Nystagmus in non-alcoholic Wernicke encephalopathy

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DESCRIPTION

A 46-year-old woman with no history of alcohol use presented with blurry vision and difficulty walking. She reported that her eyes were unfocused when texting messages on iPhone. She had been eating only ice cream for three months, felt extremely fatigued and lost 3 kg after her mother’s death. She could not keep herself upright and sought medical attention. Her medical and family history was unremarkable. She did not smoke or use illicit drugs.

Her height was 153 cm and weight was 38.8 kg. Her temperature was 37.8°C, blood pressure was 106/73 mm Hg, pulse was regular at 110 beats per minute, respirations were 20 per minute and oxygen saturation was 97% on ambient air. She was alert and oriented to time, place and person; appeared cachexic with masked facies, and was barely responsive to questions. Neurological examination revealed impaired eye movements with horizontal nystagmus, spontaneous upbeat nystagmus and upgaze-evoked spontaneous upbeat nystagmus (video 1). Strength and sensation to light touch were intact. Deep tendon reflexes were symmetrical. Cerebellar dysmetria were absent. Romberg testing could not be performed. Physical examination was otherwise unremarkable.

Laboratory studies showed normal electrolyte levels, kidney and liver function, total protein, albumin and haemoglobin A1c. Complete blood count showed mild leucocytosis and increased haematocrit. Thyroid function tests, serum levels of vitamin B12, folic acid, copper and zinc were normal. Thiamine level was measured. Brain MRI revealed high intensity in bilateral thalami on diffusion-weighted imaging, T2-weighted images and fluid-attenuated inversion recovery (FLAIR) images (figure 1). Ocular dysfunction, gate disturbance and MRI findings suggested Wernicke encephalopathy (WE). Intravenous administration of thiamine ameliorated her symptoms, improved the nystagmus and she became expressive in 2 weeks. The patient was discharged ambulatory on day 19. The vitamin B1 level before the treatment turned out to be low at 17.0 ng/mL (reference range 21.3–81.9).

WE is a neurological disorder and associated with thiamine deficiency. Thiamine, also known as vitamin B1, is a coenzyme for several enzymes in organic pathways and plays a key role in cerebral energy homoeostasis. Deficiency of thiamine causes neurological complications and develops in chronic alcoholism, severe malnutrition, hyperemesis gravidarum, intestinal obstruction, gastrointestinal surgery (bariatric surgery, gastrectomy and colectomy), cancer chemotherapy, haemodialysis and malignant diseases. In this case, eating ice cream exclusively for 3 months may have exacerbated thiamine deficiency because glucose oxidation decreases thiamine levels.

The clinical triad seen in WE includes altered mental status, ophthalmoplegia and gait ataxia, although the presence of all features is uncommon. Non-alcoholic WE may demonstrate a more acute presentation than alcoholic WE, and ocular dysfunction tends to be the first presentation in non-alcoholic WE.

Horizontal gaze-evoked nystagmus tends to be the first presentation in non-alcoholic WE. In chronic alcoholism, severe malnutrition, hyperemesis gravidarum, intestinal obstruction, gastrointestinal surgery (bariatric surgery, gastrectomy and colectomy), cancer chemotherapy, haemodialysis and malignant diseases.

Learning points

- Wernicke encephalopathy (WE) develops in chronic alcoholism, severe malnutrition, hyperemesis gravidarum, intestinal obstruction, gastrointestinal surgery (bariatric surgery, gastrectomy and colectomy), cancer chemotherapy, haemodialysis and malignant diseases.
- Non-alcoholic WE may demonstrate a more acute presentation than alcoholic WE, and ocular dysfunction tends to be the first presentation in non-alcoholic WE.
- Characteristic features of MRI include increased intensity of bilateral medial thalami, the mammillary bodies, tectal plate, periaqueductal area and around the third ventricle on T2 and fluid-attenuated inversion recovery images.
nystagmus is the typical ocular manifestation of WE, and vertical gaze-evoked nystagmus is less common.6

MRI is helpful in diagnosing WE. Characteristic features of MRI include increased intensity of bilateral medial thalami, the mammillary bodies, tectal plate, periaqueductal area and around the third ventricle on T2 and FLAIR images.2

WE is a medical emergency and under-recognised especially in non-alcoholic patients. Prolonged neurological impairment such as Korsakoff syndrome could be prevented with thiamine.1

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