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Total parenteral nutrition caused Wernicke's encephalopathy accompanied by wet beriberi

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Statistical Analysis C
Data Interpretation D
Manuscript Preparation E
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Patient: Female, 17
Final Diagnosis: Wernicke's encephalopathy
Symptoms: Blurred vision • dizziness • nystygmus • tachycardia
Medication: —
Clinical Procedure: —
Specialty: Neurology

Objective: Mistake in diagnosis

Background: Wernicke's encephalopathy (WE) is an acute and life-threatening illness which is not only seen in alcoholics, but also in persons with poor nutrition lacking thiamine (vitamin B1).

Case Report: Here, we presented a case of WE in a patient who received parenteral nutrition without complement of thiamine. Besides neuropsychiatric problems, she also manifested prominent cardiovascular abnormalities, which were consistent with wet beriberi.

Conclusions: This case emphasizes the need for thiamine supplementation in prolonged total parenteral nutrition, and also highlights the awareness of WE in persons with parenteral nutrition lacking thiamine. More importantly, we call for attention to wet beriberi in such persons.

MeSH Keywords: Parenteral Nutrition, Total – adverse effects • Thiamine Deficiency • Beriberi • Wernicke Encephalopathy

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Background

Wernicke's encephalopathy (WE) is a neurological emergency that is lethal if not diagnosed or treated properly. Although it is most associated with alcoholism, it can accompany other disorders that may be easily ignored, for example, any situation that can cause deficiency of thiamine (vitamin B1): vomiting, dietary imbalance, fasting, and other gastrointestinal diseases. The main clinical symptoms are the classical triad of oculomotor abnormalities, ataxia, and mental-status changes. Additionally, cardiovascular abnormalities, which are also called "wet beriberi", can be obvious simultaneously. We here report one case with WE and wet beriberi during total parenteral nutrition, but who was initially misdiagnosed.

Case Report

A 17-year-old female patient exhibited schizophrenia and was prescribed olanzapine, chlorpromazine, and aripiprazole 2 years ago. After about 2 years of maintenance therapy, she was admitted into the department of gastrointestinal surgery for fever, constipation, and abdominal pain, without change of diet habits. The diagnosis was incomplete intestinal obstruction, thus she was treated with antibiotics and gastrointestinal decompression. She was also given parenteral nutrition, which contained glucose, lipid, amino acid, vitamin B6, vitamin K1, vitamin C, ATP, and coenzyme A. Three weeks later, she presented dizziness, blurry vision, horizontal gaze-evoked nystagmus of the right eye, and unsteady walking, as well as tachycardia (120 beats per minute). After another 3 days, eye movement disorder – incomplete abduction in both eyes – occurred. In the meanwhile, funduscopy found bilateral retinal hemorrhage. At that time, Guillain-Barré syndrome or viral encephalitis was suspected, whereas lumbar puncture showed normal pressure, normal cell count and chemical tests. However, the condition was still getting worse and 4 days later her blood pressure decreased to 75/40 mmHg. Therefore, she was maintained with dopamine, and her heart rate was 140 beats per minute, she became somnolent, and had dysarthria. Then the cerebral MRI scan showed symmetrical hyperintense signals on T2-weighted imaging (T2WI) and fluid-attenuated inversion recovery (FLAIR) in the bilateral thalamus, mid-brain, pons, and medulla (Figure 1A, 1B). When we considered the typical MRI results combined with the total parenteral nutrition lacking thiamine, we suspected the patient had WE. Thiamine (100 mg/d) was immediately administered intravenously. After treatment, dramatic improvements occurred. On the next day she regained consciousness, and her blood pressure and heart rate returned to normal without use of vasoactive drugs. With a daily supplement of thiamine, her condition improved quickly. One week after thiamine therapy, her symptoms of somnolence, dysarthria, dizziness, nystagmus, eye movement problems, and ataxia all vanished. A follow-up cerebral MRI 1 month later in another hospital found that the abnormal signals all disappeared (Figure 1C, 1D).

Discussion

WE was initially described in malnourished alcoholic persons. However, it can be induced by any medical conditions producing malnourishment, including hyperemesis gravidarum, thyrotoxicosis, peptic ulcer, stomach cancer, gastric bypass, anorexia nervosa, re-feeding after starvation, chronic hemodialysis, and prolonged parenteral nutrition [1]. The biological half-life of thiamine is 9–18 days and has a minimal storage which is about 30 mg [2]; thus, in malnutrition it easily becomes depleted. As a result, in the presented case, the onset of WE was 21 days after thiamine starvation. The prevalence of WE associated with prolonged parenteral nutrition lacking thiamine supplementation is rather high [3,4].

The clinical symptoms are the triad of mental confusion, oculomotor dysfunctions, and ataxia. However, only 10% of patients present all 3 of these [5] and 19% of patients do not show any of them. Mental changes range from apathy, irritability, mental sluggishness, damaged memory and orientation, to stupor and coma. Oculomotor dysfunctions include nystagmus and ophthalmoparesis. Nystagmus is often the first oculomotor abnormality and usually is evoked by horizontal gaze. Rectus palsy and even complete ophthalmoplegia can be encountered. Ptosis and retinal hemorrhage occasionally occur. Ataxia affects the trunk and lower limbs the most and displays as difficulty with standing and walking. Sometimes wide-base, small, and slow gait are seen.

Apart from neuropsychological manifestations, thiamine deficiency can also lead to cardiovascular dysfunction called "wet beriberi", which is characterized by tachycardia and hypotension. It is frequently mistaken for acute coronary syndrome, septic shock, and autonomic nervous disease if the doctors are not familiar with the clinical manifestation spectrum of thiamine deficiency. Wet beriberi has rarely been described in WE in the literature. In Hahn's report [4], patients with WE and wet beriberi received the exact diagnosis as late as 16 days after onset and 14 days after admission. With increased knowledge, wet beriberi is getting more attention in thiamine deficiency [6]. Similarly, the case presented here showed obvious cardiovascular disease – tachycardia and hypotension – which added to the diagnostic evidence of WE. Luckily, the right decisions were made in time, so she had a relatively good prognosis. It reminds us again that we need sufficient theories and clinical experiences to avoid delayed diagnosis of WE.

Because only one-third of patients present all of the typical triad, adjuvant examinations are important in diagnosis. MRI is

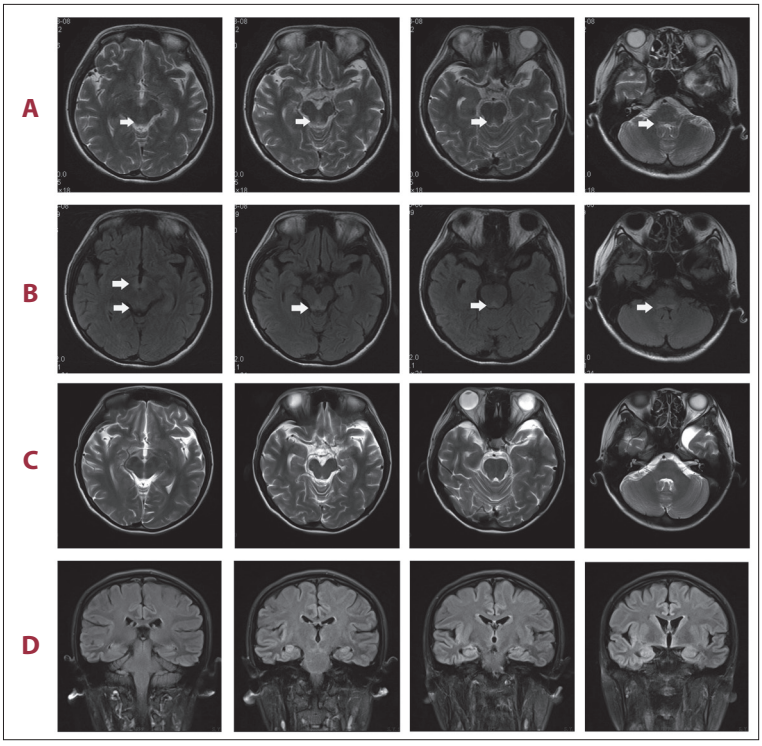

Figure 1. MRI images of the patient for the first time (**A, B**) and one month later (**C, D**). Arrows show the abnormal signals on T2-weighted imaging (T2WI) (**A**) and FLAIR axial (**B**) images in the bilateral thalamus, mid-brain, pons and medulla, but no abnormal signal is found on T1-weighted imaging (T1WI). MR images done one month later (**C**) show that the unusual signals in thalamus, mid-brain, pons and medulla once found on T2WI have all gone. FLAIR coronal images (**D**) are negative, but FLAIR axial images were not done in the second hospital.

particularly valuable in the diagnosis of WE, with a sensitivity of 53% and a specificity of 93%. As a result, MRI can be used to confirm the diagnosis, but a normal MRI finding cannot rule out WE. The most common MRI findings include hyperintense signals in the midline around the third ventricle, cerebral aqueduct, floor of the forth ventricle, and cerebral cortex in FLAIR imaging symmetrically and bilaterally [7]. Similar signal changes can also be found on T2WI, but the lesions are not as distinct because of the high signal of CSF, which is in accordance with the case reported here. In some cases, MRI images show no abnormalities even when clinical symptoms are extremely severe. The MRI images in our patient revealed relatively mild changes; as a result, with prompt high-dose thiamine therapy, hyperintense signals in T2WI vanished. This phenomenon demonstrate that the imaging changes are reversible if patients are treated properly in the acute stage. Other tests, like measurement of erythrocyte thiamine transketolase activity or thiamine/thiamine pyrophosphate concentration in serum, contribute to the diagnosis but are not routinely carried out in most institutions.

WE is easy to misdiagnose if the doctors do not have good knowledge of it [8]. A considerable number of cases are undiagnosed, as proven by autopsy studies. The case reported here was misdiagnosed initially.

After diagnosis, treatment with thiamine should be started immediately so as to prevent permanent neurologic injury. It is recommended to supplement thiamine intravenously or intramuscularly. In the meanwhile, other vitamins should be replaced as well. As the encephalopathy is usually precipitated by a carbohydrate load, carbohydrate should not be given if WE is presumptive, until supplementation of thiamine. Generally, response to treatment is rapid. Nystagmus typically resolvers quickly within several days, but ophthalmoplegia may last for 2 weeks. In contrast, ataxia improves relatively slowly, while cognitive function recovers most slowly and even can persist for life [9]. The current case received the right treatment in time, and had a good outcome.

Conclusions

Our report suggests requirements for nutrition; thiamine, along with other vitamins, is recommended for the population at risk of developing WE, including those with total parenteral nutrition. In the presence of neuropsychological symptoms in these at-risk patients, awareness of WE is necessary. In addition, we at the same time noticed some symptoms other than the WE triad – wet beriberi – that are rarely described in WE.

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