Shoshin beriberi: a rare diagnostic problem

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Summary A 30 year old man with a 15 year history of alcohol abuse presented with symptoms and signs of circulatory shock, severe disturbances of renal and liver functions, and metabolic acidosis. The cardiovascular and metabolic features were attributable to Shoshin beriberi. He recovered completely after treatment with thiamine.

Cardiovascular beriberi is caused by thiamine (vitamin B₁) deficiency and usually presents as predominantly right sided heart failure associated with a high cardiac output or less frequently as cardiovascular collapse, a condition known as Shoshin beriberi or acute pernicious beriberi.¹² The high cardiac output is caused by peripheral vasodilatation.³

Case report

A 30 year old man with a 15 year history of alcohol abuse abruptly stopped drinking because of nausea and general debility. Two weeks later he was admitted in a somnolent state after several episodes of circulatory collapse. On examination his blood pressure was 80/40 mm Hg and rectal temperature 36.8°C; he had tachypnoea, peripheral cyanosis, and jugular venous distension at the angle of the jaw. His pulse rate was regular at 80/min. An ejection systolic murmur grade 1/6 was heard in the third left interspace; the lung fields were clear. The liver was palpable 2 cm below the right costal margin; some tibial oedema was present. Relevant laboratory results showed: haemoglobin 13.5 g/dl, haematocrit 38%, erythrocyte sedimentation rate 3 mm in the first hour, and white blood cell count 9.7×10⁹/l with a normal differential count. The serum urea concentration was extremely high (90.4 mmol/l; 5.4 g/l) as were the serum creatinine (587 μmol/l; 66.3 mg/l) and potassium (7.0 mmol(mEq)/l) concentrations. Serum sodium (129 mmol(mEq)/l) and chloride (89 mmol(mEq)/l) concentrations were low. Enzyme activity of aspartate aminotransferase (560 U/l), alanine aminotransferase (400 U/l), and lactate dehydrogenase (926 U/l) was increased. Arterial blood gases while breathing room air were compatible with metabolic acidosis: pH 7.29, Pco₂ 2.1 kPa, base excess -16 mmol/l, and bicarbonate 8 mmol(mEq)/l with a Po₂ value of 16.5 kPa.

A chest radiograph showed an enlarged heart with mild pulmonary venous congestion, and the electrocardiogram sinus rhythm with non-specific ST segment changes. The patient was given symptomatic treatment to correct the acidosis and hyperkalaemia. Dopamine (up to a dose of 10 μg/kg/min) was given intravenously, which resulted in an increase in pulse rate to 120/min but a fall in blood pressure to 70/40 mm Hg. In view of the known alcohol abuse 100 mg thiamine was injected intravenously. A few hours later his blood pressure had risen to 95/40 mm Hg, and a high cardiac output state developed with bounding pulses and pistol shot sounds as well as Duroziez's sign over the femoral arteries. The cyanosis disappeared. At cardiac catheterisation using a Swan-Ganz thermodilution catheter the high cardiac output was confirmed and the peripheral vascular resistance was found to be low (Table). The patient was treated with thiamine and a sodium restricted diet. The cardiac output and peripheral resistance gradually returned to normal values, but right atrial and pulmonary wedge pressures were still raised on the third day (Table) when the Swan-Ganz catheter had to be removed because of a temperature rise to 39°C.

Within a week of admission the electrocardiogram and renal and liver function tests returned to normal. The chest radiograph showed no pulmonary congestion after two weeks, and the heart size returned to normal a few weeks later. Erythrocyte transketolase activity before treatment was 4.3 U/mmol (normal 8.8-12.5 U/mmol) of haemoglobin and the thiamine pyrophosphate effect was 22% (normal 5-25%). On retesting four weeks later the transketolase activity was 9.8 U/mmol of haemoglobin and the thiamine pyrophosphate effect 3%.

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Discussion

The two most important criteria in diagnosing cardiovascular beriberi are a high cardiac output with predominantly right sided heart failure and recovery after treatment with thiamine.4 The syndrome is easily recognisable in the oriental type of beriberi in which the thiamine deficiency results from the patient eating polished rice.5,6 In the occidental type of beriberi, which is found almost exclusively in alcoholics eating a thiamine deficient diet, however, a high cardiac output may be absent1,7 and left sided heart failure may often be found in the early clinical stages.7,8 Thiamine treatment sometimes results in a paradoxical lowering of the cardiac output.9,10 It has been suggested that the differences between the two types of beriberi may be attributable to alcoholic cardiomyopathy.6,7,11

Blacket and Palmer have shown that the high cardiac output results from dilatation of the arterioles of the skeletal musculature, which induces a compensatory vasoconstriction in other areas such as the hands and the kidneys.1 In cases of Shoshin beriberi, in which peripheral resistance may be extremely low,1,2 this may lead to peripheral cyanosis and severe disturbances of renal function.

A rise in pyruvate and lactate concentrations, caused by a thiamine deficiency induced block in carbohydrate metabolism, produces metabolic acidosis. The accompanying tachypnoea results in a high arteriovenous Po2, which is an unusual finding in the presence of peripheral cyanosis.

Thiamine deficiency may be diagnosed on the basis of the so called thiamine pyrophosphate effect, which occurs when the activity of the thiamine pyrophosphate requiring enzyme transketolase of erythrocytes is low and rises sharply after thiamine pyrophosphate is added in vitro.12 Nevertheless, depressed transketolase activity without a high thiamine pyrophosphate effect—as in our patient—may occur in alcoholics with liver disease and in patients with longstanding thiamine deficiency.13

References

1 Majoor CLH, Hillen HFP. Cardiale beriberi met melkzuur-acidose en cardiovasculaire collaps (Shoshin), een bij alcoholici niet zeldzaam ziektebeeld dat gemakkelijk wordt miskend. Ned Tijdschr Geneeskd 1982; 126: 749–57.