Reversible Pulmonary Hypertension, Lactic Acidosis, and Rapidly Evolving Multiple Organ Failure as Manifestations of Shoshin Beriberi

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Figure 1. (A) Twelve-lead electrocardiogram on admission. ST-segment depression was found in leads II, III, aVF, V5, and V6, and negative T-waves were found in leads V1–5. (B) Chest X-ray on admission and 1 week after admission. A prominent pulmonary trunk (arrow), moderate cardiomegaly, and decreased vascular markings were found on admission, and those changes improved 1 week after admission. (C) Color and continuous-wave Doppler echocardiography on admission. Moderate tricuspid regurgitation was found (Upper), and the estimated systolic pulmonary artery pressure determined by obtaining the maximum velocity of moderate tricuspid regurgitation on continuous-wave Doppler echocardiography was 70 mmHg (Lower). PA, pulmonary artery; RA (V), right atrium (ventricle).
25-year-old man presented to the emergency department with a 2-day history of progressive dyspnea and fatigue, along with generalized weakness, loss of appetite, and slight fever, and had been in an oliguric state for 12h. He had no history of medication or alcohol use. On admission his body temperature was 34.8°C, and he had clammy, cyanotic, and icteric skin, a regular tachycardia (108 beats/min), hypotension (76/55 mmHg), tachypnea (50 breaths/min), and reduced oxygen saturation (90%). He had jugular vein distension and the pulmonic second heart sound was accentuated. Prominent pitting edema was observed in both lower extremities. Electrocardiogram (ECG) demonstrated sinus tachycardia, with ST-segment depression in leads II, III, aVf, V5, and V6, and negative T-waves in leads V1–5, and chest X-ray showed a prominent pulmonary trunk, moderate cardiomegaly, and decreased vascular markings (Figures 1A, B). Echocardiography showed marked right atrial and ventricular dilatation and flattening of the interventricular septum with a D-shaped deformation of the left ventricle in later systole and throughout diastole, indicating severe right ventricular overload (Figure 2A; Supplementary Movie 1). The absence of RV hypertrophy with an end-diastolic RV free wall thickness of <5 mm indicated acute progression of RV overload (Figure 2A; Supplementary Movie 1). Moderate tricuspid regurgitation was found, and the estimated systolic pulmonary pressure determined on continuous-wave Doppler echocardiography was 70mmHg (Figure 1C). The left ventricular (LV) end-diastolic internal dimension was 44 mm, and the LV ejection fraction was 63%. The cardiac output was observed to be increased at 7.8L/min on Flo Trac™ sensor (Edwards Lifesciences, Irvine, CA, USA), indicating hyperdynamic circulation.

Blood gas indicated pronounced lactic acidosis with an arterial pH of 6.905, pCO₂ of 12.6 mmHg, pO₂ of 158.4 mmHg, HCO₃ of 2.8 mmol/L, lactate of 27 mmol/L, and base excess of –27.2 mmol/L. On laboratory investigation, aspartate aminotransferase was 439 U/L, alanine aminotransferase was 356 U/L, lactate dehydrogenase was 1,280 U/L, total bilirubin was 5.8 mg/dl, %prothrombin time was 25%, ammonia was 209 μg/dl, blood urea nitrogen was 18.9 mg/dl, creatinine was 2.03 mg/dl, creatine phosphokinase was 729 U/L, with a muscle brain fraction of 48 U/L, troponin-T 0.219 ng/ml (normal <0.1 ng/ml), C-reactive protein 0.47 mg/dl, and pro-B type-natriuretic peptide level 31,102 pg/ml (normal <125 pg/ml), demonstrating renal and hepatic failure. Thyroid function was within normal limits, and serology for B and C hepatitis was negative. Acute progression of the episodes and severe pulmonary hypertension suggested that the renal and hepatic failure were the result of circulatory collapse.

The patient became drowsy shortly after arrival. Because of the presence of severe lactic acidosis with a consciousness disturbance, the emergency physician first suspected a neurological disorder due to a thiamine deficiency. Thiamine (100 mg) was immediately administered transvenously, and a vasoactive agent (dobutamine 5 μg·kg⁻¹·min⁻¹) was also administered to support the perfusion. Doppler examination

![Figure 2. Two-dimensional echocardiography. Note (A) the marked right atrial and ventricular dilatation and flattening of the interventricular septum with D-shaped deformation of the left ventricle (short-axis view) in later systole and throughout diastole on admission, indicating severe right ventricular overload. (B) Those changes disappeared 7 days after admission. LA (V), left atrium (ventricle); RA (V), right atrium (ventricle).]
of the lower extremities was normal. He continuously received a daily dose of 100 mg of thiamine.

The lactic acidosis and body temperature improved during the initial 12 h followed by a blood pressure and urine output increase within the next 24 h. He completely recovered from the multiple organ failure within 2 days, and all the laboratory data also gradually improved. Ventilation–perfusion scan performed on the second day was normal. On repeat echocardiography 7 days after admission, the pulmonary hypertension had disappeared (Figure 2B; Supplementary Movie 2). The patient was found to be a vegetable refuser when his history was taken from his family. During the 22-day admission the patient lost 23 kg and was discharged from the hospital without the need for medications. He has done well under thiamine supplements during an 11-month follow-up period.

Shoshin beriberi, a subtype of beriberi involving the cardiovascular system, is appropriately designated as “a rapidly curable hemodynamic disaster”. It is characterized by hypertension, tachycardia, and lactic acidosis. Various ECG abnormalities, including biphasic or inverted T-waves, ST-segment elevation, and QT prolongation, elevation of the serological markers of myocardial injury, and multiple organ failure can occur in this setting. Severe pulmonary hypertension can also occur due to an increased pulmonary arterial blood flow and elevated LV end-diastolic pressure as in the present case. These ECG changes and acute progression of the pulmonary hypertension may require a differential diagnosis of pulmonary embolism.

The diagnosis of shoshin beriberi is always difficult because of its rarity, constraints of the laboratory facilities, and lack of available tests in the emergency situation. A standard laboratory diagnosis of thiamine deficiency is made by assessing the erythrocyte transketolase activity at baseline and severe pulmonary hypertension quantitatively and precisely. This modality is also handy, and could be performed in the emergency room or at the patient’s bedside. Quick and serial echo measurements are helpful for diagnosis, treatment, and follow up, as in the present case.

References
2. Park JH, Lee JH, Jeong JO, Seong IW, Choi SW. Thiamine deficiency as a rare cause of reversible severe pulmonary hypertension. *Int J Cardiol* 2007; **121**:e1–e3.
5. Tran HA. A 74-year-old woman with increasing dyspnea. Wet beriberi with fulminating (Shoshin) cardiac failure and elevated troponin I. *Arch Pathol Lab Med* 2006; **130**:e8–e10.

Supplementary files

**Movie 1.** Two-dimensional echocardiography (short axis view) on admission.

**Movie 2.** Two-dimensional echocardiography 1 week after admission. Please find supplementary file(s); http://dx.doi.org/10.1253/circj.CJ-10-0202