WERNICKE’S ENCEPHALOPATHY: RARE COMPLICATION OF HYPEREMESIS GRAVIDARUM

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ABSTRACT Wernicke’s encephalopathy is a rare disease resulting from severe thiamine deficiency (vitamin B1). Alcoholic subjects are most often affected, but it may occur outside of this addiction. Magnetic resonance imaging is the gold standard imaging modality to confirm the diagnosis. This pathology manifests in T2-weighted hyperintensity in the periaqueductal, thalami and mammillary bodies. A suitable treatment makes it possible to avoid heavy sequelae. We report the case of a 23-year-old woman, pregnant with 12 weeks of amenorrhea, with diplopia, headache and abundant vomiting, referring to Wernicke’s encephalopathy.

RÉSUMÉ L’encéphalopathie de Wernicke est une pathologie carentielle causée par un déficit profond en Thiamine (vitamine B1). Elle survient le plus souvent sur un terrain alcoolique, mais parfois elle est de diagnostic difficile et dont l’évolution en l’absence de traitement conduit à des séquelles cognitives sévères. L’imagerie par résonance magnétique est l’examen de référence permettant de confirmer le diagnostic par la présence d’hypersignaux T2 au niveau périaqueducal, des thalami, et des corps mamillaires. Nous rapportons l’observation d’une femme de 30 ans ayant des vomissements abondants lors du premier trimestre de la grossesse (hyperemesis gravidarum), à l’origine d’une encéphalopathie de Wernicke symptomatique.

Mots cles: Encéphalopathie de Gayet Wernicke, vomissements pendant la grossesse, vitamine B1

KEYWORDS Gayet Wernicke’s encephalopathy, vomiting during pregnancy, vitamin B1

Introduction
Gayet-Wernicke encephalopathy (WE) is a rare and severe neurological complication due to a thiamine deficiency causing brain lesions of the medial limbic circuit. It can also be located in the periventricular regions around the third and fourth ventricles, the aqueduct of Sylvius and the mammillary bodies. This serious neurological pathology has mortality reaching 30% if not diagnosed and treated in time [1]. WE is a known complication of chronic alcoholism. Still, it should also be known that it can occur outside of this addiction [2,3]. We report a case of non-alcoholic Gayet-Wernicke encephalopathy complicating incoercible vomiting in a pregnant woman.

Observation
A 23-year-old patient who was 12 weeks pregnant, with no significant history of disease or drug use, was referred to the emergency department for neurological symptomatology made of diplopia, headache and incoercible vomiting. During her hos-
pitalization, the patient experienced worsening her neurological condition by installing a confusional syndrome with significant temporospatial disorientation.

The neurological examination revealed significant memory disorders, static and kinetic cerebellar syndrome, abolition of osteo-tendon reflexes and hypoaesthesia, mainly in the lower limbs. The biological balance showed moderate hepatic cytoly-
sis, the moderate elevation of pancreatic enzymes, and metabolic alkalosis with hypokalemia.

An encephalic MRI showed the appearance of hypersignals at the periaqueductal level, the mammary bodies, the two thalami and around the 3rd ventricle very evocative of an EGW. Parenteral vitamin B1 supplementation (1g/24h) with vitamin B6 was introduced and then relayed orally (vitamin B1 500 mg*2/d) with the appearance of Clinical Improvement. A 60-month MRI was introduced and then relayed orally (vitamin B1 500 mg*2/d) with the appearance of Clinical Improvement. A 60-month MRI was introduced and then relayed orally (vitamin B1 500 mg*2/d) with the appearance of Clinical Improvement. A 60-month MRI was introduced and then relayed orally (vitamin B1 500 mg*2/d) with the appearance of Clinical Improvement.

Discussion

Wernicke’s encephalopathy was first described by Wernicke in 1881 [4]. It’s a pathology related to a thiamine deficiency, most often underdiagnosed with a clinical prevalence of 0.04% to 0.13% against 0.8% and 2.8% in pathology [5,6]. This under-diagnosis is due to deceptive clinical forms in patients not identified as at risk [7,8] and frequent atypical presentations.

One in five patients shows none of the classical signs of this condition; moreover, these signs are sometimes difficult to differentiate acute or chronic ethyl intoxication, the main risk area of this disease pathology in the western world [7,8]. This B1 vitamin deficiency can complicate other pathological conditions such as malnutrition, anorexia nervosa, gastrointestinal tumour, chemotherapy or prolonged parenteral nutrition without thiaminic supplementation. In our patient, hypovitaminosis B1 was secondary to incoercible vomiting.

Hyperemesis gravidarum complicates 0.5 to 2% of pregnancies [9]. This sd is defined by deep vomiting in the first trimester of pregnancy, leading to weight loss, Extracellular dehydration and metabolic alkalosis with Transient hypokalaemia hyperthyroidism may be observed and contribute to hypokalaemia’s severity [10]. In the literature, the first case of hyperemesis gravidarum has been reported by Henderson in 1914 and rare sporadic cases have been described since [11]. Togay-Isikay and al.’s article updated 30 clinical cases of Gayet-Wernicke’s encephalopathy in the context of hyperemesis gravidarum published between 1968 and 2000 [12]. In 1997, Olindo et al. demonstrated the association of this syndrome with centropontin myelinoysis in pregnancy vomiting [13]. The diagnosis of GWE is primarily clinical with the classic triad [14] which associates psychic disorders (confusional syndrome, apathy, bradypsism, hypersonnia), eye disorders (horizontal or multiple nystagmus, Oculomotor paralysis by the development of III and VI) and disturbances of balance, related to central vestibular syndrome and cerebellar syndrome; however, this triad is complete only in 30% of the cases.

Other symptoms can also be related, such as hypothermia, hypotension, tachycardia, hallucinations, headache, and abdominal discomfort. Dysarthria, dysphagia, hypotonia of the lower limbs, deafness, myoclonus, dyskinesia, dystonia, epilepsy, disorders psychosis with auditory hallucinations and delusions of persecution or bulimia have also been described. Peripheral neuropathy is often associated but rarely sought [14]. Korsakoff syndrome is described in 80% of the cases during a GWE [14].

MRI shows abnormalities in 60% of cases in imaging, implying that normal imaging does not exclude the diagnosis [15,16]. In the days following the first clinical signs, hypersignals in T2, FLAIR and diffusion, typical by their location and symmetry around the Sylvius aqueduct, the 3rd ventricle (V3), the medial face of the thalamus and especially at the level of the mammary tubers, can be observed. Furthermore, these lesions take on the contrast inconsistently after injection of Gadolinium chelate [18,19]. Atypical locations have been reported, with signal anomalies, that can make the diagnosis difficult [19]. In 1998, Antunez et al.’s work had shown that the T2-FLAIR hypersignal in the peri-aqueductal regions and thalamic dorso-median nuclei favored such encephalopathy with a specificity of 93% [15]. The diagnosis remains based on clinical signs and, above all, on significant improvement after treatment with thiamine. The scanner itself has not proved its diagnostic utility. Finally, blood tests for vitamin B1 deficiency require access to specialized laboratories and results are obtained only late, making them of limited use in clinical practice. For the treatment, vitamin B1 therapy should be introduced rapidly, parenterally, for some until vomiting stops and normal nutrition restarts, for others until the end of the pregnancy. The reversibility of the disorders and the prognosis depend essentially on the neurological signs’ duration before the treatment’s introduction.

Conclusion

Our patient’s clinical context made us suspect Wernicke’s encephalopathy in the first place. MRI facilitated the diagnosis by finding FLAIR hypersignals in a region of interest (peri-aqueductal, thalamus or mammary body). This deficiency should be mentioned in view of any neurological manifestations.
for any pregnant patient in the first trimester and who has pregnancy vomiting with a thiamine deficiency to avoid irreversible sequelae.

**Funding**

This work did not receive any grant from funding agencies in the public, commercial, or not-for-profit sectors.

**Conflict of Interest**

There are no conflicts of interest to declare by any of the authors of this study.

**References**

1. Phayphet M, Rafat R, Andreux F, Parraga E, Rondeau E, Hertig A. Hyperemesis gravidarum: une cause rare de l’encephalopathie de wernicke. Presse Med. 2007;36(12 Pt 1):1759–61. [PubMed] [Google Scholar]

2. Ogerson PR, Rahman A, Nestor S, Brick J. Wernicke encephalopathy in nonalcoholic patients. Am J Med Sci. 2002;323(2):107–11. [PubMed] [Google Scholar]

3. Reuler JB, Girard DE, Cooney TG. Wernicke’s encephalopathy. N Engl J Med. 1985;312(16):1035–9. [PubMed] [Google Scholar]

4. Wernicke C. Lehrbuch der Gehirnkrankheiten fur Aerzte und Studierende. Fischer édit Berlin. 1881;2 [Google Scholar]

5. Sechi G, Serra A. Wernicke’s encephalopathy: new clinical settings and recent advances in diagnosis and management. Lancet Neurol. 2007;6(5):442–55. [PubMed] [Google Scholar]

6. Mc Candless DW. New-York: Springer; 2009. Metabolic encephalopathy. [Google Scholar]

7. Zacconi G, Santa Cruz D, Bertolini M, Rovira A, Gallucci M, Carolo C, et al. MR imaging findings in 56 patients with Wernicke encephalopathy: nonalcoholics may differ from alcoholics. AJNR Am J Neuroradiol. 2009;30(1):171–6. [Article PMC gratuit] [PubMed] [Google Scholar]

8. Ebels EJ. Underlying illness in Wernicke’s encephalopathy: analysis of possible causes of under diagnosis. EurNeurol. 1974;12(4):226–228. [PubMed] [Google Scholar]

9. Massou S, EL Fazazi H, Atmani M, Azendour H, Belyamani L, Kamili ND. Myopathie hypokaliémique: complication rare des vomissements gravidiques. Ann Fr Anesth Reanim. 2009;28(7-8):713. [PubMed] [Google Scholar]

10. Goodwin TM, Montoro M, Mestman JH. Transient hypothyroidism and hyperemesis gravidarum: clinical aspects. Am J Obstet Gynecol. 1992;167(3):648–52. [PubMed] [Google Scholar]

11. Henderson D. Korsakow’s psychosis occurring during pregnancy. Bulletin of the Johns Hopkins Hospital. 1914;25:261–70. [Google Scholar]

12. Togay-Isikay C, Yigit A, Mutluer N. Wernicke’s encephalopathy due to hyperemesis gravidarum: an under-recognised condition. Aust N Z J Obstet Gynaecol. 2001;41(4):453–6. [PubMed] [Google Scholar]

13. Olindo S, Smadja D, Cabrè P, Mehdoei H, Heinzlef O. Encéphalopathie de Gayet-Wernicke et myélinolyse centropontine induites par des vomissements gravides. Rev Neurol. 1997;153(6-7):427–9. [PubMed] [Google Scholar]

14. 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(4):341–5. [Article PMC gratuit] [PubMed] [Google Scholar]

15. Antunez E, Estruch R, Cardenal C, Nicolas JM, Fernandez-Sola J, Urbano-Marquez A. Usefulness of CT and MR imaging in the diagnosis of acute Wernicke’s encephalopathy. AJR Am J Roentgenol. 1998;171(4):1131–7. [PubMed] [Google Scholar]

16. Fei GQ, Zhong C, Jin L, Wang J, Zhang Y, Zheng X, et al. Clinical characteristics and MR imaging features of non-alcoholic Wernicke encephalopathy. AJNR. 2008;29(1):164–9. [Article PMC gratuit] [PubMed] [Google Scholar]

17. Chu K, Kang DW, Kim H, Kim HJ, Lee YS, Park SH. Diffusion-weighted MR abnormalities in Wernicke’s encephalopathy: reversible cytotoxic edema. Arch Neurol. 2002;59(1):123–7. [PubMed] [Google Scholar]

18. Schroth G, Wichmann W, Valavanis A. Blood-brain-barrier disruption in acute Wernicke encephalopathy: MR findings. J Comput Assist Tomogr. 1991;15(6):1059–61. [PubMed] [Google Scholar]

19. Vargas MI, Lenz V, Bin JF, Bogorin A, Abu Eid M, Jacques C, et al. Imagerie par résonance magnétique (IRM) des complications encéphaliques de l’alcoolisme. J Radiol. 2003;84(4 Pt 1):369–79. [PubMed] [Google Scholar]

20. Spruill SC, Kuller JA. Hyperemesis Gravidarum Complicated by Wernicke’s Encephalopathy. Obstet Gynecol. 2002;99(5 Pt 2):875–7. [PubMed] [Google Scholar]