It is illegal to post this copyrighted PDF on any website. Got 2 Good Eyes But You Still Can't See: An Atypical Case of Wernicke's Encephalopathy

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ernicke's encephalopathy was first described in 1881 as encephalopathy, ophthalmoplegia, and gait ataxia.¹ Thiamine deficiency was later identified as the underlying cause.² Today, Wernicke's encephalopathy is most commonly identified in patients with alcohol use disorder, though eating disorders are also implicated.^{3,4} We present a case of atypical Wernicke's encephalopathy in a woman with avoidant/restrictive food intake disorder (ARFID) presenting with bilateral ophthalmoplegia and normal serum thiamine levels.

Case Report

A 24-old-woman was brought to the hospital for acute onset of bilateral blurry vision. The deficit began 6 days prior to admission when she noticed worsening eyesight over hours. She was seen by an optometrist on the day of presentation, who urged her to seek care in the emergency department.

Information obtained later in her course revealed that she had a 70-lb weight loss over the past year after restricting her eating and a diagnosis of ARFID (*DSM-5* criteria). She had presented to the emergency department multiple times over the past 2 years in the setting of starvation related to abdominal pain. Previous workup with abdominal film, computed tomography, esophagogastroduodenoscopy, ultrasound, and barium swallow had been unremarkable, leading to psychiatric evaluation and diagnosis of ARFID.

On presentation, her pulse was elevated at 113 bpm. She was oriented and attentive. The ophthalmologic examination was notable for near total restriction of all eye movement without nystagmus. She was blind in her right eye and had 20/800 vision in her left eye. Both eyes had normal ocular pressure, with edema and well perfused vessels. The

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neurologic examination was revealing for hyperalgesia over her lower extremities without gait ataxia.

Blood chemistry showed a sodium level of 147 mmol/L and potassium level of 2.9 mmol/L, and blood count revealed a hemoglobin level of 10.3 g/dL. Renal and thyroid function and albumin, liver enzyme, copper, vitamin E, niacin, vitamin B_{12} (600 pg/mL), and serum thiamine (158.4 nmol/L) levels were all within normal limits. Toxicology was negative. Vitamin A (12.8 µg/dL), vitamin D (22 ng/mL), and folate (5.4 ng/mL) levels were low. Magnetic resonance imaging (MRI) of the brain and orbits was normal, but fundus imaging showed optic disc swelling with peripapillary hemorrhages (Figure 1). Cerebrospinal fluid studies, including venereal disease research laboratory, lactate, and cytology, were normal. Paraneoplastic panel, ganglioside antibody, anti-GQ1b, and HIV test results were negative, and angiotensin-converting enzyme level was normal.

Despite a normal serum thiamine level on initial evaluation, the patient's history of ARFID, significant weight loss, and evidence of ophthalmoplegia led to empiric treatment with 500 mg of intravenous thiamine every 8 hours for 5 doses followed by 100 mg daily. Within 24 hours, her vision returned to 20/20 and her bilateral ophthalmoplegia resolved. She required prolonged hospitalization after medical stabilization to facilitate transfer to an inpatient eating disorder facility to treat ARFID.

Since her discharge, she had 1 additional admission requiring a gastrostomy tube for starvation and has since been seen in outpatient therapy for ARFID. She has had no return of return of Wernicke's encephalopathy on maintenance thiamine 100 mg daily by gastrostomy tube.

Discussion

Wernicke's encephalopathy is an acute life-threatening condition caused by thiamine deficiency that classically presents with the triad of encephalopathy, ophthalmoplegia, and gait ataxia.⁵ However, these 3 symptoms occur together in less than one-quarter of cases.⁶ Causes include alcohol use, nutritional deficiencies, and impaired gastrointestinal absorption.⁷ Most often, Wernicke's encephalopathy is a clinical diagnosis and requires 2 of the following 4 signs: (1) dietary deficiency, (2) oculomotor abnormalities, (3) cerebellar dysfunction, and (4) altered mental state.⁸

This report represents the first published case, to our knowledge, of Wernicke's encephalopathy in ARFID.⁹ Although Wernicke's encephalopathy has been seen in eating disorders, the majority of these cases have presented with

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Figure 1. Fundus Imaging Showing Bilateral Optic Disc Swelling With Peripapillary Hemorrhages



the classic clinical triad.⁴ Our patient presented with isolated bilateral ophthalmoplegia and optic disc swelling, a less recognized manifestation of Wernicke's encephalopathy.¹⁰ This case also demonstrates that serum thiamine levels may not reflect brain levels of thiamine.¹¹ Despite lacking many of the classic laboratory, imaging, or clinical sings, our patient

met 2 of the 4 Caine criteria⁸ for Wernicke's encephalopathy and responded clinically to empiric thiamine within 24 hours. Finally, this case highlights that clinical suspicion of Wernicke's encephalopathy in all at-risk patients warrants immediate administration of high-dose intravenous thiamine.^{12,13}

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has been de-identified to protect anonymity.

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