Case Report

Visual loss and optic neuropathy associated with Wernicke's encephalopathy in hyperemesis gravidarum

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Abstract

Wernicke's encephalopathy (WE) is a potentially fatal neuropsychiatric syndrome precipitated by thiamine deficiency due to a variety of causes such as chronic severe alcoholism, starvation, and prolonged intravenous feeding. WE has also been observed rarely in the clinical setting of hyperemesis gravidarum. Here, we report the case of a 34-year-old pregnant woman who presented with reduced vision, gaze-evoked nystagmus, and postural imbalance preceded by 2 weeks of hyperemesis. Fundus examination showed features consistent with papillitis. Magnetic resonance imaging (MRI) showed T2WI, FLAIR, and diffusion-weighted imaging MR images showing hyperintensity in dorsomedial thalami and periaqueductal grey matter with diffusion restriction. She was diagnosed with WE based on history, clinical examination findings, and MRI findings and was treated with injectable thiamine. She showed marked improvement in vision and nystagmus within 3 days. Our case is a rare presentation of WE in a pregnant woman with hyperemesis gravidarum.

Keywords: Hyperemesis gravidarum, optic neuropathy, pregnancy, Wernicke's encephalopathy

Introduction

Thiamine (vitamin B_1) is an essential nutrient that plays a significant role in glucose metabolism.[1] Wernicke's encephalopathy (WE) results from thiamine deficiency due to various causes such as heavy alcohol consumption, gastrointestinal disorders and surgeries (i.e. gastric bypass), acquired immune deficiency syndrome, hemodialysis, malignancies, other systemic diseases, anorexia nervosa or other psychiatric conditions, infections, shock, prolonged nutritional deficiencies, or the administration of intravenous glucose prior to thiamine in malnourished or alcohol-dependent individuals.[2] Moreover, WE has also been observed rarely in the clinical setting of hyperemesis gravidarum (HG).[3] WE clinically manifests as nystagmus and ophthalmoplegia, mental status changes, and unsteadiness of gait and carries a high rate of morbidity and mortality when goes unrecognized.[4] Optic neuropathy with visual loss is a rare manifestation of WE. We report a case of visual loss with optic neuritis in a pregnant woman with HG.

Case Report

A 34-year-old pregnant woman (G3P2L2) was admitted with reduced vision preceded by 2 weeks of hyperemesis and decreased food intake. She had no history of seizures, loss of consciousness, headache, or fever. She was conscious and afebrile. Her pulse rate was 92/min and blood pressure was 110/60 mmHg. Her gait was ataxic, and ocular examination revealed gaze-evoked nystagmus. Her vision was light perception (right eye) and finger counting (left eye). There was no neck stiffness. Examination of fundus showed features consistent with papillitis in both

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Access this article online

Quick Response Code:

Website: www.jfmpc.com

DOI: 10.4103/jfmpc.jfmpc_121_19

How to cite this article: Palakkuzhiyil N, Rehiman S, Manoj PP, Hameed S, Uvais NA. Visual loss and optic neuropathy associated with Wernicke's encephalopathy in hyperemesis gravidarum. J Family Med Prim Care 2019;8:1243-5.

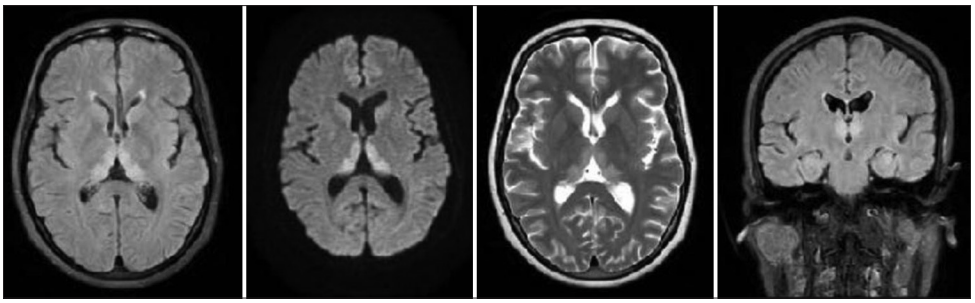

Figure 1: T2WI, FLAIR and DWI MR images showing hyperintensity in dorsomedial thalami and periaqueductal grey matter with diffusion restriction

eyes (hyperemia and swelling of the disc with blurred disc margins and distended veins). Magnetic resonance imaging (MRI) showed T2WI, FLAIR, and diffusion-weighted imaging MR images showing hyperintensity in dorsomedial thalami and periaqueductal grey matter with diffusion restriction [Figure 1]. The diagnosis of WE was made based on her clinical presentation and radiological findings. She was treated with high-dose intravenous thiamine injection (500 mg 8 hourly). Her vision improved within 3 days and nystagmus was resolved. At the time of discharge, she was much better, but ataxia persisted.

Discussion

Thiamine (vitamin B_1) is a water-soluble vitamin found naturally in animal and plant products and its requirement for the body is directly related to total caloric intake.[5] However, the average daily requirement of thiamine is around 1.5 mg. and its demands is increased during hyper metabolic states such as pregnancy.[5,6] There are more than 40 cases of WE associated with HG described in the published literature.[3]

Our patient suffered optic neuropathy, visual loss, and ataxia following HG, and the diagnosis of WE was made based on clinical history, ocular, and neurological findings including fundoscopy, characteristic MRI findings, and a marked improvement in clinical manifestations with thiamine administration. Our case did not show features suggestive of confusion. However, a necropsy study of 131 patients with WE showed that only 16% of the cases exhibited all three features at presentation.[7] The common ocular manifestations of WE reported in the literature are nystagmus, ophthalmoplegia, conjugate gaze palsy, and amaurosis. Optic neuropathy and papilloedema have also been reported infrequently in patients with WE.[8] Chitra and Lath reported the case of a 25-year-old lady in her 20th week of her pregnancy presenting with a visual acuity of 6/60 in both eyes; and abduction restriction, nystagmus, retinal hemorrhages, and macular edema in both eyes following nausea and vomiting for 3 months.[9]

The diagnosis of WE is mainly clinical, and a low thiamine level in blood may not be present in all cases. The erythrocyte thiamine diphosphate level assessed by high-pressure liquid chromatography is found to be a better marker of thiamine deficiency.[10] Furthermore, the characteristic MRI findings in our case of hyperintensity on FLAIR and T2-weighted images in the periaqueductal area have high specificity and moderate sensitivity in the diagnosis of WE.[2]

Once a diagnosis of WE is made clinically, thiamine replacement should be started immediately to prevent significant cognitive consequences of untreated WE. The traditional recommendation is a parenteral dosage of 100 mg of thiamine per day.[11] However, there are other treatment protocols also such as 500 mg of thiamine hydrochloride infused over 30 min, three times a day for 2–3 days, and an additional 250 mg of thiamine given intravenously or intramuscularly daily for 3–5 days, or until resolution of symptoms, which was also found to be effective in the treatment of WE.[12]

Conclusion

Visual loss due to optic neuropathy is a relatively rare clinical picture of WE. Clinicians should bear in mind this clinical presentation while evaluating patients with HG as prompt recognition, and treatment of WE results in good prognosis.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

References

1. Osiezagha K, Ali S, Freeman C, Barker NC, Jabeen S, Maitra S, *et al.* Thiamine deficiency and delirium. Innov Clin Neurosci 2013;10:26-32.
2. Flynn A, Macaluso M, D'Empaire I, Troutman MM. Wernicke's encephalopathy: Increasing clinician awareness

of this serious, enigmatic, yet treatable disease. Prim Care Companion CNS Disord 2015;17:10.4088.

3. Ashraf VV, Prijesh J, Praveenkumar R, Saifudheen K. Wernicke's encephalopathy due to hyperemesis gravidarum: Clinical and magnetic resonance imaging characteristics. J Postgrad Med 2016;62:260-3.
4. Kantor S, Prakash S, Chandwani J, Gokhale A, Sarma K, Albahrani MJ. Wernicke's encephalopathy following hyperemesis gravidarum. Indian J Crit Care Med 2014;18:164-6.
5. Gratton SM, Lam BL. Visual loss and optic nerve head swelling in thiamine deficiency without prolonged dietary deficiency. Clin Ophthalmol 2014;8:1021-4.
6. Carrodeguas L, Kaidar-Person O, Szomstein S, Antozzi P, Rosenthal R. Preoperative thiamine deficiency in obese population undergoing laparoscopic bariatric surgery. Surg Obes Relat Dis 2005;1:517-22; discussion 522.
7. Harper CG, Giles M, Finlay-Jones R. Clinical signs in the Wernicke-Korsakoff complex: A retrospective analysis of 131 cases diagnosed at necropsy. J Neurol Neurosurg Psychiatry 1986;49:341-5.
8. De Wardener HE, Lennox B. Cerebral beriberi (Wernicke's encephalopathy). Review of 52 cases in a Singapore prisoner-of-war hospital. Lancet 1947;i: 11-7.
9. Chitra S, Lath KV. Wernicke's encephalopathy with visual loss in a patient with hyperemesis gravidarum. J Assoc Physicians India 2012;60:53-6.
10. Ferdinands. MD, Seneviratne. J, White O, Visual deterioration in hyperemesis gravidarum. Med J Aust 2005;182:583-4.
11. Donnino MW, Vega J, Miller J, Wlsh M. Myths and misconceptions of Wernicke's encephalopathy: what every emergency physician should know. Ann Emerg Med 2007;50:715-21.
12. Hope LC, Cook CCH, Thomson AD: A survey of the current clinical practice of psychiatrists and accident and emergency specialists in the United Kingdom concerning vitamin supplementation for chronic alcohol misusers. Alcohol Alcohol 1999;34:862-7.