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CASE REPORT Open Access

A case of Gayet-Wernicke encephalopathy with misleading symptoms

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Abstract

Background: The dramatic evolution of incoercible vomiting in pregnancy is at the origin of a rare and serious neurological complication known as EGW; its symptomatology is polymorphic posing a diagnostic problem. MRI is the test of choice for screening. Early vitamin supplementation is essential to improve the maternal-fetal prognosis.

Case presentation: A 35-year-old parturient, G5P1, without particular pathological history presented incoercible vomiting at 8 weeks, which led to dehydration, undernutrition, and impairment of general condition. Neurological signs: headaches, confused scored at 14 on the Glasgow scale, amnesia, a cerebellar syndrome, sensory-motor deficit. Ophthalmological examination revealed: edema in the eye fundus, a decrease in visual acuity, nystagmus, and ophthalmoplegia. The biological analysis showed moderate hepatic cytolysis.
Given the polymorphic neurological symptomatology presented by our parturient, several differential diagnoses were evoked: intracranial hypertension (headache - vomiting - decrease in visual acuity - papillary edema), brain stroke (sensory-motor deficit, ground "pregnancy"), deficit in vitamin B12 (sensory-motor deficit, confusion, memory impairment), and GWE (nystagmus-confusion-ataxia). Cerebral MRI has allowed us to establish the diagnosis of GWE.

Conclusion: All pregnant with severe hyperemesis gravidarum should be supplemented with thiamine prior to glucose administration to prevent and improve the prognosis of GWE.

Keywords: Gayet-Wernicke encephalopathy, Hyperemesis gravidarum, Thiamine, Magnetic resonance imagery

Background

Hyperemesis gravidarum is the most severe form of nausea and vomiting in pregnancy, and it can be complicated by Gayet-Wernicke encephalopathy (GWE) due to low thiamine reserves, as well as increased needs (Goodwin 2008). GWE is characterized by classical triad encephalopathy, ophthalmoplegia, and/or nystagmus with ataxia. Treatment is mainly based on hospitalization, vitamin therapy, intravenous hydro-electrolytic disorder correction, and antiemetic (Eboué et al. 2006).

We report a case of Gayet-Wernicke encephalopathy where the clinical diagnosis is difficult in the face of rich and heterogeneous symptomatology and discuss the diagnostic difficulties, treatment modalities, and prognosis of such pathology.

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Case presentation

It is about the parturient R. N, young adult, G5P1, with a progressive pregnancy at 14 weeks of amenorrhea, without particular pathological history, was admitted to our department for incoercible vomiting with impairment of the general state. The onset of symptomatology seems to date back to a month and a half marked by the appearance of immediately abundant vomiting, becoming more and more frequent during the day and rebel to the symptomatic treatment. These vomitings were accompanied by dehydration, severe malnutrition with weight loss of 15 kg in 1 month (Buzby index at 78), and a progressive alteration of the general state, hence her hospitalization at our structure in the Gynecological Obstetrics Department of the Bab El Oued University Hospital Center classified as level II.

At admission, we found an asthenia patient, headaches, confused scored at 14 on the Glasgow scale, pale skin-

mucosa, signs of dehydration (skin folds, dry mouth, excavated eyeballs, tachycardia at 124 beats/min), blood pressure at 90/60 mm Hg, polypnea at 24 cycle/min, without edema, and evolving in an apyretic context.

Neurological examination revealed the presence of anterograde amnesia, a cerebellar syndrome with ataxia (movement coordination disorder), a sensory-motor deficit with muscular weakness and paresthesias of the limbs and trunk, and an abolition of osteo-tendinous reflexes (patellar, achillian).

Ophthalmological examination revealed: nystagmus, ophthalmoplegia, stage II edema in the eye fundus, and a decrease in visual acuity to 1/20 ddc.

The bioassay showed anemia at 9.8 g/dl, hypokalemia at 2.58 mmol/L, hyponatremia at 130 mmol/L, hypochloremia at 78.3 mmol/L, hypoproteinemia at 53.64 g/l with hypoalbuminemia at 31.7 g/l, glycemia at 0.87 g/l, and correct renal function. The liver function test: SGOT at 49 IU/L and a slight increase in SGPT at 84 IU/L. Total bilirubin level was 11.3 mg/L, with direct bilirubin at 6.6 g/L, ultra-sensitive TSH collapsed at 0.03 mUI/L with FT4 at 29.92 pmol/l, anti-PTO antibody testing came back negative, and normal cervical ultrasound. Hemostasis test including prothrombin levels at 85% and activated cephalin time at 42" (witness: 33").

Given the polymorphic neurological symptomatology presented by our patient, a cerebral MRI was requested to support the diagnosis, which revealed bilateral and symmetrical pulvinar signal abnormalities presenting as T2 hypersignal and flair, related to Gayet-Wernicke encephalopathy (Fig. 1).

The treatment consisted of a rehydration by the isotonic saline and antiemetic (metoclopramide, ondansétron), potassium supplementation, and the starting of intravenous vitamin B1 treatment at the dose of 1 g/d during 10 days, then relays 500 mg/d orally during the remainder of the pregnancy.

A few hours after the beginning of the specific treatment (vitamin B1), the evolution was marked by the spectacular improvement of the visual disorders; the parturient started to discern shapes and colors again.

After 4 days, the evolution was marked by the disappearance of confusion and ataxia, improvement of the level of the visual acuity to 8/10 at the right and the left eye, and disappearance of the papillary edema at the eye fundus.

The pregnancy was continued until full term, without complications leading to a vaginal delivery of a healthy newborn female baby with a good Apgar (8/10–10/10) and a weight at 2400 g.

After 6 months of evolution, the patient still presented some memory disorders, tingling, a slight visual blur, and moderate ataxia. At 1 year old, Ms RN had presented a good general state, with a weight gain of 12 kg and disappearance of neurological signs.

Discussion

Incoercible vomiting, or hyperemesis gravidarum (HG), complicates 0.3 to 3.6% of pregnancies (Einarson et al. 2013) and is the leading cause of hospitalization in the first trimester of pregnancy. They are associated with significant morbidity and high cost for the society (Gazmararian et al. 2002).

Serious HG can give the following presentation: weight loss ˃ 5%, prolonged vomiting resistant to usual treatment, ketonuria, skinfold, low blood pressure, signs of intra- and extra-dehydration neurological disorders, and biological signs (hyponatremia, hypokalemia, hypochloremia, liver cytolysis) (Niebyl 2010).

Our parturient had therefore presented clinical and biological signs in favor of a state of severe hyperemesis

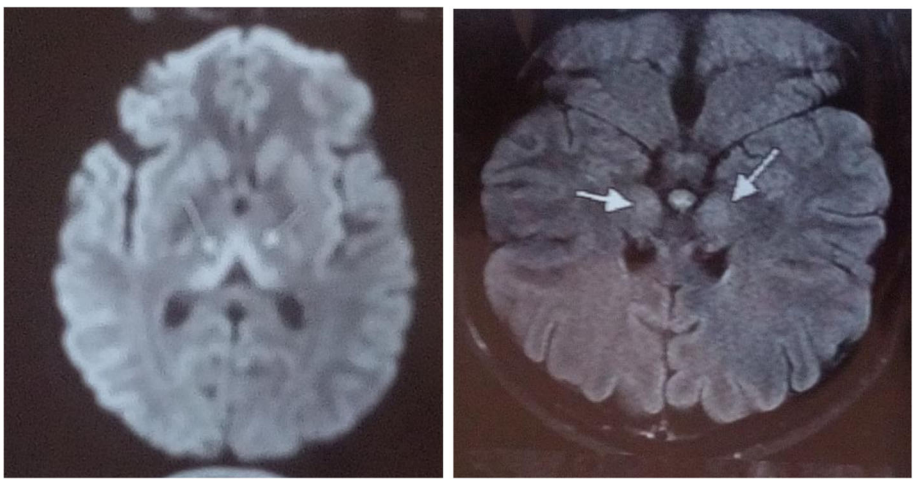

Fig. 1 Image of the brain showing symmetrical hyperintensity in bilateral pulvinar nuclei of the thalamus on the MRI

gravidarum, complicated by disparate neurological symptoms.

Given the polymorphic neurological symptomatology presented by our parturient, several differential diagnoses were evoked:

- Intracranial hypertension: benign, expansive process, front (headache - vomiting - decrease in visual acuity - papillary edema stage II at the eye fundus bottom of the eye).
- Brain stroke: ischemic or hemorrhagic [sensory motor deficit - amnesia - ground "pregnant woman"].
- Deficit in vitamin B12: sensory-motor deficit, confusion, memory impairment...
- Hyperemesis gravidarum: incoercible vomiting causing dehydration, malnutrition, hepatic cytolysis. Complicated by GWE, with the evocative triad (nystagmus-confusion-ataxia).

The diagnosis of Gayet-Wernicke encephalopathy is primarily clinical with the classic triad found in 66% of cases: ocular abnormalities (93% of cases, mainly with nystagmus), confusion (80% of cases), and ataxia (76% of cases). More rarely, we can observe a decrease in osteotendinous reflexes, a decrease in tone or dysarthria (Reuler et al. 1985). The clinical trial was found in our patient associated with sensory-motor's signs and abolition of osteo-tendinous reflexes.

The reference imaging examination to confirm Gayet-Wernicke's diagnosis of encephalopathy is cerebral MRI with a sensitivity of 53% and specificity at 93%. Typical damage affects the floor of the 4th ventricle, around the Sylvius aqueduct, the vermis, the 3rd ventricle, the thalamus (middle parts), the hypothalamus, and the mamillary bodies (Omer et al. 1995).

GWE is a neuropsychiatric syndrome due to a deficiency of thiamine, potentially fatal but preventable; vitamin B1 or thiamine is a water-soluble vitamin belonging to the vitamin B family; and it is a non-caloric organic substance, essential to our metabolism and not produced by our body (Gardian et al. 1999).

Thiamine deficiency leads to brain damage mainly in meso-diencephalic regions rich in dependent thiamine energy processes.

The exhaustion of thiamine may be worsened by hyperthyroidism or by intravenous administration of glucose without vitamins (Gardian et al. 1999), as was the case with our patient who presented with thyrotoxicosis with serum glucose infusions without vitamin supplementation at the beginning of management.

Gayet-Wernicke encephalopathy is a medical emergency. The substitute treatment should be early, as soon as the diagnosis is suspected, and should not be delayed by vitamin dosing to allow the reversibility of neurological impairment.

There is no consensus on the amount of thiamine to be delivered, the duration of treatment, and the number of doses given per day. It is especially appropriate to introduce vitamin B1 therapy rapidly, by parenteral means, for some until vomiting stops and normal feeding resumption, for others until the end of pregnancy (Gardian et al. 1999). Our parturient had been treated with vitamin B1 intravenously at a dose of 1 g/d for 10 days, then relayed 500 mg/d orally during the remainder of the pregnancy.

The introduction of thiamine may change the evolution of GWE. Ocular symptoms usually begin to improve 1–6 h after the start of treatment (Gardian et al. 1999), ataxia, and confusion within days to weeks. Residual impairment such as nystagmus or ataxia has been documented in approximately 60% of cases. In the absence or inadequately managed GWE can lead to coma, death (17–20%), or long-term Korsakoff syndrome (Sechi and Serra 2007).

The evolution of our patient is marked by the gradual improvement of symptoms over a 1-year period.

The fetal prognosis according to Spruill and Kuller's article was favorable in the various published cases when treatment was started early. Rare or exceptional cases of malformation, abortion, or death in utero have been reported in the literature (Spruill and Kuller 2002). Our parturient pregnancy was completed, resulting in a vaginal delivery of a baby with moderate growth retardation (below the 10th percentile), and without any malformations.

Conclusion

All pregnant women with severe hyperemesis gravidarum should be supplemented with thiamine prior to glucose administration to prevent and improve the prognosis of Gayet-Wernicke encephalopathy.

Abbreviations

GWE: Gayet-Wernicke encephalopathy; MRI: Magnetic resonance imaging; g/d: Gram per day

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Authors' contributions

HM developed the idea and the design of the study. BD revised literature, collected the data, FN analyzed the data, and HM wrote and critically revised the manuscript. The authors read and approved the final version of the manuscript.

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