Wernicke encephalopathy and beriberi disease presenting as STEMI-equivalent

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Abstract

Thiamine deficiency is commonly associated with malnutrition, alcoholism and bariatric surgery. Thiamine deficiency can manifest in different ways, especially in developing countries: as peripheric neuropathy, as Wernicke encephalopathy or as beriberi disease. The authors present the case of a 72-year-old male, with a hiatal hernia that led to thiamine deficiency due to malnutrition. The initial clinical manifestation was an ST-elevation myocardial infarct equivalent, an ECG with a shark-fin pattern that evolved to a Wellens type B pattern. The patient evolved with severe altered mental status. A Wernicke encephalopathy diagnosis was confirmed by MRI; the patient was medicated with high-dose thiamine, with quick recovery, both neurologic and cardiac. The clinical history and response to treatment confirm the diagnosis of Wernicke encephalopathy and Beriberi disease.

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Introduction

Thiamine deficiency is commonly associated with malnutrition, alcoholism and bariatric surgery. It is very rare to be clinically significant in developed countries, where cereals and rice are supplemented with vitamins. Thiamine is a coenzyme required for glucose generation and for adenosine triphosphate (ATP) production in the Krebs cycle; thiamine is also important in propagating nerve impulses and maintaining myelin sheath, in a still unknow way [1]. The human body reserves of thiamine normally last 3 weeks; it is present in meat, vegetables, whole grains, and nuts. Classically, thiamine deficiency is associated with developing countries populations, where polished white rice (without the bran layer) and milled cereals are the primary food source [2]. Thiamine deficiency can manifest in different ways: as peripheric neuropathy, as Wernicke encephalopathy or as beriberi disease. The authors present a clinical case of an unusual manifestation of thiamine deficiency.

Case Report

A 72-year-old male, with known medical history of hypertension, former smoker, with esophageal diverticula, submitted to an esophageal surgery 20 years ago, was admitted to the General Surgery ward due to abdominal pain and anorexia with 3 weeks of evolution. The blood samples were unremarkable. The patient clinic was attributed to a large hiatal hernia: the patient was prescribed a diet without solid food and an esophageal surgery was scheduled at that hospitalization. A pre-op ECG was obtained, revealing sinus rhythm, with no abnormalities. At the 17th day of hospitalization, the patient presented with altered mental status and disorientation; the patient also complained of chest pain, nausea and sweating. At physical examination, he was hypotensive and tachycardiac, with marked bilateral edema. Arterial blood gas revealed a respiratory alkalosis with hyperlacticaemia. An ECG was performed: atrial rhythm, with ST elevation in the anterior wall (Figure 1). A few

minutes lateral, the ECG evolved, now revealing a dramatic ST elevation with QRS complex fused with T waves, in a pattern known as shark-fin, normally associated with acute coronary occlusion (Figure 2). Patient was given 5000 units of heparin and 250 mg of ASA. While under observation from Cardiology, the patient became asymptomatic and the ECG evolved again, now showing sinus rhythm with Wellens type B pattern (Figure 3). Blood samples revealed increased high sensitivity troponin I (1284 ng/L, normal range <34 ng/L). A transthoracic echocardiogram was performed, revealed preserved biventricular systolic function, with apical akinesia and hypokinesia of the medium segment of the inferior septum. As the patient was asymptomatic at that moment, it was interpreted as an acute myocardial infarct without persistent ST-segment elevation. The patient was admitted in the Cardiac ICU: while under observation, the patient presented again with changes in altered mental status: disorientation, aphasia, nystagmus and changes in sensitivity. Cerebral angio-CT did not show any acute events. No metabolic disorders were found. It was interpreted as a focal absence crisis and the patient was medicated with levetiracetam 1000 mg tid. A few hours later, patient had a Glasgow Coma Scale of 8, with Cheyne-Stokes breathing pattern, Babinski sign and nystagmus. He was hypothermic (34°C), hypotensive and tachycardiac. Arterial blood gas analyses showed aggravated respiratory alkalosis, with increased lactate (6 mmol/L). A cerebral and body CT was performed, not revealing any relevant new data. Lumbar punction revealed only mild protein increase, with augmented lactate. The patient was medicated with aggressive fluid therapy, lacosamide and clonazepam. An urgent cerebral MRI was requested: there was a diffuse, bilateral and symmetric areas of increased T2 signal in the thalamus, hypothalamus and mamillary bodies, highly suggestive of Wernicke encephalopathy (Figure 4). There were also recent ischemic injuries compatible with an embolic mechanism. The patient was medicated with high dose of thiamine (500 mg tid iv) and Levetiracetam, presenting a quick clinical response, with nearly complete resolution of the neurologic and cardiac symptoms. A diagnosis of Wernicke encephalopathy, caused by mal-nutrition and thiamine deficiency due to a hiatal hernia, was assumed. The ischemic cerebral injuries were attributed to the low- cardiac output status.

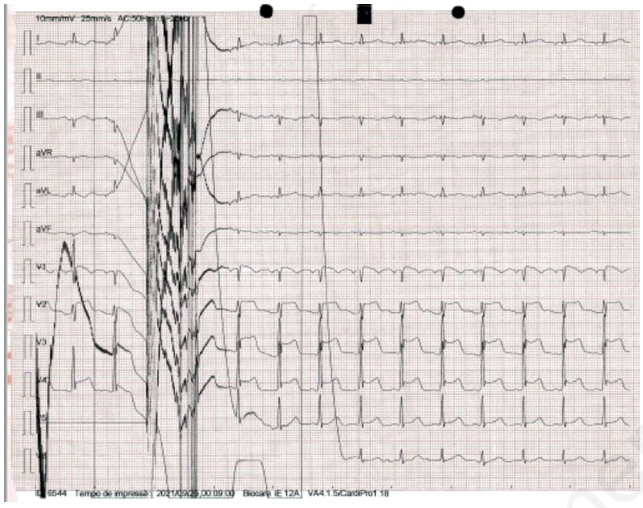

Figure 1. ECG revealing atrial rhythm, with ST elevation in the anterior wall.

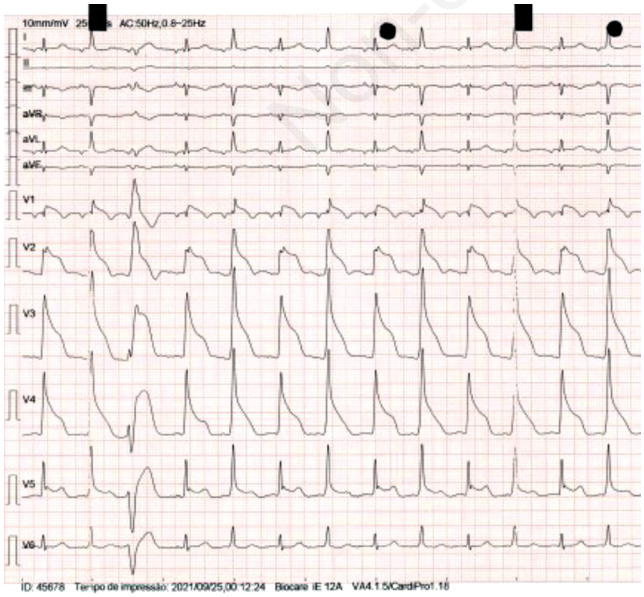

Figure 2. ECG revealing a shark-fin pattern: a dramatic ST elevation with QRS complex fused with T waves.

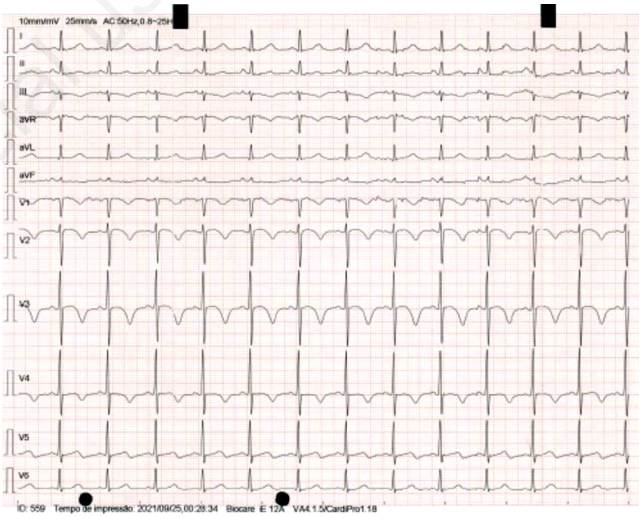

Figure 3. ECG revealing sinus rhythm with Wellens type B pattern.

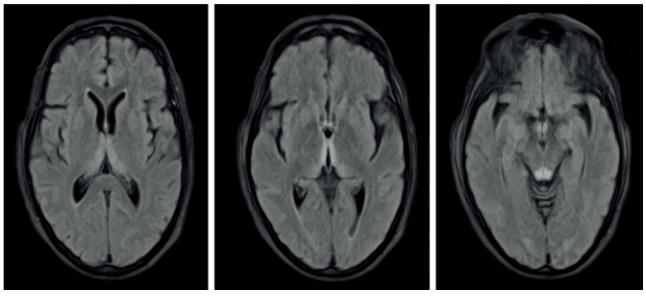

Figure 4. Cerebral MRI revealing a diffuse, bilateral and symmetric areas of increased T2 signal in the thalamus, hypothalamus and mamillary bodies, highly suggestive of Wernicke encephalopathy.

After clinical stabilization, the patient performed coronary angiogram, that showed one vessel coronary artery disease: 80% lesion on the medium segment of the right coronary artery, with non-obstructive disease of the left anterior descending and circumflex arteries. An echocardiogram was repeated, revealing preserved left ventricle systolic function and no wall motion abnormalities; of note, there were elevated left ventricle filling pressures. An ECG was also repeated: sinus rhythm, with no repolarization alterations. The patient was discharged with medical therapy for coronary artery disease and long-time supplementation for thiamine deficiency. During a 12 months follow-up, the patient was in NYHA functional class I and without angina. The patient is still waiting esophageal surgery. A cerebral MRI was repeated, showing nearly complete recovery. In retrospective, the most likely diagnosis to explain the cardiac clinic is Beriberi disease.

Discussion

Wernicke encephalopathy is the neurologic complication of thiamine deficiency. It is an acute syndrome that needs emergent treatment. It is commonly associated with alcoholism, malnutrition and gastric surgery. It typically presents as encephalopathy (altered mental status, profound disorientation, coma or even death), oculomotor dysfunction (specially with horizontal nystagmus) and ataxia. Less frequently, it can present with hypotension, hypothermia and peripheric neuropathy. The diagnosis is clinical: CT lacks sensitivity; MRI is the most sensible exam. Treatment consists of immediate high-dose iv thiamine in short-term, and low dose thiamine at long-term. Glucose and furosemide administration should be avoided; diuretics increase urinary thiamine excretion and glucose leads to increased consumption of thiamine [3]. Wernicke encephalopathy is associated with high mortality if untreated. The prognosis is good when treated; there is a very quick recovery; nevertheless, residual ataxia and memory loss are relatively common. Cardiac manifestations of Wernicke encephalopathy can occur. ECG changes are relatively common, mostly repolarization abnormalities, tachycardia, QT prolongation, low voltage. There are 2 known cases of ST elevation without coronary artery disease [4,5]: in both of them, resolution occurred with thiamine administration. None of the patients performed cardiac MRI (of note, the cases are from 2005 and 2008). The precise mechanism of the ST elevation is unknown.

Beriberi disease is typically described as Heart Failure with high cardiac output (due to decreased systemic vascular resistance). Patients normally present with tachycardia, elevated left ventricle filling pressures and pulmonary congestion. Nevertheless, the cardiac output can also be normal or low in one third of patients. Patients can also present with cardiomegaly, myocardial infarction with non-obstructive coronary arteries, coronary vasospasm or pulmonary hypertension [6]. It was previously common in oriental developing countries; since 1980 it is also very rare, even there. The first reports of beriberi disease in contemporary literature are attributed to Wenckebach [7]. The pathophysiologic mechanism of Beriberi is attributed to accumulation of adenosine and lactate (due to impaired production of ATP) and its direct vasomotor depression, with reduction in systemic vascular resistance and a compensatory high-output state [8]. There are no pathognomonic findings of Beriberi. Also, there are very few recent reports in developed countries, which adds to the difficulty in establishing an accurate diagnosis in severely ill patients, delaying treatment and potentially leading to misdiagnosis. In our patient, the diagnosis was established due to the altered mental status and Wernicke encephalopathy diagnosis; the good response to thiamine and the clinical context of hiatal hernia confirms the diagnosis. The patient's coronary artery disease (only affecting the right coronary artery) was interpreted as bystander and not as the culprit for the severe ECG changes, normally associated with left anterior descending artery occlusion. Thiamine dosing in whole blood testing was not performed in our patient. However, thiamine blood testing is rarely performed in the acute care setting; the result is often delayed and thiamine levels are not required for definitive diagnosis. Treatment should not be delayed [4]. The diagnosis criteria of beriberi disease were described by Blankenhorn [9] in 1946 and were the following: cardiomegaly in sinus rhythm; edema and elevated venous jugular pressure; peripheric neuropathy; minor ECG repolarization abnormalities (ST-T); no other apparent cause; clinical history of malnutrition; good response to thiamine or necropsy confirmation. One should not forget that the diagnostic capacity in 1946 was very different from nowadays; there was no echocardiography, no MRI, no coronary angiography and no high sensitivity troponin. In very severe cases, with hypotension, tachycardia, lactic acidosis and multiple organ dysfunction, it is called Shoshin beriberi. The treatment, in both cases, consists of immediate high-dose iv thiamine – the ideal treatment dosage is still not consensual [10]. The prognosis is also good, with a very quick recovery. A diagnosis of beriberi should be suspected in patients with unexplained cardiomyopathy and poor nutritional intake (even with Western diet) or alcoholism. The prevalence of thiamine deficiency in patients with heart failure is estimated to be 30% [11]; thiamine supplementation in these patients will likely be beneficial [12].

References

1. Wiley KD, Gupta M. Vitamin B1 thiamine deficiency. In: StatPearls [Internet]. Treasure Island: StatPearls Publishing; 2022.
2. Yamashita N, Aikawa T. Dutch research on beriberi: I. Christiaan Eijkman's research and evaluation of Kanehiro Takaki's diet reforms of the Japanese navy. Nihon Ishigaku Zasshi 2017;63:3-21.
3. Kuzume D, Inoue Y, Morimoto Y, et al. [Diuretics-induced beriberi polyneuropathy: a case report].[Article in Japanese]. Rinsho Shinkeigaku 2022;62:641-3.
4. Daly MJ, Dixon LJ, A case of ST-elevation and nystagmus--when coronary thrombosis is not to blame. QJM 2009;102: 737-9.
5. Kawano H, Koide Y, Toda G, Yanok. ST-segment elevation of electrocardiogram in a patient with Shoshin beriberi. Intern Med 2005;44:578- 85.
6. Coelho LS, Hueb JC, Minicucci MF, et al, Thiamin deficiency as a cause of reversible cor pulmonale. Arq Bras Cardiol 2008; 91:e7-9.
7. Wenckebach KF. [Das Beriberi-Herz: Morphologie, Klinik, Pathogenese].[Book in German]. Julius Springer: Berlin; 1934.
8. Akbarian M, Yankopoulos NA, Abelmann WH. Hemodynamic studies in beriberi heart disease. Am J Med 1966;41:197-212.
9. Blankenhorn MA, Vilter CF, Scheinker IM, et al. Occidental beriberi heart disease. J Am Med Assoc 1946;131:717-26.
10. Helali J, Park S, Ziaeian B, et al. Thiamine and heart failure: Challenging cases of modern-day cardiac beriberi. Mayo Clin Proc Innov Qual Outcomes 2019;3:221-5.
11. Hanninen SA, Darling PB, Sole MJ, et al. The prevalence of thiamin deficiency in hospitalized patients with congestive heart failure. J Am Coll Cardiol 2006;47:354-61.
12. Jain A, Mehta R, Al-Ani M, et al. Determining the role of thiamine deficiency in systolic heart failure: A meta-analysis and systematic review. J Card Fail 2015;21:1000-7.