

Case Report

Celiac disease in a teenager revealed with Wernicke Encephalopathy

Amira Bouayad^{1,2*}, Chaimae Khodriss^{1,2}, Sihame Tantani³,
Hicham Fadel⁴, Siham Alaoui Rachidi^{1,5} and Meriem
Elbahloul^{1,2}

¹Faculty of Medicine and Pharmacy Tangier, Abdelmalek Essaâdi University, Tangier, Morocco

²Department of Ophthalmology, Mohammed 6 University Hospital of Tangier, Morocco

³Department of Pediatrics, Mohammed 5 Hospital of Tangier, Morocco

⁴Department of Neurology, Al Kortobi Hospital of Tangier, Morocco

⁵Department of Radiology, University Hospital of Tangier, Morocco

Abstract

Background: Wernicke encephalopathy is an acute neurological condition defined by a clinical triad of ophthalmoplegia, ataxia, and confusion. This disease is due to thiamine deficiency.

Case presentation: After persistent diarrhea and vomiting, a 15-year-old boy presented up-beating nystagmus, with binocular vertical diplopia and unstable gait. An etiological workup revealed a celiac disease. Magnetic resonance imaging showed bilateral periaqueductal region lesions.

Treatment and discussion: Because of suspected Wernicke's encephalopathy, the patient was treated with thiamine replacement and significant improvement of symptoms took place. Wernicke encephalopathy is a serious medical disorder with enormous morbidity and mortality. Evaluation should include patient history with a physical and ophthalmologic examination and laboratory workup with appropriate imaging.

Conclusion: Undiagnosed celiac disease can lead to malabsorption of vitamin B1 causing acute symptoms of Wernicke encephalopathy.

Introduction

Wernicke encephalopathy (WE) is a neurological emergency resulting from thiamine deficiency. It presents with varied neurocognitive manifestations, typically involving mental status changes, gait, and oculomotor dysfunction [1]. The prevalence of WE in the general population is approximated from 0.4% to 2.8% [2]. And it carries an approximated 20% mortality [1].

It is a disease that is usually, but not entirely seen in those with persistent alcohol excess [1]. Among the other causes, celiac disease is an immune-mediated situation characterized by small intestinal enteropathy and systemic symptoms due to malabsorption and/or immune activation [3].

Successful management of this disease requires early identification and efficient medical interventions to prevent the high morbidity and mortality associated with this disease. Hence, we hereby report a case of celiac disease recognized by Wernicke encephalopathy.

Case report

We report the case of a teenager with Wernicke encephalopathy complicating celiac disease.

A 15-year-old boy presented to the emergency department with a 3-day history of binocular vertical diplopia and unstable gait. His previous ocular history was unremarkable. His medical history was significant for treating *Helicobacter pylori* gastritis 'with infection eradication therapy' and uncontrollable vomiting and diarrhea. There is no history of toxic habits that were mentioned. His family medical and ocular histories were noncontributory.

Upon ocular examination, his visual acuity(VA) measured 20/20 in both eyes, ocular motility examination revealed small amplitude and moderate frequency up-beating nystagmus that increased slightly on lateral gaze, on up gaze, and on downgaze and a bilateral sixth cranial nerve palsy (Figure 1). Pupillary light reflexes were brisk with no relative afferent

More Information

*Address for correspondence: Amira Bouayad, Faculty of Medicine and Pharmacy Tangier, Abdelmalek Essaâdi Department of Ophthalmology, Mohammed 6 University Hospital of Tangier, Morocco, Email: Bouayad2228@gmail.com

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Figure 1: Ocular motility examination revealing bilateral sixth cranial nerve palsy.

pupil defect. Biomicroscopy found normal anterior segment structures. Dilated fundus evaluation was normal, including both optic nerve appearances.

A Lancaster Test confirmed the bilateral sixth cranial nerve palsy, classified as stage 3 (Figure 2).

On general examination, the patient was apyrexia, asthenic, and anorexic, and had a normal heart rate, and hypotension of 100/60mmHg. He had puberty delay and epigastric tenderness.

Neurological examination revealed an ataxic gait, tendon reflexes, and muscle strength were normal. There were no signs of confusion or delirium. There were no focal neurological signs, but a slight decrease in proprioceptive sensation especially vibratory sensitivity in lower limbs. The remaining cranial nerves were intact.

An etiologic work-up, which included an upper-gastrointestinal endoscopy with biopsy that revealed chronic gastritis and celiac disease.

Brain magnetic resonance imaging shows a defined T2 hyperintense area in the periaqueductal region (Figure 3).

A lumbar puncture was performed with pressure measurement and it was normal.

Laboratory investigations, including complete blood count, erythrocyte sedimentation rate, C reactive protein, fasting blood sugar, and kidney function tests were all within the normal range. A serum electrolyte typically shows hyponatremia. Furthermore, a high value of antitransglutaminase antibodies (AntiTG IgA and AntiTG IgG) was disclosed.

We have concluded that unknown celiac disease was liable for Wernicke encephalopathy.

The severity of the disease made that the supplementation should be anticipated without waiting for the thiamine results. However, the patient refuses any parenteral form. Therefore, he was given oral Thiamine 200mg, three times/day.

The patient was also followed in the gastro-enterology pediatric department for his celiac disease.

Within two weeks, diplopia, and lateral rectus palsies began to improve, his ataxic gait disappeared completely but nystagmus remains persistent (Figure 4). Whole-blood thiamine level, repeated after supplementation, was normal (83nmol/L versus 41nmol/L before).

He remains well on follow-up at 1 month, with a normal Lancaster test (Figure 5).

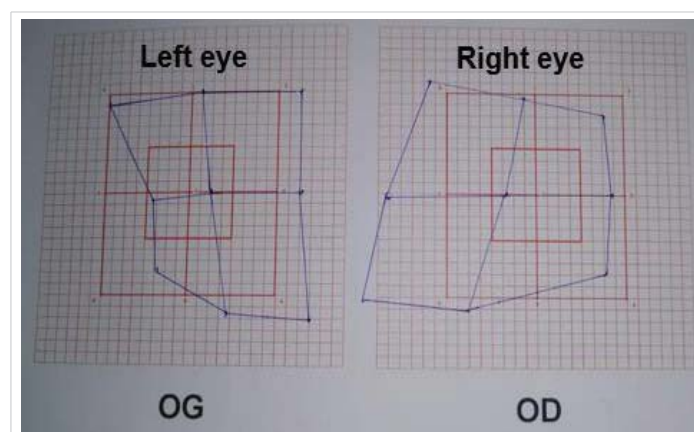


Figure 2: Lancaster test showing the bilateral sixth cranial nerve palsy.

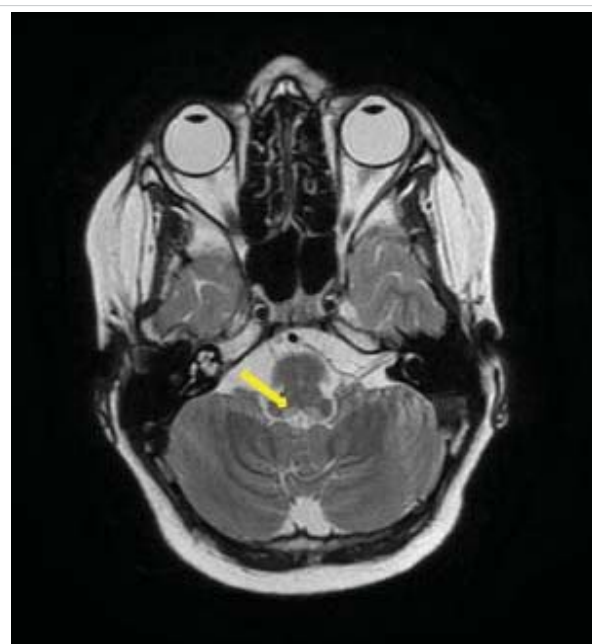


Figure 3: Axial T2-weighted magnetic resonance image with hyperintense areas in the periaqueductal region (yellow arrow).



Figure 4: After treatment, the examination shows normal ocular motility.

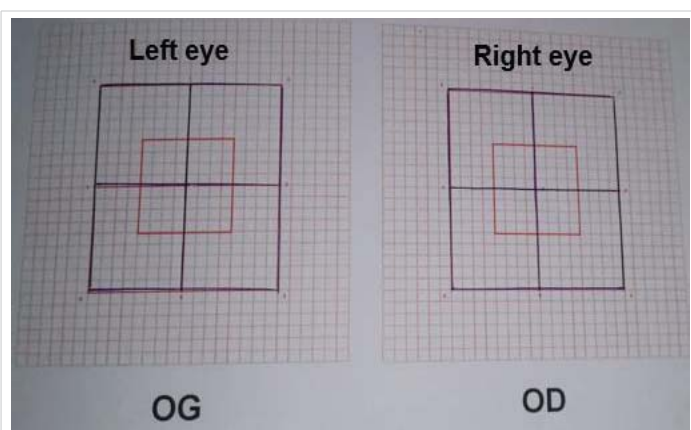


Figure 5: A normal Lancaster test on follow-up.

Discussion

Wernicke's encephalopathy is a result of thiamine deficiency, mostly associated with chronic alcoholism. It can also be included in other causes, such as the surgical treatment of gastrointestinal diseases [4], hyperemesis gravidarum [5], acquired immunodeficiency syndrome (AIDS) [1], Crohn's disease [4], anorexia nervosa [1], malnutrition and unbalanced diets [4].

In our case, uncontrollable vomiting and signs of celiac disease caused vitamin depletion in the patient.

Typically, the syndrome comprises changes in mental state, gait ataxia, and ophthalmoplegia [6]. However, the full triad has been rarely described [6]. In fact, the major ophthalmological signs in WE comprise abduction paresis, horizontal and vertical gaze palsy, internuclear ophthalmoplegia, horizontal and vertical nystagmus, and optic neuropathy [5]. Our patient presented with bilateral abduction palsy, up-beating nystagmus, and gait ataxia.

The brain MRI has outcomes in diagnosis and follow-up examinations to monitor prognosis. Particular damage in the thalamus, especially medial and intralaminar nuclei, inferior colliculus, and other vulnerable areas has been discerned, where thiamine is processed [4].

In the present case, the periaqueductal region was abnormal which was correlated with the clinical findings.

WE remains a clinical diagnosis and a medical emergency, that should be treated immediately. Treatment should not wait or be based on laboratory or imaging results [1]. That being said, if treated correctly ophthalmological signs begin to improve gradually. Nystagmus and ocular motility abnormalities can be recovered in 1-2 weeks. However, gait disorders may endure in up to 60% and just 20% of cases have entirely memory rehabilitation [7].

Conclusion

Bilateral Ocular motility disorders, gait instability, and nystagmus should raise the suspicion of WE, especially when there is a predisposition. Thiamine dosage and brain MRI help with diagnosis. Nevertheless, immediate treatment is crucial and can prevent sequelae.

Authors' contributions

All the authors contribute to the management of this case and the redaction of this manuscript. All the authors have given their approval to the final version of the manuscript.

Ethical considerations

The authors declare that they have the patient's consent to appear in a journal article.

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