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A Woman with Alcohol Use Disorder, Wernicke-Korsakoff Syndrome, and Central Pontine Myelinolysis: A Case Report and Review of the Literature

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Introduction

We present a case report of a woman with a history of alcohol use disorder who was found to have both Wernicke-Korsakoff Syndrome (WKS) and Central Pontine Myelinolysis (CPM) on MRI. After a discussion of the case, we review the literature of the clinical manifestations, diagnostic criteria, and prognostic indicators of functional outcomes for both WKS and CPM. We compare and contrast the two diseases individually and simultaneously, as occurred in our patient. Central pontine myelinolysis (CPM) is a demyelinating disorder of the brainstem that typically occurs with the rapid correction of serum sodium in hyponatremic patients. Diagnosis of both WKS and CPM requires a high index of suspicion. Though overlapping in some aspects of clinical manifestations, the prognosis and functional outcomes can differ between the two diseases so it is important to distinguish between the two processes.

Neurologic exam revealed horizontal nystagmus, poor concentration and recall, tangential conversation with confabulation, and wide-based gait. Extensive work-up showed no abnormalities in the complete metabolic panel, CBC, TSH, Vitamin B12, HbA1c, ammonia, urinalysis, urine toxicology, and EEG. She tested negative for HIV, Lyme disease, and Syphilis. Lumbar puncture revealed normal protein, glucose, and negative gram stain and culture. CSF was negative for West Nile Virus, HSV, and VZV. The notable findings on work up were: a low thiamine level of 6 nmol/L (normal 8-30 nmol/L), and T2-FLAIR MRI of the brain with atypical white matter signal alteration of the central pons consistent with central pontine myelinolysis.

The constellation of symptoms (nystagmus, confusion, ataxia, and amnesia) in the setting of alcohol use disorder and low thiamine led to a working diagnosis of Wernicke-Korsakoff Syndrome. However, MRI findings in WKS typically reveal abnormalities in the mammary bodies and periventricular regions, not the pons. Central pontine myelinolysis is usually iatrogenic from rapid correction of serum sodium. On further questioning, it was discovered that the patient was hospitalized at an OSH (Outside Hospital) for Legionnaire's Disease six months prior to presentation. It was hypothesized that she suffered a rapid correction of hyponatremia at that time which resulted in mild, unrecognized central pontine myelinolysis. Outside medical records were reviewed, but there was no evidence for hyponatremia to support this hypothesis. On review of the literature, central pontine dysfunction on MRI has been associated with WKS.

Intravenous thiamine was given, and the patient was discharged home with oral thiamine, outpatient cognitive therapy, and alcohol cessation management. On her post-discharge follow-up, she showed improvement in her short-term memory and returned to baseline shortly after discharge, which is seen in only a minority of patients with WKS.

Interestingly, the patient re-presented to the ED six months after hospitalization with a similar presentation of acute onset confusion, this time with a normal thiamine level. Her work-up was unrevealing and MRI was unchanged, revealing stable white matter signal alteration of central pons. She was again admitted to the hospital and treated with IV thiamine for WE with improvement in her mental status during her hospital stay.

When WE is not treated, around 80% of patients will advance to the chronic form of the disease, Korsakoff Syndrome (KS). Clinical manifestations of KS include the classic WE triad as well as amnesia and confabulation. KS generally carries a poorer prognosis compared to WE, as KS is typically irreversible despite treatment with thiamine and alcohol cessation. In contrast to patients with WE, patients with KS rarely recover and will require some form of social support.

Central pontine myelinolysis is a demyelinating disorder of the brainstem and typically occurs with the rapid correction of serum sodium in hyponatremic patients . As with WKS, CPM is also seen in the setting of nutritional and electrolyte stress, including chronic alcohol use disorder and malnourishment . The diagnosis of CPM is based on correlation of clinical findings with radiologic studies, in which an MRI reveals a symmetric area of central pontine demyelination .The treatment of CPM is usually supportive. Interestingly, the clinical presentation of CPM can vary from asymptomatic, to neuropsychiatric, to paralysis, coma, or death. The neurological findings that were seen in our patient include disinhibiting, confusion, impaired cognition, and gait instability. It is important to note that it is difficult to distinguish the mild forms of CPM from WKS as both diseases present with similar neurological findings.

While CPM can vary from incidental finding to a fatal diagnosis, there are only a few studies that examine the prognostic indicators and functional outcomes. A retrospective cohort study of 25 patients with CPM demonstrated that higher GCS (Glasgow Coma Scale) scores ($\geq$ 11), better scores in functional scales, less severe hyponatremia ($[Na^+]$ > 115 mEq/L), and absence of superadded hypokalemia predict favorable outcome . Another article reviews the homeostatic mechanism by which severe acute hyponatremia (serum $[Na^+]$ dropping to < 120 mEq/L in $\leq$ 48-hours) causes a more serious presentation compared to a

slowly progressive chronic hyponatremia . If untreated, severe acute hyponatremia may lead to brain edema, irreversible neurologic damage, respiratory arrest, brainstem herniation, and even death. In contrast, in chronic or less severe hyponatremia, symptoms usually do not arise until the serum [Na^{+}] falls below 120 mEq/L and they are initially nonspecific (e.g., headache, lethargy, nausea) .Ultimately, however, the rate of sodium correction is more important than the nadir sodium concentration in the development of CPM. In addition to treating the patient for WKS with thiamine, CPM in the setting of WKS also necessitates assessment of the patient's functional status. Only a few small studies exist that examine the predictors of disease severity and functional outcomes in WKS and CPM. There is very little literature about patients with both concurrent WKS and CPM as in our patient. This case is important because it highlights the subtle and overlapping differences between WKS and CPM from a clinical, diagnostic, and outcomes perspective.

Keywords: Alcohol use disorder; Wernicke-Korsakoff Syndrome; Central Pontine Myelinolysis; Encephalopathy; Hypothyroidism