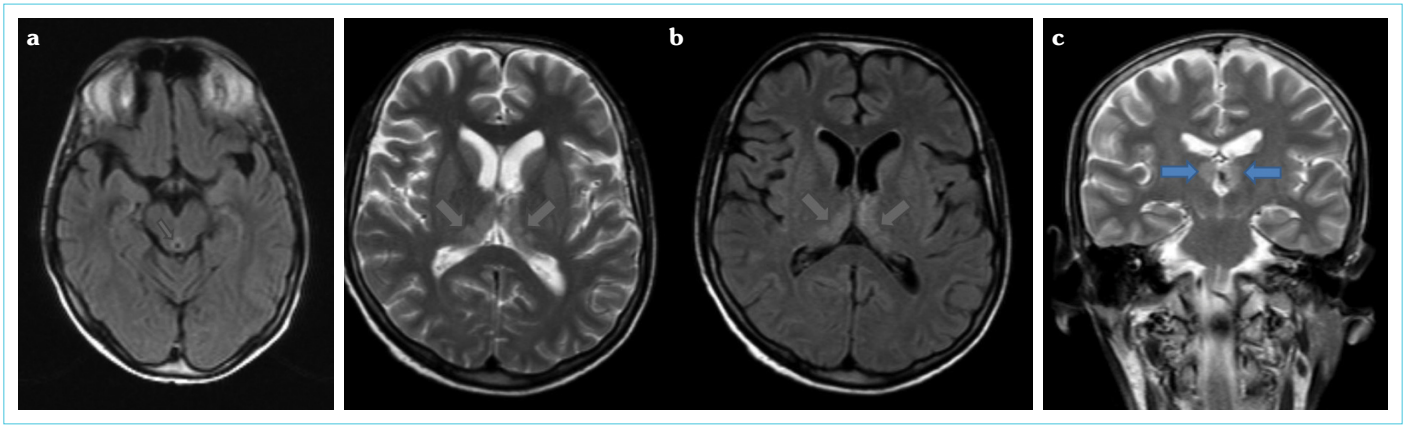

Figure 1. a–c. (a) A hyper-intensity was observed in the surrounding aqueduct on an axial FLAIR sequence. (b) On axial T2-weighted and FLAIR images, a symmetrical, bilateral hyper-intensity was observed in the subthalamic nucleus localization at dorsomedial of thalamus. (c) A symmetrical, bilateral hyper-intensity was observed in the subthalamic nucleus localization at dorsomedial of thalamus on coronal T2-weighted images

rial blood gases returned to normal (pH, 7.44; pCO_2, 30.5; pO_2, 87.4; lactate, 1.81; HCO_3, 20.5). The patient was discharged back to the ward after a 2-day stay in ICU.

DISCUSSION

WE is a disorder characterized by an acute/subacute onset of a clinical triad including confusion, ophthalmoplegia, and ataxia. Although WE is typically observed in chronic alcoholism, it may also occur due to thiamine deficiency resulting from gastrointestinal surgery, systemic infection, or dietary deficiency (imbalanced diet/thiamine-free TPN) (4). The presence of all three findings is helpful in early diagnosis.

The magnetic resonance imaging is the most important diagnostic tool for early diagnosis, prognostication, and identification of atypical cases and has a 53% sensitivity and 93% specificity (5). In our patient, there was a hyper-intensity surrounding the aqueduct detected on axial FLAIR sequences, while a bilateral, symmetrical hyper-intensity at dorsomedial of thalamus on DWI images was observed, but there was no increase in the signal intensity at the same localization on ADC images. In addition, there was also a symmetrical, bilateral hyper-intensity in the subthalamic nucleus localization at dorsomedial of thalamus on axial T2-weighted and FLAIR images, while a symmetrical, bilateral hyper-intensity was observed in the subthalamic nucleus localization at dorsomedial of thalamus on coronal T2-weighted images (arrows). These findings were considered to be compatible with WE.

The prevalence of WE is 1%–3%, as can be concluded mainly from autopsy studies. Several studies show that the prevalence rates on clinical studies are lower than autopsy studies because the diagnosis is usually overlooked and unnoticed (2). If it is unidentified in the acute/subacute phase, hypotension, tachycardia, the ST segment, and T-wave changes on electrocardiogram (ECG) (heart failure as a finding of cardiac beriberi), profound hypothermia and lactic acidosis, myosis and unresponsiveness to light, stupor, coma, and death will occur in further stages. The diagnosis of WE is particularly important in surgical patients because hemodynamic instability (hypotension, tachycardia, hypothermia, ECG changes, altered mental status, acidosis, or hyperventilation that cannot be explained by hypovolemia) can easily be confused with postoperative complications.

The prevention of WE depends on adequate nutrition. In adults, the estimated thiamine requirement is 1.4 mg/day or 0.5 mg/1000 calories. Decreased thiamine levels are related to reduced intake and appetite, nausea, vomiting, diarrhea, oral thrush, which is common in cancer, hyperemesis gravidarum, severe psychiatric disorders, and bariatric and gastrointestinal surgery. These patients usually receive artificial nutrition, especially parenteral. As it was reported in the literature, most of the cases who received TPN without thiamine develop nonalcoholic WE (5–7).

If symptoms suggestive of WE are observed, thiamine hydrochloride should be given empirically (8). However, there is no consensus about the optimal dose, duration, and frequency of thiamine treatment. In many case reports, it was stated that 100–200 mg/day thiamine in nonalcoholic patients and a high-dose (1500 mg/day, 3×500 mg) in alcoholic patients is adequate for recovery. In addition, intravenous administration of thiamine is also recommended. Thiamine treatment should be continued until no more progress is observed in symptoms and findings. A persistent neurological dysfunction in WE is common. Both an early diagnosis and timely treatment are very important in the prevention of permanent brain injury (8). As thiamine has no potential adverse effects, treatment should be given promptly even in case of suspected thiamine deficiency. Francini-Pesenti et al. retrospectively evaluated 5,411 adult cases who received TPN, and 7 cases developed WE due to TPN. The response to thiamine treatment was reported in 6 cases, but neurological sequelae were only reported in 1 case (9). Similarly, Nishimoto et al. reported that 11 patients with suspected WE received high-dose parenteral thiamine (≥500 mg IV thiamine) for a median of 3 days. Eight out of 11 patients (73%) showed resolution of symptoms (4).

As a result, WE should be considered in patients with a history of alcohol abuse, malnutrition, and long-term TPN, when there is an onset of mental confusion, ataxia, or ocular findings. In that case, it should be kept in mind that intravenous thiamine supplementation could reverse all these findings, and if there

is any suspicion, it should be empirically started according to recommendations. Also, it should not be ignored that WE may develop in TPN without appropriate thiamine supplementation. Therefore, parenteral solutions should be routinely supplied with vitamin preparations.

Informed Consent: Written informed consent was obtained from the patient for the publication of the case report and the accompanying images.

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