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WERNICKE-KORSAKOFF SYNDROME IN PRIMIPAROUS: CASE REPORT

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ABSTRACT

Wernicke-Korsakoff syndrome results from the combination of Wernicke's encephalopathy and Korsakoff's syndrome, and is associated with thiamine (vitamin B1) deficiency. Wernicke's encephalopathy is characterized by ataxia, ophthalmoparesis, and mental confusion, with a higher prevalence in severe alcoholics, although it can also occur due to malnutrition, liver diseases, anorexia nervosa, among other causes. Korsakoff's syndrome, on the other hand, is a chronic condition, diagnosed by confabulatory amnesia and difficulties in organizing events in time. Thiamine deficiency impairs energy production in nerve cells and affects brain function. Changes in the central nervous system, especially in the limbic circuits, are seen on imaging. Hyperemesis gravidarum, characterized by intense nausea and vomiting during pregnancy, can lead to malnutrition and thiamine deficiency, with the risk of developing Wernicke-Korsakoff syndrome, as demonstrated in this report by a primiparous woman from a tertiary service in the interior of São Paulo.

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INTRODUCTION

Wernicke-Korsakoff syndrome is a pathology that arose from the junction of two others: Wernicke's encephalopathy and Korsakoff's syndrome. Initially described in the 19th century, it is commonly known that this neuropsychiatric pathology is associated with thiamine deficiency, a fat-soluble vitamin of the B complex (B1) with a prevalence of 0% to 2% worldwide.[1,3,5,6,7,8].

Wernicke's encephalopathy presents as a classic triad of acute form with ataxia, ophthalmoparesis with nystagmus and mental confusion and can be clinically identified through delirium in severe alcoholic patients, as this population is more associated with chronic deficiency of the vitamin in question, however, other fundamental causes have been described to explain vitamin B1 deficiency, such as: severe malnutrition, hyperemesis gravidarum, prolonged parenteral nutrition, malignant diseases such as terminal cancers, liver diseases, anorexia nervosa, immunocompromise, intestinal diseases, bariatric surgery, etc.2,3.

Korsakoff's syndrome, in turn, presents in a more chronic alteration and its diagnosis consists of a broad psychiatric evaluation that, with the help of the DSM-5, characterizes this pathology as confabulatory amnesia, a major neurocognitive disorder with structural alterations of the central nervous system. The dysfunction may also present with difficulty in identifying the temporal structuring of events3.

Vitamin B1 is responsible for catalyzing energy generation through decarboxylation of branched-chain amino acids and acts as a coenzyme for transketolase reactions. Thus, thiamine, as vitamin B1 is called, also plays a poorly understood role in nerve conduction5. Early changes in astrocytes and microglia were observed in thiamine deficiency, with loss of glutamate transporters. On imaging studies, there is "altered signal in several components of the limbic circuits, including the paraventricular regions of the thalamus, the hypothalamus, the mammillary bodies, the periaqueductal region, the floor of the fourth ventricle, and the midline cerebellum"[3].

During pregnancy, up to 90% of women experience nausea and the exact causes and mechanism remains unclear. Some theories involve an increase in chorionic gonadotropin and high levels of estrogen for the appearance of this symptom. Hyperemesis gravidarum refers to extreme cases of nausea and vomiting in the first trimester of pregnancy, and can extend into advanced gestational stages. As a consequence, significant loss of electrolytes, dehydration, and even malnutrition occur if not well managed. Thus, the diagnosis arises from weight loss associated with dehydration and intense vomiting when discarding possible differential diagnoses4.

Understanding the relationship between pregnancy and volume loss and malnutrition in the face of hyperemesis gravidarum, it is possible to elucidate the broad relationship with thiamine deficiency and some reported cases of Wernicke-Korsakoff Syndrome in pregnant women6,7,8. Thus, this report brings to light another case of a young pregnant woman diagnosed with this pathology in a hospital in the interior of São Paulo, in order to assist the scientific community in the management of such a disease with a potential unfavorable outcome if it is not treated quickly and adequately.

METHODOLOGY

This is a case report study, whose information was collected through a review of medical records. In parallel, to support the ideas discussed in this article, a literature review was carried out in scientific databases such as PubMed. The production of this scientific article followed the regulations proposed by the National Research Council (CONEP).

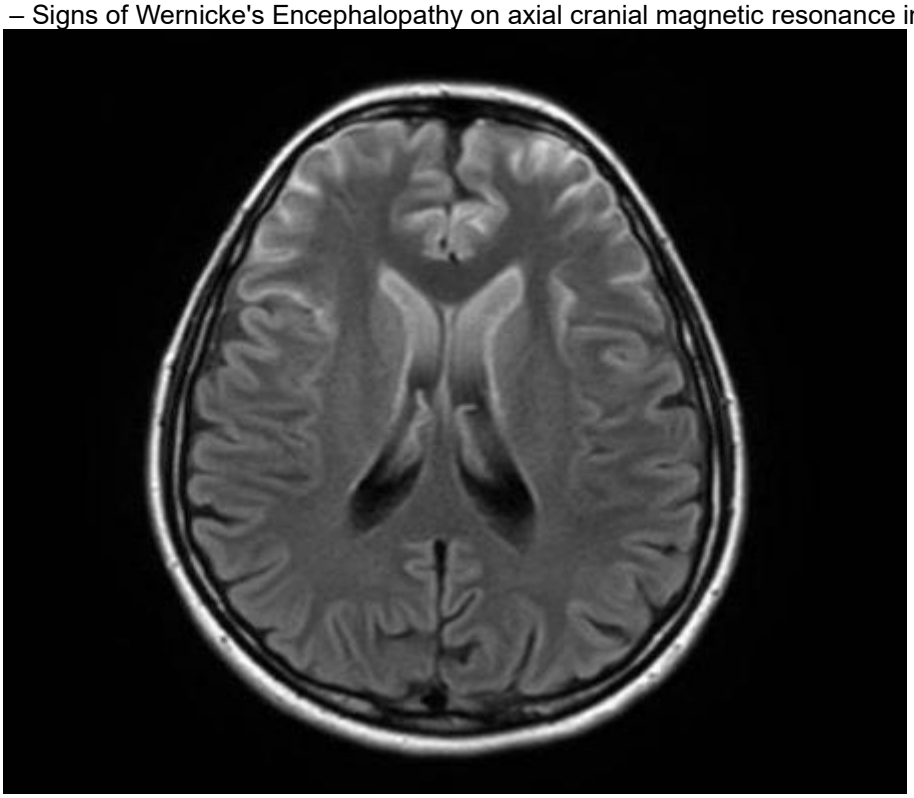
CASE REPORT

A 20-year-old patient, G1P0AO, 17 weeks and 1 day (by USG performed at 12 weeks) was referred to the tertiary service for evaluation by the neurology team due to mental confusion and recent memory loss. A companion also reports significant weight loss since the patient discovered the pregnancy (approximately 20 kg). He did not present obstetric complaints. He denies previous comorbidities.

The patient was admitted to the Gynecology and Obstetrics ward, with daily follow-up by the neurology team, who advised the performance of cranial MRI, electroencephalogram, cerebrospinal fluid puncture and laboratory tests, to investigate the etiology of mental confusion and memory loss. The blood count showed normocytic and normochromic anemia, and cranial MRI showed signs of Wernicke's encephalopathy (Figures 1 and 2), with no cerebrospinal fluid abnormalities and negative serology.

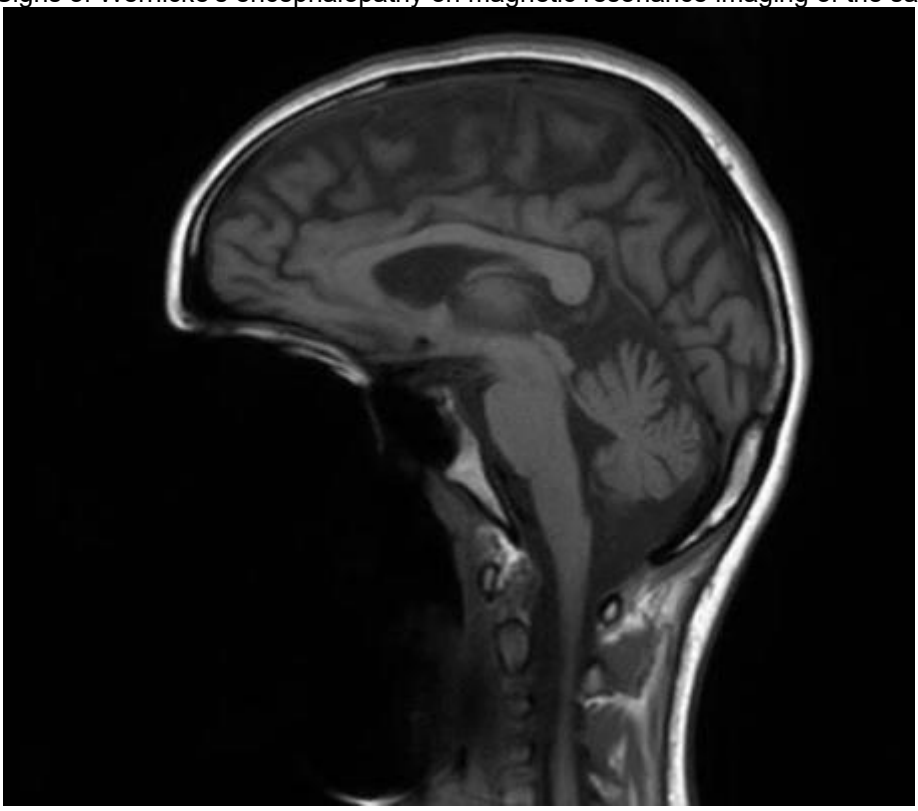
Thiamine and ferrous sulfate replacement was initiated. Throughout the treatment, the patient showed significant clinical improvement, with no new episodes of mental confusion. Regarding the obstetric part, the patient had no complaints, and obstetric ultrasonography was performed on the day of discharge, which showed no changes. After the end of all treatment, the patient is released by the neurology team for outpatient and usual prenatal follow-up.

Figure 1 – Signs of Wernicke's Encephalopathy on axial cranial magnetic resonance imaging.



Source: The authors

Figure 2 - Signs of Wernicke's encephalopathy on magnetic resonance imaging of the sagittal skull.



Source: the authors.

DISCUSSION

Thiamine deficiency is the main cause of Wernicke-Korsakoff syndrome (SWK), and more than 90% of cases occur in alcoholics9. In this clinic, little is said about pregnant

women with this vitamin deficiency, but this is not an unknown pathology for gynecology and obstetrics.

SWK begins primarily with Wernicke's encephalopathy and later progresses to Korsakoff's syndrome, the latter having irreversible potential and with considerable risk to the patient's life. The symptoms, despite being well described in the literature, are presented in practice as nonspecific, since the classic triad (ataxia, mental confusion and ophthalmoparesis associated with nystagmus) is present in only 16% of the cases7. Here we report a primiparous woman who was admitted to the health service with considerable mental confusion. In addition to this fact, memory loss was a significant factor for clinical diagnosis, since this symptom is not part of the triad described, but belongs to the broad spectrum of neurological symptoms that help in the diagnosis of WKS.

Considering the insidious character of Wernick's encephalopathy and the chronicity of Korsakoff's syndrome, it was not expected that this pregnant woman would develop clinical conditions for both dysfunctions. However, the confabulatory amnesia presented at the patient's admission is recognized as Korsakoff's symptomatology, according to the DSM-53. This fact may suggest a history of malnutrition from a time long before the pregnancy itself.

Obstetrics is aware of the association of SWK with hyperemesis gravidarum. By analyzing the connection between pregnancy, increased body volume, and malnutrition associated with hyperemesis gravidarum, it is possible to clarify the high occurrence of cases of Wernicke-Korsakoff Syndrome in pregnant women6,7,9. A history of excessive vomiting and anorexia aid in the diagnosis when it occurs in obstetric patients.

Thiamine, one of the essential vitamins, has a recommended daily dose of 5 mg, obtained through various foods, since the human body stores 25 to 30 mg of this vitamin. During pregnancy, the need for thiamine triples, and the daily intake is indicated at about 15 mg8. This patient did not present symptomatological alterations at the beginning of pregnancy that could diagnose a case of hyperemesis gravidarum, however, in underworld countries such as Brazil, it is still common to observe cases of malnutrition due to dietary deficiency, corroborated by the concomitant alteration of the blood count suggesting iron deficiency anemia, also caused by poor diet, and the considerable weight loss of the same patient reported. In other words, a long history of vitamin deficiency did not require excessive vomiting to culminate in a potentially fatal pathology, only high body demand could already lead to this outcome.

Thiamine supplementation as a routine was not found in the current recommendations for the management of pregnant women during prenatal care in Brazil, as

this is not a common situation in the country, despite malnutrition being a significant problem. All the literature studied around thiamine deficiency in pregnant women revolves around isolated case reports. In all, only one was identified as occurring in Brazil6. This, in turn, agrees that there are no precise protocols in the management of pregnant women with SWK.

Some studies, such as Dingwall, Kylie M et al (2022)[9] evaluate prophylactic thiamine supplementation in alcoholic patients, suggesting doses of up to 200mg intravenously in three applications. However, we must take into account that thiamine is essentially part of the generation of cellular energy by the Krebs cycle, which justifies the neurological alterations in patients with this vitamin deficiency, since the central nervous system has a high energy demand5. However, the demand on patients during the gestational course is even greater. When associated with a picture of deficient intake, the consequences can be even more serious. Therefore, in the present Brazilian study identified and referenced here, it suggests an international protocol for intravenous supplementation of up to 500 mg of thiamine three times a day during pregnancy6.

Considering the scarcity of randomized studies and clinical trials regarding the situation described above, this report aims to assist in the management of obstetricians regarding pregnant women with Wernicke-Korsakoff Syndrome, observing the positive outcome regarding the protocol adopted to treat this specific pregnant woman reported here. The patient progresses with significant improvement of symptoms and is discharged from the hospital without sequelae after a few days of intravenous thiamine supplementation, and can have his prenatal care completed on an outpatient basis.

CONFLICTS OF INTEREST

The authors state that there is no potential conflict of interest that could compromise the impartiality of the information presented in this scientific article.

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