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CASE REPORT

General Medicine

Wernicke Korsakoff syndrome in a teenage female as a complication of COVID-19

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Abstract

Wernicke encephalitis (WE) is usually associated with alcohol use disorder and caused by a deficiency in thiamine. Classic findings include confusion, ataxia, and ophthalmoplegia. This case is a unique presentation of WE in a 14-year-old female related to prior coronavirus disease infection. She had persistent dysgeusia and developed thiamine deficiency. She presented with confusion, ataxia, and changes in speech. She had a prolonged hospitalization but was discharged to an inpatient rehab facility with persistent symptoms. It is prudent to include thiamine deficiency in the differential for patients with any symptoms of WE and a history of nutritional deficiency.

1 | INTRODUCTION

Wernicke encephalitis (WE) is a neurological disorder secondary to profound thiamine (vitamin B1) deficiency that can lead to permanent neurological damage and even death. Chronic nutritional deficits lead to thiamine deficiency with alcoholism being a classic cause of WE. The classic findings of WE include gait ataxia, ophthalmoplegia, and altered mental status. Unfortunately, this triad is only present in up to 33% of cases leading to it being underdiagnosed and undertreated.¹⁻⁴ A total of 23% of cases of WE are non-alcoholic with a total prevalence from all causes of up to 2.8% in developed countries.^{1,3,4} We present the case of a 14-year-old female who developed thiamine deficiency, leading to a diagnosis of WE, secondary to poor oral intake after a change in her taste from a coronavirus disease 2019 (COVID-19) infection.

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2 | CASE REPORT

A 14-year-old previously healthy female presented with her parents to the emergency department with concerns based on 3 days of worsening ataxia, confusion, inappropriate speech, urinary incontinence, and changes in level of consciousness. Her parents stated her symptoms were intermittent but had become progressively worse. History was significant for a 50-pound unintentional weight loss following ongoing dysgeusia after COVID-19 infection 10 months prior, as well as recent treatment with fluconazole for oral thrush 1 week prior to presentation with little improvement in her symptoms. They reported her having normal eating habits and not trying to lose weight before having COVID. Due to her weight loss, her body mass index changed from 36 to 27 after her illness.

On initial examination, her cranial nerves were intact, including extraocular movements, although she was ataxic with dysmetria on

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finger-to-nose testing bilaterally, had a wide base gait with mild ataxia, and was noted to have difficulty following commands during the examination. Vital signs and initial laboratory testing including lumbar puncture and head computerized tomography were unremarkable. After neurologic consultation, she was started on high dose Solu-Medrol for suspected acute disseminated encephalomyelitis.

Her clinical course abruptly changed with neurological failure and hemodynamic collapse, and a significant lactic acidosis of 11.6 mmol/L. She was obtunded, without a gag reflex, in significant respiratory distress, unable to tolerate secretions, hypotensive, and tachycardic. She was intubated and started on norepinephrine and epinephrine to maintain her blood pressure. Electroencephalography showed no epileptiform activity and echocardiogram showed normal cardiac function. Magnetic resonance imaging (MRI) of the brain revealed symmetric non-enhancing areas of signal alteration involving the thalami, hypothalamus, periaqueductal gray, and inferior colliculi consistent with WE. Her parents were further questioned about her daily oral intake, and they reported for the past 3 weeks she would eat 1–2 saltine crackers and drink a small amount of water due to a foul taste in her mouth.

The patient was started on high dose thiamine: 500 mg 3 times daily for 2 days, 250 mg daily for 3 days, followed by 100 mg 3 times daily throughout her inpatient admission. Prior to this, her thiamine level was found to be significantly low at 20 with a normal range being 70–180 nmol/L. She was weaned off norepinephrine later that day and extubated on day 6 of her pediatric intensive care unit (PICU) admission. Her course was complicated by development of severe hypertension and status epilepticus requiring reintubation. Repeat MRI now showed nearly symmetrical cortical involvement of the posterior pole and watershed regions that can be a rare manifestation of WE or an atypical posterior reversible encephalopathy syndrome (PRES). The neurologist believed these changes were PRES. She was treated with a nicardipine drip and anti-epileptics.

Once stabilized on oral anti-hypertensives and anti-epileptics, she was transferred out of the PICU to the general floor for another 6 days before being discharged to a rehab facility. Her neurological recovery at that time included significantly improved ataxia and she followed all directions. Ongoing deficits included ophthalmoplegia, difficulty swallowing, and she did not speak to staff. Her parents reported her conversing appropriately with them occasionally.

3 | DISCUSSION

WE was first named by Carl Wernicke in 1881 after observing 2 alcoholic patients and 1 patient who developed pyloric stenosis after attempting suicide by drinking sulfuric acid leading to critical nutritional deficiencies.^{5,6} Although classically associated with alcoholism, multiple other etiologies have been linked to WE including hyperemesis gravidarum, bariatric surgery, anorexia, intestinal disorders with malabsorption, thyroid disease, and malignancy.^{1,4,7,8} The typical triad of WE, including gait ataxia, ophthalmoplegia, and altered mental status, is only present in up to 33% of cases leading to it being under-

diagnosed and undertreated.^{1–4} The consequence of being unrecognized is progression to Korsakoff syndrome that manifests with permanent memory loss and confabulation and even death in up to 20% of patients.^{1,3,5,7}

The diagnosis of WE can be made exclusively from history and physical examination but MRI can also reveal structural changes in the paraventricular regions of the thalamus, hypothalamus, mammillary bodies, periaqueductal region, and floor of the 4th ventricle. However, it has a low sensitivity of 53% and changes on MRI suggest a later stage of the disease.^{1,3,5} Other disorders associated with thiamine deficiency are wet beriberi and dry beriberi. The former causes cardiac dysfunction (most commonly high output cardiac failure) and the latter causes neurologic dysfunction (most commonly peripheral neuropathy).^{5,7,9}

Non-alcoholic-associated WE is associated with a relatively small percent of the total cases in developed countries. Ota et al³ stated 50% of cases are related to alcohol use disorder, but other studies found on autopsy that 23% of identified WE were people without excessive alcohol intake. WE has been identified and treated for patients with current COVID infection because severe illness itself leads to thiamine depletion.^{10–12} In this case, the patient developed thiamine deficiency secondary to poor oral intake following a change in her taste after having COVID-19 approximately 10 months prior to her presentation. COVID has been documented to cause dysgeusia for more than 3 months post COVID in some people.^{13–15} She had substantial weight loss over this time period but her neurologic symptoms started when her nutritional intake became even more sparse. She presented with initial symptoms of dry beriberi but progressed to wet beriberi additionally with significant hypotension and lactic acidosis.

Thiamine is water soluble and 30–50 mg can be stored in the body with a half-life of 10–18 days.^{8,16,17} Because thiamine is directly related to metabolism, daily intake is based on caloric intake with 1–2 mg recommended daily for the average adult. Depletion can occur in 2–3 weeks with the symptoms of WE appearing in 4–6 weeks.^{6,7,16,18} In this case, our patient likely already had significant thiamine depletion before limiting her diet to crackers and was then profoundly depleted causing her initial symptoms to appear. On initial presentation, she did not have ophthalmoplegia but was ataxic and her mental status rapidly declined.

Although WE classically shows abnormalities on MRI near the thalamus, hypothalamus, mammillary bodies, and 4th ventricle, it can also show abnormalities in other parts of the brain as well.^{1,5} The pathophysiology of thiamine deficiency in the brain is the development of cytotoxic and vasogenic edema from dysfunction of Krebs cycle and pentose phosphate pathway. The accumulation of lactate, alanine, and glutamate causes lower intracellular pH and disruption of cellular homeostasis. Highly metabolic areas of the brain are implicated first.^{1,5} Treatment of possible acute disseminated encephalomyelitis with high dose Solu-Medrol potentially led to hyperglycemia that worsened these metabolic derangements, precipitating her neurological and hemodynamic collapse. She was treated with high dose thiamine but subsequently developed PRES which is a known complication of WE. PRES develops from dysregulation of cerebrovasculature causing vasogenic edema.¹

Thiamine has a good safety profile and, even in high doses, toxicity has not been seen. Monitoring for anaphylaxis is recommended when given intravenously.^{3-5,9,18} The prognosis of appropriately treated WE is variable, although there is no consensus of what appropriate treatment is at this time. Full neurological recovery is possible but often some deficits persist such as memory impairment, learning deficits, ataxia, and nystagmus.^{5,18}

4 | CONCLUSION

WE is a devastating syndrome that, if left untreated, can lead to permanent neurologic damage or even death. It is advised to consider WE with any patient who presents with only 1 of the following: ataxia, impaired ocular motor function, history of nutritional deficiency, or altered mental status. If more than 1 of these symptoms is present, there is little harm to begin empiric treatment with high-dose thiamine. Neurological sequelae can be prevented with early recognition and treatment.

CONFLICTS OF INTEREST

None.

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