AN UNUSUAL PRESENTATION OF GAYET-WERNICKE ENCEPHALOPATHY IN A PREGNANT WOMAN

UNE PRESENTATION INHABITUELLE DE L'ENCEPHALOPATHIE DE GAYET-WERNICKE CHEZ UNE FEMME **ENCEINTE**

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Abstract

Gayet-Wernicke encephalopathy is an uncommon neurologic disease that can cause severe morbidity and mortality, if untreated. A 37-year-old woman at 30 weeks of pregnancy was referred to our hospital for severe acute and unusual headache. Clinical examination found a conscious woman. A meningeal syndrome and a palsy of the right sixth cranial nerve were objected. The patient had an urgent cerebral computed tomography scan with radioprotection measures for the fetus. The CT scan was normal. Cerebral magnetic resonance imaging was performed the following day. It showed a symmetrical bilateral hypersignal of the mamillary bodies on Fluid-Attenuated Inversion Recovery sequences. Gayet-Wernicke encephalopathy was suspected. The patient had an urgent intravenous supplementation of Thiamine. The headache decreased gradually. An oral supplementation of thiamine was continued until the end of pregnancy. She gave birth to a healthy baby girl.

Key - words: Gayet-Wernicke encephalopathy; Thiamine; Headache; Pregnant

Résumé

L'encéphalopathie de Gayet-Wernicke est une maladie neurologique peu fréquente qui peut entraîner une morbidité et une mortalité graves si elle n'est pas traitée. Une femme de 37 ans, à 30 semaines de grossesse, a été adressée à notre hôpital pour des céphalées aiguës et inhabituelles. L'examen clinique a révélé une femme consciente. Un syndrome méningé et une paralysie du sixième nerf crânien droit ont été objectivés. La patiente a subi en urgence une tomodensitométrie cérébrale avec des mesures de radioprotection pour le fœtus. Le scanner était normal. Une imagerie par résonance magnétique cérébrale a été réalisée le lendemain. Elle a montré un hypersignal bilatéral symétrique des corps mamillaires sur les séquences de récupération par inversion atténuée par le fluide. Une encéphalopathie de Gayet-Wernicke a été suspectée. Le patient a reçu en urgence une supplémentation intraveineuse en thiamine. La céphalée a diminué progressivement. Une supplémentation orale en thiamine a été poursuivie jusqu'à la fin de la grossesse. La patiente a accouché d'une petite fille en bonne santé.

Mots-clés : Encéphalopathie de Gayet-Wernicke ; Thiamine ; Céphalées ; Grossesse.

ملخص

يعتبر الاعتلال الدماغي غايت فيرنيك مرضا عصبيا غير شائع و يمكن أن يؤدي إلى مراضة و وفيات خطيرة إذا لم يقع علاجه.
أحيلت امرأة تبلغ من العمر 37 سنة، حامل في الأسبوع 30، إلى المستشفى بسبب صداع حاد غير معهود. كشف الفحص السريري عن وجود امرأة واعية. وقع كشف وجود متلازمة سحائية و شلل بالعصب القحفي السادس الأيمن.
خضعت المريضة لفحص طارئ بالأشعة المقطعية للدماغ مع تدابير حماية من الإشعاع للجنين.
كان الكشف الدماغي المقطعي عاديا. وقع اعتماد الكشف الدماغي بالتصوير الطبي بالرنين المغناطيسي في اليوم الموالي ليبين حصول شدة ثنائية متناظرة للأجسام الثديية على متواليات استعادة الانعكاس المضعف بالسوائل. تم الاشتباه في الاعتلال الدماغي غايت فيرنيك. تلقت المريضة بصفة استعجالية مكملات التيامين في الوريد. و منها نقصت حدة الصداع بصفة تدريجية.
وقع استمرار تكميل التيامين عبر أخذه عن طريق الفم حتى نهاية الحمل. ثم أنجبت المريضة طفلة سليمة.

الكلمات المفاتيح: الاعتلال الدماغي غايت فيرنيك ; التيامين ; الصداع ; الحمل.

INTRODUCTION

Gayet-Wernicke encephalopathy (W.E) is an uncommon neurologic disease caused by deficiency in thiamine or vitamin B1. It is a serious condition, with severe morbidity and mortality[1]. It is underdiagnosed in children and adults[2]. W.E is frequently associated with chronic alcohol intake[3]. However, many other conditions can be the cause such as: bariatric surgery, starving, intractable vomiting, chronic renal failure.... Diagnosis of W.E is clinical and magnetic resonance imaging (MRI) helps confirm it[2]. Treatment is simple but must be urgent in order to prevent severe sequelae, such as Korsakov syndrome, and death.

CASE REPORT

A 37-year-old woman (gravida: 3, para: 2) at 30 weeks of pregnancy was referred to our hospital for severe acute headache. The pain started suddenly and was so intense and unusual that the patient stopped immediately her activity and laid down. Few episodes of vomiting followed shortly with a sensation of relief. Clinical examination found an alert and conscious woman. She had no fever. A meningeal syndrome and a palsy of the right sixth cranial nerve were objected. The diagnoses of subarachnoid hemorrhage and cerebral venous thrombosis were suspected and the patient had an urgent cerebral computed tomography (CT) scan with radioprotection measures for the fetus. The CT scan was normal (Figure 1). A lumbar puncture was performed and the cerebrospinal fluid analysis was normal. Magnetic resonance imaging (MRI) was performed the following day. It showed a symmetrical bilateral hypersignal of the mamillary bodies on Fluid-Attenuated Inversion Recovery (FLAIR) sequences (Figure 2). W.E was suspected. When we asked the patient again for more specific details, she reported recurrent episodes of vomiting and poor oral intake for the last 3 weeks that were neglected. The patient had an urgent intravenous supplementation of 500 mg of Thiamine followed by intravenous perfusion of 200 mg of Thiamine three times a day for five days then twice a day for five other days. Headache decreased gradually and disappeared during the first week after treatment. An oral supplementation of thiamine was continued until the end of pregnancy. She had a vaginal birth of health baby girl.

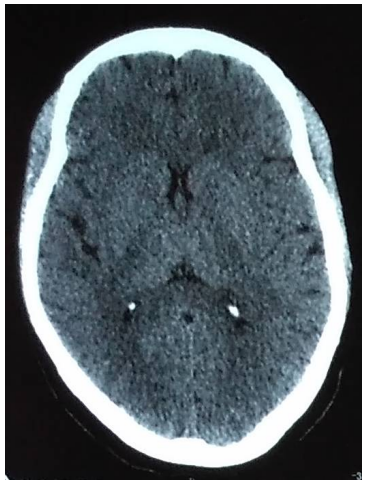

Figure 1: Axial view of a cerebral computed tomography without contrast agent injection showing no signs of acute subarachnoid hemorrhage

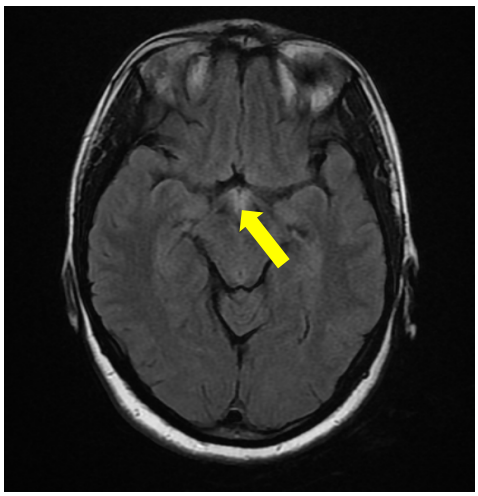

Figure 2 : Axial FLAIR weighted cerebral MRI showing a symmetrical hyperintense signal in the mamillary bodies (yellow arrow)

DISCUSSION

W.E was first described by Wernicke in 1881[4]. It was associated for a long time with chronic alcohol intake for most of the cases. It was only in the forties that it was related to thiamine deficiency[5]. W.E is underdiagnosed in both children and adults[1,6]. The prevalence of the disease in autopsy studies is higher than in clinical ones[7]. Many conditions can cause the disease, mainly chronic alcohol intake but also hyperemesis gravidarum, bariatric surgery, malnutrition and starvation, and hemodialysis….

Vitamin B1 is a water-soluble vitamin that cannot be produced in the human body[4,8]. A balanced diet is sufficient to provide the daily requirements of thiamine which is about 5mg. The jejunum is the site where it is absorbed. Depending on its concentration in the intestines, the absorption can proceed in two different ways. When the concentration is low, an active process absorbs the thiamine while a passive mucosal process is activated when the concentration is high[4,8]. The absorbed vitamin B1 is then stored in the body, mostly in the muscles, with a maximum storage capacity up to 30mg[4].

Thiamine is crucial to the human body as it is a co-enzyme of three essential enzymes: ketoglutarate dehydrogenase complex, pyruvate dehydrogenase complex and transketolase[9]. A deficit in thiamine causes a deficit in these enzymes leading to serious sequalae. A decrease in the ketoglutarate dehydrogenase activity induces a decrease in adenosine 5-triphosphate production and results in an increase of lactate levels and neuronal excitotoxicity[10]. Thiamine deficiency affects selective regions in the brain with high concentration and turnover of the vitamin. The symptoms start when the blood concentration of thiamine drops. It happens after the body storage is depleted, which takes about two to three weeks after the deficiency[10].

The disease was characterized by a clinical triad consisting of ataxia, confusion and ophthalmic disorders[1]. However, this triad is only found in 16% of the cases and most of the patients develop the ophthalmic disorders only, as they are the most frequent (93%)[1]. Caine et al. proposed new criteria to diagnose W.E, which require the presence of two out of four of the following signs: dietary deficiencies; oculomotor abnormalities; cerebellar dysfunction, and either an altered mental state or mild memory impairment[11].

The diagnosis of W.E is clinical and brain MRI can be a good help to confirm it especially when the clinical criteria are not satisfied. It has a sensibility of 53% and a specificity of 93%[12]. It typically shows in T2-weighted and FLAIR sequences a bilateral symmetrical hypersignal in the thalamus, hypothalamus, mamillary bodies, periaqueductal region, the fourth ventricle and the midline cerebellum. Such typical MRI pattern is found in 58% of the cases[12,13].

W.E is a medical emergency. Treatment consists of intra venous injection of thiamine to allow an optimum body intake. There is no consensus concerning the optimal dose, or the frequency of admission or the duration of treatment[2].

In 2016, Boulanger et al. tried to identify the consensual treatment for W.E[14]. For curative purpose, 500 mg of thiamine is administered intravenous three times a day for three to five days[14]. If there is a good response, the treatment is continued with 250 mg intravenous three times a day for three to five more days[14]. For prophylactic purpose in ambulatory patients, it consists of 100 to 300 mg of daily oral intake. For hospitalized patients, the posology is 250 to 300 mg intravenous for three to five days followed by 300 mg daily oral intake until returning to a balanced diet[14].

W.E is a serious condition and must be diagnosed and treated without delay. Patients risk sequelae that can be serious such as Korsakoff syndrome, which affects 80% of patients who previously had W.E[15]. The mortality of this disease is estimated to 17%[15].

W.E has been associated with hyperemesis gravidarum. However, this case is unusual since the acute onset of the headaches and the meningeal syndrome have not been reported before in the English literature, to the best of our knowledge, in association with W.E. We searched PubMed and Google Scholar using the following MeSH "Headache", "Meningism" and "Wernicke Encephalopathy". We did not find any report stating them in patients with the disease. Such unusual presentation can be misleading and cause delay to the actual diagnosis.

CONCLUSION

W.E is a serious disease with severe sequelae and high mortality. It is frequent but underdiagnosed. Treatment must be administered immediately upon suspicion. Prophylaxis is a necessity especially in patients that are predisposed.

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