

CLINICAL VIGNETTE

Acute Wernicke Encephalopathy with Treatment Response

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Introduction

Wernicke encephalopathy is a classically described neurologic manifestation of chronic alcohol toxicity. While physicians often treat with thiamine empirically in the setting of chronic alcohol use or withdrawal, it is rare to see a classic manifestation of the disorder in addition to a rapid response to treatment.

Case

Patient Presentation

A 67-year-old male was brought to the ED by his mother and sister due to their concern about a decrease in his functional level over the past week. They reported significant disorientation over the prior week with difficulty completing his activities of daily living. They also noted frequent falls and instability when walking. Of note, the patient was relatively unconcerned by his condition and only came to the ED to appease his family.

The patient's sister informed the house staff that he had a long alcohol history that involved drinking 12 beers a day along with up to one third of a brandy bottle for 30 years. The patient disputed the daily amount but agreed that he had been drinking consistently for the prior 30 years. He reported that his last drink was the morning of admission.

He acknowledged some differences over the last couple of weeks. He had diplopia when watching TV and also needed to hold onto furniture while walking to avoid falling. This subjective lack of balance was not accompanied by sensations of the room spinning or lightheadedness. He did not report any disorientation but acknowledged a significant memory deficit over the prior six months that has accelerated recently. He reported being unable to recall events from the day prior.

He lives in a back room on his mother's rental property. He retired from his job as a furniture mover 2 years ago and spent most of his days drinking while watching TV. He was unable to walk to the bottom of the hill where the nearest liquor store was located but had been getting rides from the store owners to purchase alcohol.

While his substance abuse history was lengthy, the patient was previously sober for 18 months approximately one year ago

with the first 12 months occurring while in a residential treatment facility. He reported that he had not tried to quit drinking outside of his one episode of sobriety. He did not articulate any specific reasons for drinking other than habit.

Vital Signs and Physical Exam

On presentation, the patient was afebrile and normotensive without tachycardia or tachypnea. He was alert and oriented, lying in bed speaking with the medical staff.

His neurologic exam was notable for inability to abduct either eye past the midline, consistent with a bilateral sixth nerve palsy. He had significant vertical nystagmus with up beating on vertical gaze and down beating on downward gaze. His speech was notable for moderate latency. Visual fields were intact to direct confrontation. All other cranial nerves were intact. He had normal muscle tone and bulk although abnormal reflexes. Patellar reflexes were 4+ bilaterally and triceps, biceps and brachioradialis were all 3+ bilaterally. He had normal sensation and a negative Romberg test. Finger to nose and heel to shin testing were also intact. Gait was wide based with small steps. The patient was unable to complete toe, heel, or tandem walk due instability.

He had poor skin turgor and dry mucous membranes. Cardiovascular, pulmonary, and abdominal exams were unremarkable. Strong distal pulses were noted with warm and well perfused extremities.

The patient's labs on admission were notable for a normal hemoglobin slightly elevated white blood cell count at 11.81 k/microliter. Transaminases were elevated with AST and ALT of 167 U/L and 180 U/ respectively. Lactate levels were elevated on admission at 3.3 mmol/L. Albumin and INR were within normal limits. His urine toxicology screen was negative for addictive substances and serum ethanol levels were undetectable.

Hospital Course

Upon presentation to the emergency department, there was initial concern for Wernicke's encephalopathy given the patient's significant drinking history and classic presentation.

There was also significant concern for potential alcohol withdrawal given his undetectable serum ethanol level.

He was initially treated with intravenous thiamine and normal saline while in the emergency department. Intracranial hemorrhage was thought unlikely based on clinical presentation. While in the ED, CT scan was read, showing lack of acute hemorrhage. MRI was pending and the patient was admitted to a monitored general medicine bed where he received intravenous thiamine 500 mg every 8 hours with maintenance half normal saline. It was thought that his elevated liver enzymes were most likely due to alcohol, though not elevated in the typical two to one ratio of AST over ALT.

His neurologic deficits persisted when admitted to the floor and consisted of a bilateral lateral rectus palsy preventing the patient from abducting either eye past midline. It was felt that the diplopia was a result of other subtle cranial nerve deficits interfering with ocular coordination.

Magnetic resonance imaging of the brain showed subtle symmetric restricted diffusion defects along the posteromedial and medial aspects of both thalami and bilaterally within the hypothalamus. There was also a subtle increase in the T2 signal of the mammillary bodies. There was no intracranial hemorrhage or mass effect.

On exam, the following morning at 6AM, the bilateral rectus palsy was still apparent. However, upon reexamination on rounds 5 hours later, the bilateral rectus palsy had completely resolved with the patient able to abduct both eyes past midline. A mild horizontal bilateral nystagmus was now visible with the ability to abduct eyes bilaterally. He did not have any signs of alcohol withdrawal overnight and did not need treatment with benzodiazepines.

Discussion

Wernicke encephalopathy (Wernicke encephalopathy) is common even if not formally diagnosed, with 1.3% of autopsies worldwide showing signs of the disease.¹ The four cardinal signs include: cerebellar dysfunction, dietary deficiency, abnormal eye movements and alteration in mental status, or memory. To make the diagnosis, 2 of the 4 criteria must be met.¹ It is now recommended that a serum thiamine level be obtained to aid in diagnosis prior to thiamine administration. A retrospective study found that 85% of patients diagnosed with Wernicke encephalopathy had symmetric involvement of the medial thalami and 53% had involvement of the mammillary bodies.² It is an important condition to detect given the benign and effective treatment alongside the potentially severe outcomes, if left untreated.

While Wernicke encephalopathy is usually only thought of in the context of alcohol, other conditions can lead to its presentation. However, they do not always present in manners as classical as the above case. One case report discusses a 48-year-old woman with the combined risk factors of gastric banding and excessive alcohol consumption. She presented with nystagmus and ataxia in the absence of significant

cognitive symptoms.³ She had classic T2-FLAIR hyperintense mammillary body lesions on MRI.

Wernicke encephalopathy has also presented with gait disturbance in the setting of suppressed appetite in cancer patients. One report discussed two patients with metastatic cancer who developed the disease following two weeks of changes in appetite.⁴ Both patients did not have the classic finding of delirium, and were especially notable given the rapid manifestations following changes in appetite.

Contrasting the presentation of Wernicke encephalopathy in the setting of alcoholic and nonalcoholic causes, a large case series found cerebellar signs more likely in alcoholic patients and ocular signs are more common in non alcoholic patients.⁵ Non alcoholic patients also were diagnosed later, pointing to need for increased clinical suspicion in patients with nontraditional risk factors.

Treatment for Wernicke encephalopathy is based on an empirically established regimen of at least 500 mg intravenous thiamine for three days.⁶ A recent case series showed that 73% of patients showed symptom improvement following administration of thiamine.⁶ From clinical practice it is apparent that many clinicians treat in excess of this amount given that there is no data supporting this precise recommendation and the lack of harms of higher doses. Overall, it is important for clinicians to have a high index of suspicion for Wernicke encephalopathy in patients regardless of alcohol history and to initiate treatment without delay.

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