Malnutrição: Uma Causa Subdiagnosticada de Síndrome de Wernicke
Malnutrition: An Underdiagnosed Cause of Wernicke's Syndrome

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Resumo:

A síndrome de Wernicke é uma condição subdiagnosticada e frequentemente não identificada em vida. A principal causa é a perturbação do uso do álcool, no entanto existem outras menos comuns como a malnutrição, nomeadamente provocada por cirurgia bariátrica. As manifestações iniciais são habitualmente neurológicas e cursam com confusão, ataxia e oftalmoplegia. O diagnóstico é clínico, a confirmação laboratorial surge tardiamente e os achados em ressonância magnética corroboram o diagnóstico, mas não são patognomónicos. A reposição empírica precoce de tiamina é necessária para prevenir défices neurológicos permanentes ou mesmo morte. Apresentamos um caso de défice de tiamina associado a malnutrição em doente com obesidade e sujeita a cirurgia bariátrica recente. Assim, pretende-se alertar para a importância de manter um baixo limiar de suspeita para este défice vitamínico, mesmo na ausência de hábitos alcoólicos, e de iniciar precocemente reposição.

Palavras-chave: Cirurgia Bariátrica; Deficiência de Tiamina; Desnutrição; Encefalopatia de Wernicke.

Abstract:

Wernicke syndrome is an underdiagnosed condition, and many cases are not identified in life. The main cause is alcohol abuse, however there are others less evident as malnutrition or bariatric surgery. Generally, this disease presents with neurological symptoms such as: confusion, ophthalmoplegia and ataxia. The diagnosis is primarily clinical and often confirmed days later. Magnetic resonance imaging findings are not pathognomonic but can corroborate the diagnosis. Early thiamine treatment is necessary to avoid permanent neurological deficits or even death. We present a case of thiamine deficiency due to malnutrition in an obese patient recently submitted to bariatric surgery. The authors' purpose is to alert to the importance of maintaining a high level of suspicion for thiamine deficiency, even in patients without history of alcohol abuse for successful outcome.

Keywords: Bariatric Surgery; Malnutrition; Thiamine Deficiency; Wernicke Encephalopathy.

Introduction

There are limited thiamine body stores, depending exclusively on exogenous sources. Thiamine is absorbed mainly in the duodenum and the reserves can last for 2-3 weeks.[1] The main cause for thiamine deficiency is alcohol abuse which may lead to Wernicke syndrome. Other causes include malnutrition, fasting, starvation, unbalanced diets, bariatric surgery (gastric banding, gastric bypass, bilio-pancreatic diversion), *hyperemesis gravidarum*, cancer, parenteral nutrition, dialysis, renal diseases and others.[2] The central nervous system is the first system affected by thiamine deficiency.[1] Wernicke syndrome is a clinical diagnosis supported by brain magnetic resonance imaging (MRI) and confirmed by blood thiamine concentration. MRI typically presents abnormal bilateral T2-weighted hyperintensities in the paraventricular regions of the thalamus, hypothalamus, mammillary bodies, periaqueductal region and floor of the fourth ventricule.[3] Usually, results of blood analysis are only available after empirical treatment.[3] In this paper we report an uncommon cause of thiamine deficiency.

Case Report

24-year-old woman, history of arterial hypertension, hypercholesterolemia, obesity (body mass index 52 kg/m^2) and no previous history of ethanol consumption. Two months earlier, patient was submitted to bariatric surgery, a vertical gastrectomy. The patient did not tolerate the proposed diet progression after surgery, nor reported it, which led to a profound reduction of ingestion. Instead of complying the dietary recommendations, the patient consumed frequently soda and candies and subsequently lost 29 kg since surgery.

The patient presented to the emergency department with anorexia, vomiting, and diplopia for the previous 15 days. On a first observation, the patient was apyretic, presented exophthalmia, vertical, horizontal and rotatory nystagmus, bilateral convergent squint more evident at the left side, bilateral incomplete and asymmetric paresis of the III cranial nerve, bilateral complete paresis of IV cranial nerve, pupillary hippus, wide-based gait and posterior-anterior imbalance. At the emergency department, a cranial computed tomography (CT) with angiography revealed a hypodensity in the protuberance left paramedian

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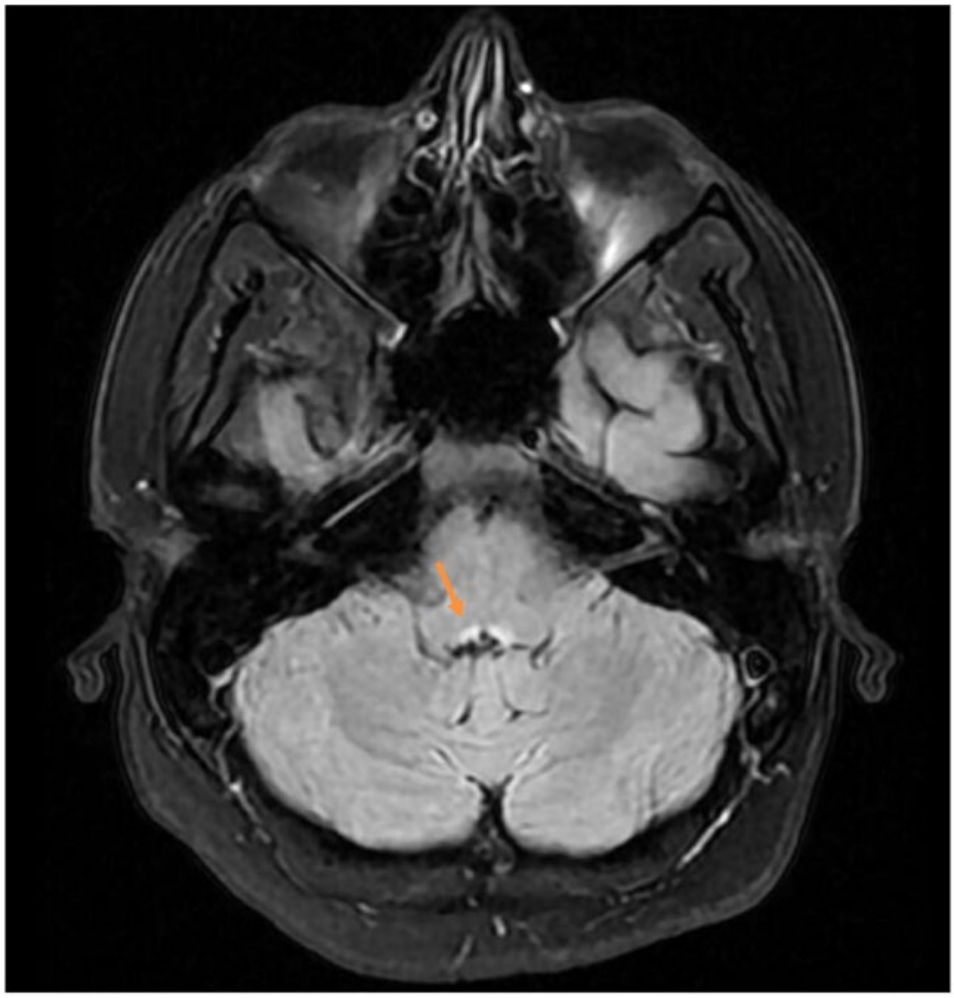

Figure 1: Magnetic resonance imaging (MRI) FLAIR of brain: bilateral hyperintensity in the periaqueductal region suggesting a metabolic lesion (arrow).

region with undetermined significance. Lumbar puncture revealed "*eau de roche*" cerebrospinal fluid, 2 lymphocytes/µL, 40 mg/dL proteins and 80 mg/dL glucose concentration (glycemia: 201 mg/dL). Due to persistent vomiting, patient had an abdominal CT and an upper endoscopy without suspicious findings. The patient was consequently admitted to a stroke unit for further investigation.

Brain MRI excluded cerebrovascular disease but showed a T2, FLAIR and SWAN bilateral hyperintensity in the periaqueductal region suggesting metabolic lesion (Fig. 1). No oligoclonal bands were found in the cerebrospinal fluid. The autoimmune study was negative (acetylcholine antibodies, antinuclear and extractable nuclear antigen antibodies). C and B hepatitis virus, human immunodeficiency virus, syphilis, cytomegalovirus serologies were negative and other infectious diseases were excluded. Sedimentation velocity was low, and it was never reported neutrophilia or neutropenia. Neurotropic virus, syphilis and bacterial screening of the cerebrospinal fluid was negative. Metabolic study found folate deficiency (1.5 ng/dL) and hypocholesterolaemia. Iron levels, thyroid hormones, B12, D, K and E vitamins were normal.

On the second day of hospitalization, after excluding other aetiologies, the patient began thiamine replacement (300 mg, 8/8 hours, intravenous) as a therapeutic trial. Symptoms improved after 48 hours. Before treatment, thiamine concentration was measured by blood sample and the results confirmed a thiamine deficiency, 22 ng/mL (reference value >26 ng/mL). Nonetheless, this sample was obtained some days after resume the recommended dietary plan, therefore overestimating the real thiamine concentration levels. Eight weeks after discharge, the patient improved but still presented with incomplete paresis of the right IV cranial nerve. Thiamine therapy was progressively decreased to 100 mg/day *per os*. Wernicke syndrome secondary to malnutrition was confirmed by the combination of symptoms, improvement after thiamine replacement, thiamine deficiency confirmed in blood sample, suggestive MRI findings and differential diagnosis exclusion.

Discussion

Thiamine deficiency is an underdiagnosed condition and differential diagnosis are wide.[4] In this case the patient was initially admitted in stroke unit due to brainstem stroke suspicion.

Many cases are not identified in life or have late diagnosis.[3] This delay compromises the beginning of treatment which leads to permanent neurological deficits or even death. The classical triad of symptoms (abnormal mental state, ataxia and ophthalmoplegia) is present only in 20% of patients.[3] Ophthalmoplegia is more frequent in non-alcoholic aetiologies.[1]

Current bariatric surgical approaches can cause several micronutrients deficiencies. Roux-en-Y-gastric bypass, laparoscopic adjustable gastric banding and jejunoileal bypass have a more profound impact on the absorption of essential vitamins as thiamine.[5] According to a systematic review from 2016, only 13 cases of Wernicke syndrome associated with sleeve gastrectomy were identified, although the authors believe most cases are underdiagnosed.[6] With increasing prevalence of bariatric surgeries, gastric sleeve became a common surgery. It can be related to thiamine deficiency, with the overall incidence estimated in 1.06 cases per 100 000 surgeries.[7] In 10% of the case descriptions, the patients reported non-compliance to the medication and dietary regimen.[8] In this case, the vertical gastrectomy did not compromise thiamine absorption, which occurs in duodenum. However, patient did not respect the dietary plan and committed many nutritional errors, provoking severe malnutrition. Eight weeks after discharge, the patient maintained some neurological deficits despite thiamine supplementation. Persisting symptoms can be due to a long period of malnutrition. Some authors describe persistent oculomotor abnormalities for months or life.[3] In western countries, malnutrition can be present in the absence of alcohol abuse, reminding health care professionals the need to be aware of other causes. European guidelines recommend having a high level of suspicion for all clinical conditions that could cause thiamine deficiency. Due to its overall safety, it is also suggested to start thiamine supplementation even if suspicion is low.[2] ■

Declaração de Contribuição

DD, AV, SL - Elaboração e redação do manuscrito
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Todos os autores aprovaram a versão final a ser submetida.

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