Shoshin Beriberi With Low Cardiac Output and Hemodynamic Deterioration Treated Dramatically by Thiamine Administration

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SUMMARY

“Shoshin beriberi”, which is a fulminant form of cardiovascular beriberi accompanied by hemodynamic deterioration with high cardiac output and decreased systemic blood pressure, caused by thiamine deficiency due to alcoholic abuse or malnutrition, is often difficult to address because of its rarity and non-specific symptoms. We here present a patient with a history of alcoholic abuse who had suffered hemodynamic deterioration with extremely low cardiac output refractory to extracorporeal membrane oxygenation and intravenous catecholamine support, which was improved dramatically by bolus intravenous thiamine administration. Such a type with low cardiac output would be the most severe form of Shoshin beriberi, and cannot be rescued without diagnostic administration of thiamine. (Int Heart J 2015; 56: 568-570)

Key word: Vitamin B1, Alcohol, Heart failure

Cardiovascular Beriberi” is typically accompanied by high cardiac output and decreased systemic blood pressure, which are caused by thiamine deficiency due to alcohol abuse or malnutrition. Although the disease has been studied from ancient times, it is often difficult to diagnose because of its rarity and non-specific symptom. Therefore, “beriberi” is an old but also new current disease, which we should re-focus on.

“Shoshin beriberi” is a fulminant form of cardiovascular beriberi accompanied by hemodynamic deterioration. However, little has been reported about this type of beriberi. We here report a patient with shoshin beriberi who was successfully saved by the rapid administration of thiamine.

CASE REPORT

A 40-year-old man with no specific medical history was admitted to our hospital by ambulance complaining of dyspnea, and presented with a heart rate of 108 bpm, systolic blood pressure of 70 mmHg, and oxygen saturation of peripheral artery of 84% under 10 L/minute of facial mask oxygenation. Chest X-rays showed cardiomegaly with a butterfly shadow pattern, and an electrocardiogram indicated right ventricular (RV) overload (Figure 1AB). Serum levels of total bilirubin 3.9 mg/dL, creatinine 2.8 mg/dL, and lactate 20.4 mmol/L were assayed, and the plasma B-type natriuretic peptide (BNP) concentration was 995 pg/mL on admission. Transthoracic echocardiography showed a left ventricular ejection fraction (LVEF) of 30% along with dilatation of the RV cavity. Hemodynamic study demonstrated cardiac output (CO) of 2.8 L/minute and systemic vascular resistance (SVR) of 400 dyn·sec·cm⁻5.

We excluded the following diagnoses: ischemic heart disease by percutaneous coronary angiography, pulmonary embolism by enhanced computed tomography, and myocarditis by endomyocardial biopsy without any infiltration (Figure 2A-C). Septic shock was excluded considering the laboratory data: white blood cell count of 8700/μL and C-reactive protein level of 1.4 mg/dL. Adrenal insufficiency was also excluded considering the laboratory data: serum levels of sodium and potassium of 136 mEq/L and 4.5 mg/dL and blood sugar level of 97 mg/dL. Anaphylactic shock was not aggressively suspected because of the absence of other cutaneous, digestive, and respiratory symptoms.

Considering the persistent hemodynamic deterioration refractory to extracorporeal membrane oxygenation (ECMO) support (Figure 3) and the additionally obtained social history of his alcoholic abuse, ie, 200 g/day of alcohol intake during the past 20 years, which was elucidated after the initiation of ECMO for the first time, “shoshin beriberi”, the fulminant form of cardiovascular beriberi caused by thiamine deficiency, was suspected.

After the first intravenous administration of thiamine of 100 mg at day 2, his systolic blood pressure increased dramatically along with rapid normalization of the lactate concentration, accompanied by withdrawal of all catecholamine infusion and ECMO support during daily intravenous administration of 200 mg of thiamine. His hemodynamics remained normal, and he was discharged from the intensive care unit on day 10 rep-
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Discussion

We experienced a middle-age male patient with a history of alcoholic abuse, who was diagnosed as having "shoshin beriberi" with acute hemodynamic deterioration accompanied by low CO and decreased SVR refractory to ECMO support, which was dramatically improved by the introduction of thiamine.

The major cause of beriberi is thiamine deficiency. Thiamine is a precursor of the thiamine pyrophosphate cofactor used in several steps in carbohydrate metabolism pathways, such as the Krebs citric acid cycle. Thiamine deficiency inhibits the function of the citric acid cycle and the hexose monophosphate shunt, resulting in insufficient oxygenation of the peripheral tissues and lactic acidosis.

Diagnosis of beriberi is often difficult because of its rarity and non-significant symptoms. The administration of thiamine is the most recommended procedure to diagnose and treat beriberi. A patient history of alcoholic abuse or malnutrition is a key to suspecting the disease, but an immediate history taking is often difficult because of consciousness disorder in patients with shoshin beriberi, as was the case with our patient. Although endomyocardial biopsy is useful to exclude other specific diseases such as myocarditis, there are no specific features indicating beriberi. A decrease in the blood concentration of vitamin B1 is a key to diagnosing beriberi, but the level of vitamin B1 sometimes is not necessarily exceedingly low as in the present patient with beriberi. Moreover, it takes days to obtain the results of these procedures, which is too late to rescue patients with hemodynamic deterioration.

Alcohol abuse, malnutrition including continuous intake of polished rice, dialysis, and long-term use of diuretics are the major causes of thiamine deficiency. The relation between alcohol abuse and thiamine deficiency is 1) malnutrition accompanied by alcohol abuse, 2) enhanced urinary excretion of magnesium, which is cofactor of thiamine, and 3) blockage of thiamine absorption through the ileum. Moreover, it takes days to obtain the results of these procedures, which is too late to rescue patients with hemodynamic deterioration.

The decrease in SVR and elevated lactate level in the present patient were compatible with previous reports. Deficiency of thiamine injures the ability to vasoconstrict, which leads to decreased SVR and systemic blood pressure. Lactic acidosis results from 1) inhibition of oxidative decarboxylation secondary to thiamine deficiency, 2) hyperventilation, and 3) regional hypoxia.

Although most cardiovascular beriberi represents high CO to compensate for decreased SVR, a few patients with shoshin beriberi suffer hemodynamic deterioration following low CO. The precise mechanism remains unknown, but di-
rect injury of the myocardium by thiamine deficiency may play a critical role.\textsuperscript{19} The reason why thiamine deficiency results in such various phenotypes should be studied in the future.

**REFERENCES**

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