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A Rare Case of Rapidly Progressing Beriberi Heart

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A 67-year-old man with schizophrenia was admitted to our hospital complaining of nausea and leg edema, and fell into a shock state the same day. The blood tests revealed high levels of the troponin I（5.2 ng/mL）and transthoracic echocardiography showed left ventricular（LV）dilatation and wall motion abnormality, he was suspected of acute heart failure caused by coronary artery disease. But coronary angiography showed no significant coronary arterial stenosis. The cardiac output was high and peripheral vascular dilatation occurred, he was treated supported by noradrenaline, but he passed away on the third day. He did not receive supplement of thiamine during this period without being given a diagnosis because he worsened rapidly. Myocardial necrosis and colliquative myocytolysis were observed in the autopsy. The thiamine level at admission was proven to be low, and was diagnosed as shoshin beriberi.

Keywords: beriberi heart, shoshin beriberi, thiamine, heart failure, colliquative myocytolysis

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INTRODUCTION

Thiamine is a water-soluble vitamin that plays an important role in carbohydrate and lipid metabolism. A thiamine deficiency can cause neuritis as well as cardiac beriberi, which is characterized by lactic acidosis, peripheral arterial dilation, and high-output heart failure［1］.

Shoshin beriberi is a fulminant form of cardiac beriberi, in which rapid deterioration occurs in a few hours, and may lead to a fatal outcome unless diagnosed and treated in the early stage［2］. However, the diagnosis of shoshin beriberi is often complicated by its rapid progression.

The causative factors of cardiac beriberi were common during the Edo period, in which the habit of eating polished white rice was prevalent. This disease is now relatively rare, but occurs sporadically in cases of alcoholism and severe dietary restrictions, and following a gastrectomy. We report here a case of shoshin beriberi in a patient who had a poorly balanced diet.

CASE REPORT

A 67-year-old man with a history of schizophrenia presented to the emergency outpatient clinic, complaining of lower leg edema and nausea for 2 days prior to admission. The state of schizophrenia was stable, but he remained unmarried placed in care home. Meals were offered, but he seldom ate vegetables. He had a smoking history（30 cigarettes a day）, but no intake of alcohol. There was no diabetes or hypertension in the past history and he did not take any medicine including supplement.

On admission, he was alert, with pitting edema of the lower extremities. His body mass index was 18.7 kg/m^2, vital signs included a blood pressure of 95/46 mmHg, heart rate of 86 beats per minute,

peripheral oxygen saturation of 99% with room air, and body temperature of 36.5°C. No heart murmurs or rales were audible. In the physical examination, the findings to suggest infection were absent. An electrocardiogram revealed sinus rhythm, complete right bundle branch block, and abnormal Q waves in lead III (Fig. 1). In chest X-ray, the cardiothoracic ratio was 60.2% and mild pulmonary congestion was observed. The blood tests revealed high levels of the troponin I (5.2 ng/mL), creatine kinase (470 IU/L), creatine kinase-myocardial band (43 IU/L), and creatinine (1.8 mg/dL); the brain natriuretic peptide level was 98 pg/mL and hemoglobin level 12.0 g/dL. There was no increase in the white blood cell count (7,450 /μL) or eosinophil count (52.1 /μL) or C-reactive protein (0.64 mg /dL), and the thyroid function was within normal range. Blood gas analysis was not performed at the time of admission.

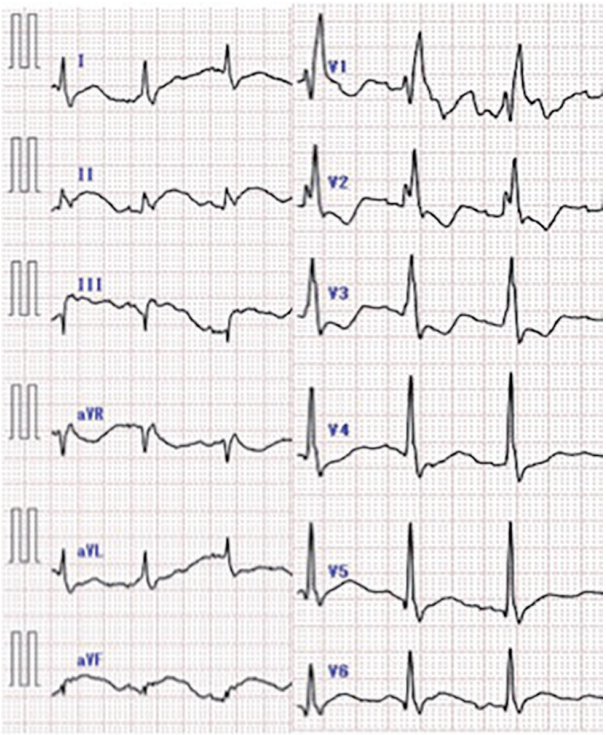

Fig. 1. An electrocardiogram on admission revealed sinus rhythm, complete right bundle branch block, and abnormal Q waves in lead III.

Transthoracic echocardiography (TTE) revealed a LV end-diastolic dimension of 59 mm, end-systolic dimension of 44 mm, hypokinesis in the basal segment of the inferior wall and mid-apical segments of the anterior wall, and LV ejection fraction was 50%. The transmitral flow and pulmonary venous flow patterns demonstrated an E-wave velocity greater than the A-wave (E > A), and S-wave velocity lower than the D-wave (S < D), respectively, and mild pulmonary hypertension was observed (tricuspid regurgitation pressure gradient = 36 mmHg). These findings suggested an increase in the LV end-diastolic pressure. TTE also showed a decreased respiratory variation in the inferior vena cava diameter (24 mm with inspiration, and 28 mm with expiration), and a minimal pericardial effusion. No intracardiac shunt was detected.

He was suspected of acute heart failure caused by coronary artery disease, elective coronary angiography (CAG) was planned because he had no ST segment elevation or chest symptoms. On the night of admission, he developed a decreased blood pressure (systolic blood pressure 60 mmHg), but blood pressure did not respond to dopamine. On day 2 of admission, an emergency CAG performed after the placement of an intra-aortic balloon pump showed no significant coronary stenosis (Fig. 2). Therefore, acute coronary syndrome was ruled out. A right heart catheterization performed immediately after the CAG revealed a pulmonary capillary wedge pressure of 21 mmHg, cardiac output of 7.91 L/min, and cardiac index of 4.63 L/min/m^2; the systemic vascular resistance was 384 dyne·ssec·cm^{-5} (normal range: 1170 ± 270 dynes·sec·cm^{-5}) and systemic vascular resistance index 656 dyne·sec·cm^{-5}·m^2 (normal range: 2130 ± 450 dyne·sec·cm^{-5}·m^2). The results of the right heart catheterization suggested high-output cardiac failure with peripheral vasodilation with a high lactate level on day 2 (5.0 mmol/L). Because of LV dilatation and wall motion abnormality in addition to this hemodynamics, we couldn't deny sepsis-induced cardiomyopathy. The patient was started on noradrenaline and a large-volume fluid replacement in combination with an antibiotic (meropenem 1 g every 12 h) and cytokine adsorption therapy as sepis, but he had persistent shock. The patient was given intravenous hyperalimentation during this period, but thiamine was not included, and there was not the oral intake because tracheal intubation was done by disturbance of consciousness and shock to be prolonged. An

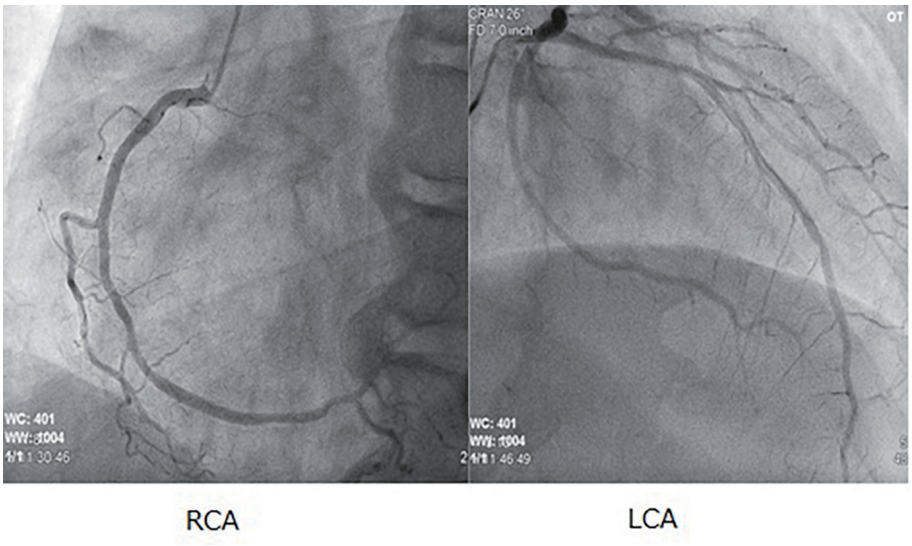

Fig. 2. Coronary angiography showing no significant stenosis in the coronary artery.

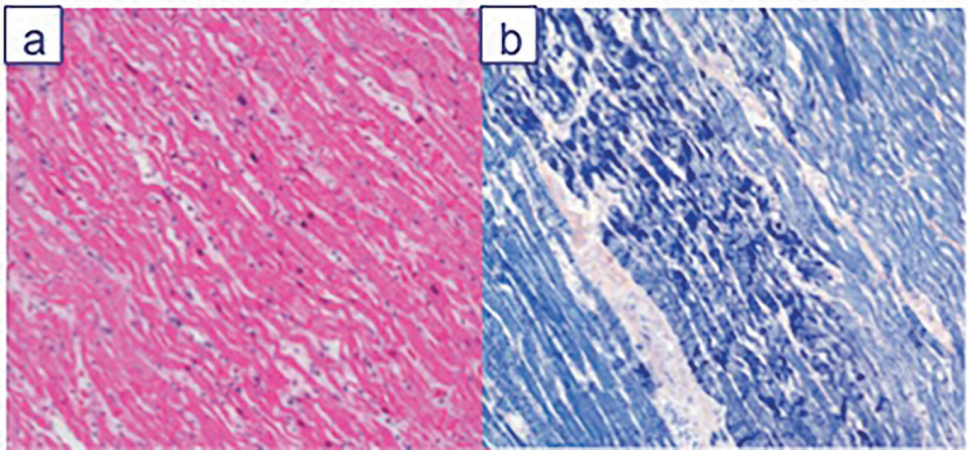

Fig. 3. Autopsy findings
(a) A lot of coalescent fresh colliquative myocytolysis was found in left ventricle. There were few inflammatory cell infiltrates in the necrotic part. Hematoxylin-eosin stain, ×100.
(b) Most of the necrosis was equivalent to contraction band necrosis, and only a little colliquative necrosis was found. Phosphotungstic acid-hematoxylin stain, ×100.

increase in lactic acid concentration (20 mmol/L) and pyruvic acid concentration (2.01 mg/dL) was observed on day 3 of admission.

The patient declined receiving any percutaneous cardiopulmonary support, and died on day 3. It turned out later that the blood and urine culture tests as well as procalcitonin and β-D-glucan tests were negative. No other findings during the course of the treatment suggested a definite infection.

Autopsy revealed a large volume of focal-to-confluent fresh coagulation necrosis in the LV, most of which was identical to contraction band necrosis accompanied by colliquative myocytolysis; there were no characteristic findings of acute myocarditis, including lymphocytes or eosinophil infiltration (Fig. 3). The patient was later found to have had a low level of thiamine (8 ng/mL) in the drawing blood on day 2. Based on these findings and his clinical course, the patient was diagnosed with shoshin beriberi.

DISCUSSION

In Japan, beriberi is one of two main folk illnesses, along with tuberculosis, observed from the Edo to early Showa periods, during which the habit of

eating polished white rice was prevalent. Thereafter, the incidence of beriberi dramatically decreased in association with the Westernization of dietary habits. Although attention has again been focused on beriberi due to an increased number of people whose diets are excessively restricted or poorly balanced or associated with alcoholism, the incidence of this illness still remains low.

Thiamine is a coenzyme required for the metabolism of pyruvate into acetyl-CoA. Thiamine deficiency leads to the accumulation of pyruvate and lactate, causing metabolic acidosis. In the cardiovascular system, this deficiency is associated with increased arteriovenous shunting, microcirculatory changes, and a decreased peripheral vascular resistance with decreased cerebral and renal blood flow. This leads to increases in the hydrostatic pressure and sodium retention, and therefore an increase in edema and the venous return, resulting in high-output cardiac failure [3]. In the present patient, a marked decrease in the peripheral vascular resistance and high-output cardiac failure occurred, with an increased lactate level and high pyruvate level within 3 days of admission.

Histopathological features in patients with cardiac beriberi include myocardial necrosis as well as colliquative myocytolysis [4]. Colliquative myocytolysis is also observed in patients with diseases presenting with congestion such as myocardial infarctions and myocarditis. In the early stage, cardiac beriberi is reversible, and is characterized by myocardial hypertrophy, vacuole formation, and stromal edema. In the chronic stage, the disease is irreversible, and is characterized by progressive fibrosis and cellular infiltration [5]. Acute myocarditis is characterized by stromal infiltration of inflammatory cells including lymphocytes and eosinophils, and degeneration and necrosis of adjacent cardiomyocytes [6]. Myocardial necrosis and colliquative myocytolysis are pathological findings of shoshin beriberi [4], our case does not conflict with shoshin beriberi.

The present patient was diagnosed with shoshin beriberi based on his medical history and the presence of a low thiamine level, lactic acidosis, high-output cardiac failure with peripheral vasodilation, and colliquative myocytolysis of the myocardium without inflammatory infiltration. Sepsis-induced cardiomyopathy also has a decrease in LV ejection fraction with asynergy, and a rise in troponin I [7], but in this patient, other findings suggesting sepsis were not observed and was negative.

Treatment of shoshin beriberi consists of thiamine replacement alone. In previously published case, none of patients diagnosed with shoshin beriberi was recovery without thiamine supplementation [4, 8, 9]. Initiation of appropriate thiamine replacement in patients with shoshin beriberi reportedly produces dramatic improvement in the hemodynamics as well as normalization of the systemic vascular resistance within 30 to 90 min after the treatment [10]. Blacket *et al*. reported that thiamine improved the mean pulmonary artery wedge pressure (from 16 to 5 mmHg) and mean pulmonary arterial pressure (from 28 to 14 mmHg) [11]. If you suspect shoshin beriberi, it is important to administer thiamine as early as possible.

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None

CONFLICTS OF INTEREST

The authors declare that there is no conflict of interest.

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