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Case report

Acute up-beating nystagmus in a pregnant woman with hyperemesis gravidarum

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ABSTRACT

Purpose: To describe a case of sudden onset of nystagmus in a pregnant patient with hyperemesis gravidarum.
Observations: Sixteen days after onset of persistent nausea and uncontrollable vomiting, a 12 week pregnant woman presented up-beating nystagmus, mild memory impairment and reduced sensitivity in the lower limbs. Laboratory tests presented thiamine deficiency and magnetic resonance imaging showed bilateral medial thalami and midbrain lesions. Because of suspected Wernicke's encephalopathy, the patient was treated with thiamine replacement and significant improvement of symptoms took place.
Conclusions and importance: Uncontrollable vomiting can lead to malabsorption of vitamin B1 causing acute onset of nystagmus.

1. Introduction

Up-beating nystagmus (UBN) is an uncommon clinical finding observed in several neurological disorders associated with lesion affecting the brainstem,[1] especially caudal pons, rostral medulla, pontomesencephalic junction[2] and thalamic area.[3] Wernicke's encephalopathy is a rare neurological disorder caused by thiamine deficiency. Classically, patients present a triad of ataxia, confusion and ocular signs.[4] Neuro-ophthalmologic findings include abducens paresis, horizontal and vertical gaze-evoked nystagmus, progressive vertical and horizontal gaze palsies, internuclear ophthalmoplegia and impaired vestibulo-ocular response.[4,5] A case is reported here in which UBN was the predominant manifestation of Wernicke's encephalopathy (WE).

2. Case report

A 23-year-old female has developed “dizziness”, weakness and tingling on both legs, for the last two weeks. She was 10 weeks pregnant when incoercible vomiting and nausea started. When she was admitted in the emergency room two weeks later, she told the neurology staff that she had difficulty to remember things and recent events. She also said that she had lost weight (6 kg) in the past two weeks.

Although she reported mild short-term memory loss, she was completely able to name, date, place and situate, during the neurological examination. It was also detected weakness on both lower limbs, without focal neurological signs and ataxia, and slight sensitivity loss in light touch.

Her visual acuity (VA) measured 20/20 in both eyes. Pupil reflexes were brisk and consensual with no relative afferent pupil defect. Slit lamp exam and ophthalmoscopy were normal, including both optic nerves appearance. External examination revealed small amplitude and moderate frequency up-beating nystagmus on primary gaze that increased slightly on lateral gaze, increased markedly on upgaze and decreased significantly in amplitude and frequency on downgaze (Video 1). Cover and uncover tests have not showed constant strabismus. The confrontation visual field test was normal.

Supplementary video related to this article can be found at http://dx.doi.org/10.1016/j.ajoc.2017.01.008.

Cerebral spinal fluid, blood cell count and capillary blood glucose, C reactive protein and erythrocyte sedimentation rate were normal. Mild hypokalemia with K^+ of 3,2mmmol/l (normal 3,5-5,0mmmol/l) and hyponatremia with Na^+ of 130mmmol/l

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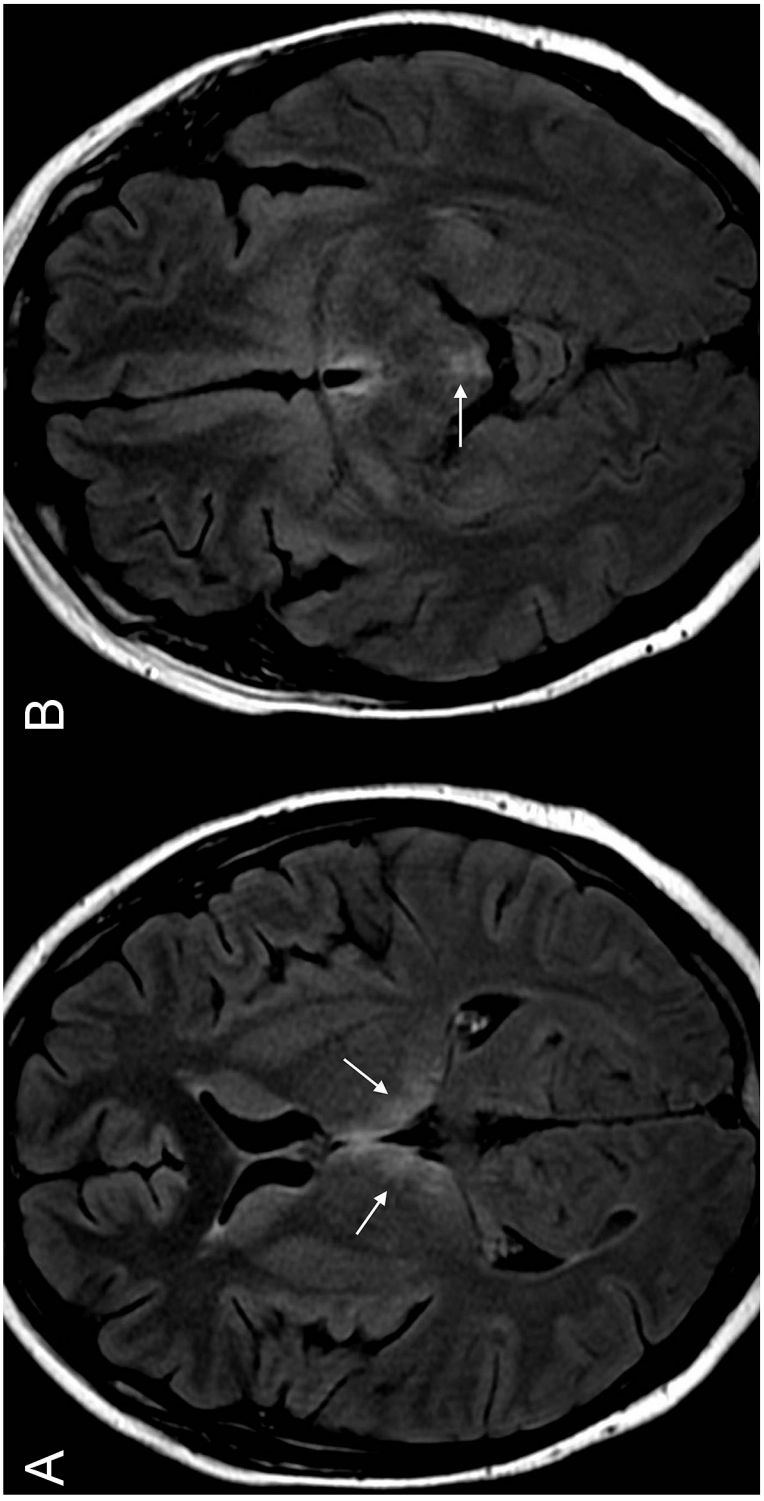

Fig. 1. Axial FLAIR (fluid attenuation inversion recovery) depicting increased signal intensity in the medial part of the thalami (A) and periaqueductal white matter (B).

(normal 135–145mmol/l) were detected in metabolic evaluation. Due to vomiting and electrolyte loss, the diagnosis of hyperemesis gravidarum was confirmed. Brain MRI to investigate the origin of nystagmus showed high signal intensity on FLAIR images at the thalamus and brainstem - midbrain, periaqueductal - (Fig. 1), which raised suspicion of Wernicke's encephalopathy (WE). Then, the thiamine levels were requested and her initial thiamine level was found to be 28nmol/l (normal 60–120nmol/l) confirming the WE.

She was treated with 500 mg thiamine IV during one week at the hospital and had a progressive improvement of neurological and ocular signs. After hospital discharge, she received 300 mg of oral thiamine for three weeks and at follow-up four weeks later the patient reported improvement of oscillopsia, but a mild degree of nystagmus continued (Video 2 and 3), which almost disappeared two months after her hospitalization. Also, both her optic disks were normal. Her thiamine levels increased and reached normal value.

3. Discussion

Initially described in 1881, Wernicke's encephalopathy consists of a triad of ataxia, mental confusion and ocular motor disfunction.[4] It is worth noting that the three symptoms are present, concurrently, in only 16% of patients which could make the diagnosis undisclosed. On the other hand, if precise and early diagnosis is made, the disease can be easily treated through thiamine replacement with prompt improvement of symptoms, otherwise it may occur progression to Korsakoff syndrome with irreversible brain damage.[6]

Thiamine, known as vitamin B1, is an essential molecule involved in several metabolic steps of chain energy cell production.[7] Thiamine deficiency demonstrated to affect brain structures with high thiamine turnover rate as cerebellum, medulla, pons, hypothalamus, midbrain and thalamic area.[8] These regions are involved in the mechanism of controlling eye movements and steady fixation which are committed during Wernicke's encephalopathy.[9] Malnutrition is the main cause of thiamine deficiency as in cases of chronic alcoholism, after bariatric surgery and anorexia. In our case, uncontrollable vomiting caused vitamin depletion of the patient. Since body storage of thiamine lies between 30 and 50mg, reservations may become extinct as early as four to six weeks.[10]

The main ophthalmological signs in WE include abduction paresis, horizontal and vertical gaze palsy, internuclear ophthalmoplegia, horizontal and vertical nystagmus and optic neuropathy.[4] In this case, the patient presented as key sign of WE an up-beating nystagmus. Damage of tegmental area of midbrain, as well as injuries in the medial region of thalamus caused UBN[11] (Fig. 1). Both lesions were shown in the MRI and correlated with the clinical findings.

Wernicke's syndrome is a medical emergency and should be treated quickly. Untreated, WE caused death in up to 20% of cases.[12] On the other hand, if treated properly, ophthalmological signs begin to improve quickly. Nystagmus and eye movements' defects can be recovered in 1–2 weeks. However, gait disorders may persist in up to 60% and just 20% of patients have complete memory recovery.[12]

4. Conclusion

Ocular motility disorders, gait instability and mental confusion should raise the suspicion of WE. Thiamine dosage and MRI aid in the diagnosis. Prompt replacement must be performed to avoid sequelae.

5. Patient consent

The patient consented to publication of the case in writing.

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Conflict of interest

The following authors have no financial disclosures: K. H., F.G, D.B.

Authorship

All authors attest that they meet the current ICMJE criteria for Authorship.

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