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WERNICKE'S ENCEPHALOPATHY AS A COMPLICATION OF CHAGAS MEGA ESOPHAGUS

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Abstract: The Wernicke-Korsakoff syndrome (WKS) is a potentially lethal disease that is associated with vitamin B1 (thiamin) deficiency. It is characterized by disordered mental status, memory deficits, psychosis, ophtalmoplegia and ataxic gait. The WKS is associated with alcohol use in about 90% of the patients. This report adds the possibility of developing vitamin B1 deficiency and WKS in patients with chagasic mega esophagus. The authors describe a case of a 63 year old man with grade IV chagasic mega esophagus. There was no past use of alcohol. He was admitted to the emergency unit of the Brasilia University Hospital with a history of acute diarrhea, altered mental status, disordered ocular movements, flaccid tetraparesis and abolished deep tendon reflexes. The plasma levels of thiamin were markedly reduced and in the magnetic resonance of the brain there were alterations usually found in acute Wernicke encephalopathy. This report highlights the possibility of considering patients with chagasic mega esophagus as at risk for developing vitamin B1 deficiency and WKS.

Key words: Encephalopathy, Wernicke, Chagas disease, thiamine.

Introduction

Wernicke's encephalopathy (WE) and Korsakoff's psychosis, witch is also known as Wernicke Korsakoff syndrome (WKS), is a neurological disease caused by a deficiency of thiamine. WKS is a chronic neurological condition that occurs frequently as consequence of WE, an acute disorder that requires urgent therapy (1, 2)

Thiamine pyrophosphate, the active form of thiamine, is a cofactor for several enzymes that metabolizes carbohydrates and it is required for the synthesis of acetylcholine. Thiamine is found in cereals and flour. It is absorbed in the duodenum, and the daily requirement depends on the metabolic rate (1).

Chagas Disease (CD) is prevalent in Brazil and one of its complications is mega esophagus. Patients in the severe stages of the disease (grades III and IV) can develop protein-calorie malnutrition and vitamin deficiencies, such as thiamine deficiency, as a result of inadequate food intake. There is no description in the medical literature of associations among CD, megaesophagus and WE It is necessary to consider thiamine deficiency one of the complications of CD (3).

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Case report

A 63 years old male farmer had been attended in the Brasilia's University Hospital with a four-day history of diarrhea and vomiting. He had attended a primary health service previously and had received treatment with intravenous fluids. After discharge he presente with acute mental confusion, inversion of the sleep-wake cycle, and visual hallucinations. There was no history of alcohol misuse. The patient had undergone partial gastrectomy as a treatment for peptic ulcer seventeen years previously. He had CD and grade IV megaesophagus (confirmed by gastrointestional endoscopy) and despite two surgical interventions he required a liquid diet.

On physical examination, the patient showed cachexia, stupor, confusion and temporal and spatial disorientation. There was an inadequate ocular convergence and horizontal and vertical nystagmus bilaterally. The patient was tetraparetic. There was generalized muscular hypotrophy and abesence of osteotendinous reflexes. Motor coordination could not be analyzed.

The serum concentration of thiamine, vitamin B12, vitamin B6 and folic acid were 16,8mcg/l (reference value – RV: 30-95), 134,6 mcg/l (RV: 130-900), 2,5 mcg/l (RV: 4-18) and 4,4 mcg/l (RV: 3-17), respectively. Magnetic resonance imaging (MRI) of the brain (figure) revealed abnormal hyperintensity of the periaqueductal white

matter, the periventricular region of the third ventricle and the mammillary bodies.

The treatment provided was an enteral diet and parenteral thiamine. The patient recovered from the acute confusion and ocular abnormality. However, three months after his admission, he contracted a pulmonary infection that caused his death.

Discussion

Wernicke Korsakoff syndrome is a potentially fatal complication of thiamine deficiency. WE is characterized by nystagmus, ataxic gait, altered conjugate eye movements, and confusion which is usually acute (4). The daily requirement for thiamine is approximately 0,5 mg per 1000 Kcal consumed. The higher the metabolic rate and intake of glucose, higher is the requirement for thiamine. The body reserve is approximately 30 to 50 mg, and the depletion occurs in 4 to 6 weeks if thiamine is not ingested daily (5).

Although it is associated most often with alcoholism, WE also occurs under conditions of poor nutrition caused by malabsorption, poor dietary intake, increased metabolic requirement, systemic malignancy or gastrointestinal surgery (6). Autopsy studies have revealed that the incidence of Wernicke lesions is higher than that predicted by clinical studies. In a neuropathological study of 131 cases of WE only 20% of the causes had been diagnosed clinically. This large discrepancy between the number of cases diagnosed clinically and those diagnosed pathologically can be attributed to the occurrence of subclinical episodes of Wernicke, given that only 16% of the patients have all the features of the classic triad. WE should be suspected in every patient with malnutrition or unbalanced nutrition who demonstrates an acute state of confusion, which is present in 80% of patients with WE (7).

Chagas disease is prevalent in Latin America, and 14 million Brazilians are infected by the parasite (8). Approximately 40% of these patients will develop the complications of chronic disease, such as megaesophagus. The disease is characterized by abnormalities in the motility and morphology of the esophagus and are caused by neuronal damages. The Chagasic megaesophagus is a frequent cause of dysphagia and can lead to severe nutritional consequences (3). In general, doctors tend to focus on the low intake of calories and protein, and fail to prescribe supplement of vitamins.

In the case reported herein, despite a previous diagnosis of grade IV Chagasic megaesophagus, the patient had not received nutritional therapy and he was not taking a vitamin supplement. Owing to the severe dysphagia, his daily calorie intake was low and his diet was deficient in thiamine. Gastrectomy is known to be a a cause of the thiamin deficiency however, in the case reported, gastrectomy had been performed 17 years previously, so this procedure cannot have been a factor. The dysphagia caused by CD was the responsible for the low intake of solid food, and hence for WE, rather than the surgery (9).

It is probably that the infusion of glucose given at the primary medical care center resulted in the manifestations of vitamin deficiency. This sequence of events generally occurs in patients with acute manifestations of WE. After an infusion of glucose, the higher metabolic requirement in cells that are depleted of thiamine causes brain damage, due to difficulties in the utilization of glucose as a metabolic substrate.

Imaging and laboratory examinations are not necessary for all patients suspected of WE and treatment should not be delayed. However, such tests might be necessary to support the diagnosis of WE and to reject other pathologies. Typical findings on MRI are areas of hyperintensity at T2 in the periaqueductal white matter, the periventricular region of the third ventricle, the medial thalamus and the corpus mammilares bodies (10).

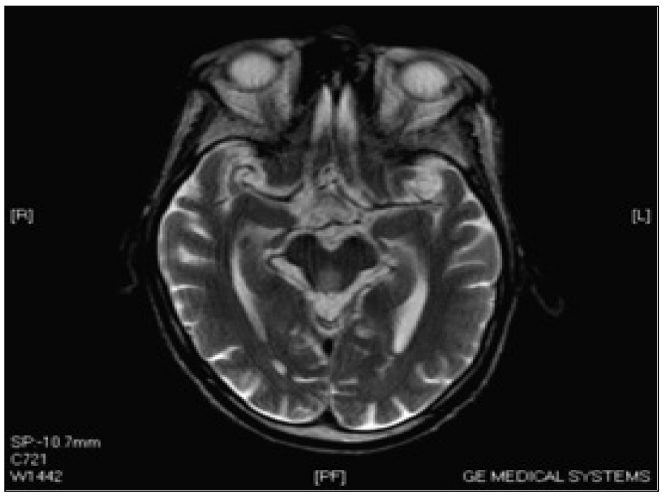

Figure 1. Magnetic resonance imaging (MRI) of the brain : abnormal hyperintensity of the periaqueductal white matter, the periventricular region of the third ventricle and the mammillary bodies

Conclusion

Wernicke's encephalopathy should be suspected in individuals with risk factors for thiamine deficiency and who present with one of the features of the classic triad. This report shows that it is necessary to consider the deficiency of micronutrients, especially thiamine, as one of the complications of Chagas disease.

Conflict of interest: There is no conflict of interest.

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