CARDIAC BERI-BERI PRESENTING WITH SHOCK

By Dhanwant Singh

SYNOPSIS

Two cases of cardiac beri-beri with shock are described. This is an unusual mode of presentation. The possible mechanisms for the shock are discussed.

INTRODUCTION

Though very rare cardiac beri-beri is still seen in Singapore. Because of its rare occurrence and atypical presentation at times, the diagnosis is often not entertained.

Cardiac beri-beri as documented in standard texts and journals presents usually as a high-output cardiac failure (Blankenhorn M.A., 1955; Wagner P.I., 1965; Akbarian M. *et al*, 1966; Robin E. *et al*, 1970). Sukumalchantra Y. *et al* (1976) recently described 6 cases of cardiac beri-beri, two of which had a low or normal output. Robin E. *et al* (1970) described a case of thiamine deficiency with a high-output cardiac failure which after therapy developed persistent low-output due to an associated. underlying cardiomyopathy. Blankenhorn (1945) stated that in late cases of cardiac beri-beri it was not unusual to find very low blood pressures and a 'shock-like' state. The response of these to thiamine was generally unsatisfactory and advocated that such cases should not be subjected to undue physiological studies.

CASE REPORT

Case 1

A 20 year old Indian female was admitted on 20th June, 1975 with a history of acute onset of breathlessness. She gave a history of having had a non-specific high fever for 1 week prior to the onset of breathlessness. She was a widow with four children and worked as a labourer.

She was known to consume toddy intermittently and had a poor nutritional history.

On admission she was acutely ill with marked hyperventilation. She appeared agitated, restless and confused. The pulse rate was 140/min. with a presence of pulses paradoxus. The blood pressure was 60/40. In the heart there was marked tachycardia, an S_3 and a great 3/6 systolic murmur along the left sternal edge. In the lungs there were bilateral crepitations. The neck veins were engorged and the liver was 4 cm. below the right costal margin and tender.

Neurologically she had bilateral foot drop with tenderness of the calves and absent knee and ankle jerks. The planter response was normal.

Investigations done including Hb., platelets, urine microscopy; urine, blood and sputum cultures; urine for porphyrins, blood for S.E.L., blood sugar, lumbar puncture and VDRL were non-contributory. The X-ray chest showed an enlarged heart with pulmonary congestion (Fig. 1). ECG showed sinus tachycardia; total white count was 16,300 with 80% polymorphs; blood urea was 196 mg%; serum electrolytes: potassium 6.4 mEq/L, sodium 120 mEq/L, chloride 80 mEq/L, serum pyruvic acid 6.2 mgm% (Normal 0.5 to 1). Arterial blood gases showed pH 7.07 (7.38-7.42), pCO_2 14 mmHg. (40), Standard Bicarbonate 7.6 (24-28), pO_2 121 mmHg (80). (Patient on oxygen) (normal values within brackets).

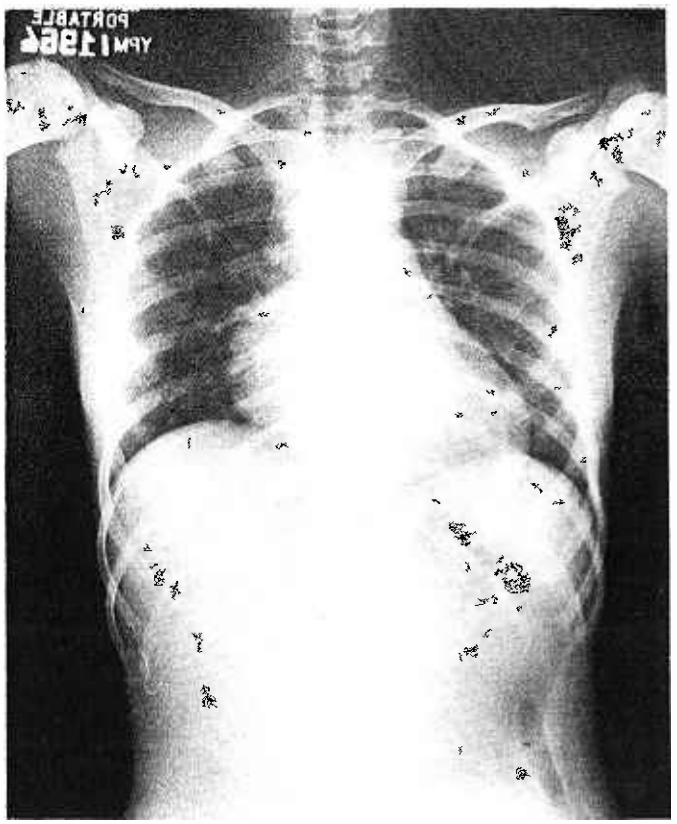

Fig. 1.

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She was diagnosed as having Metabolic Acidosis and Cardiac Beri-Beri with Peripheral Neuropathy.

She was treated with intravenous B1, sodium bicarbonate, oxygen, digoxin, diuretic, hydrocortisone and aramine.

She improved gradually and on the following day was found to be conscious, rational and non dyspnoeic. The pulse was 100/min. with a blood pressure of 100/50. The heart was in dual rhythm and the lungs were clear. Arterial blood gases showed pH 7.37, pO_2 80, pCO_2 37, Standard Bicarbonate 21.7. Her digoxin, diuretic and hydrocortisone was stopped. She was subsequently treated with B1 and a nutritious diet. By 23rd June, 1975, her blood urea had come down to 23 mg%. She was then noticed to be mildly jaundiced with a liver of 2 cm. Liver functions showed SGPT more than 200 I units, alkaline phosphatase 35 King Armstrong Units, serum proteins 5.9 gm/DL, albumin 2.3 gm/DL and S. bilirubin 4.1 mgm%, consistent with the diagnosis of Cholestatic Hepatitis secondary to congestion of the liver and hypotension.

She was up and about on the fourth day and apart from her peripheral neuropathy she continued to make satisfactory progress and was discharged on 8th July, 1975 when her liver function tests had returned to normal.

Case 2

A 40 year old Indian male was admitted on 10th March, 1976 with a history of acute onset of breathlessness and generalised body aches. He had a history of heavy alcohol consumption and had been on the binge prior to admission.

Clinically, on admission, he was restless, delirious and hyperventilating. The pulse was 100/min. with a blood pressure of 100/70 which subsequently fell to 80/40. In the heart, apart from tachycardia there were no abnormal signs or murmurs. There were a few crepitations in the left lung base. The neck veins were engorged and there was a tender, enlarged liver about 3 cm. below the right costal margin. There was minimal oedema of the legs. The knee and ankle jerks were absent. There was no calf tenderness. The fundi were normal.

Investigations done including blood urea, serum electrolytes, full blood counts, urine microscopy, urine for salicylates, urine and blood for methyl alcohol were all normal. The arterial blood gases on admission showed pH 6.9, (7.38-7.42) pCO_2 14 mmHg (40), Standard Bicarbonate 5.5 (24-28), pO_2 122 mmHg (80) with the patient on intranasal oxygen. (normal values within brackets). X-ray chest showed an enlarged heart with pulmonary congestion (Fig. 2). ECG showed sinus tachycardia. The blood sugar was 33 mg% and serum pyruvic acid was 7.8 mg%.

He was diagnosed as having Metabolic Acidosis with Cardiac Beri-Beri and Peripheral Neuropathy and was given intravenous B1, sodium bicarbonate,

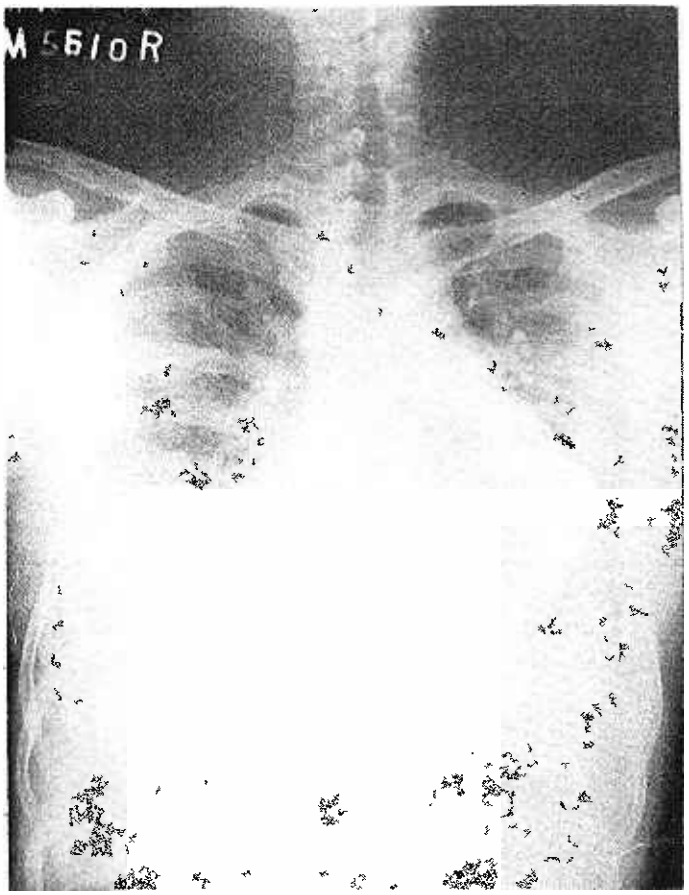

Fig. 2.

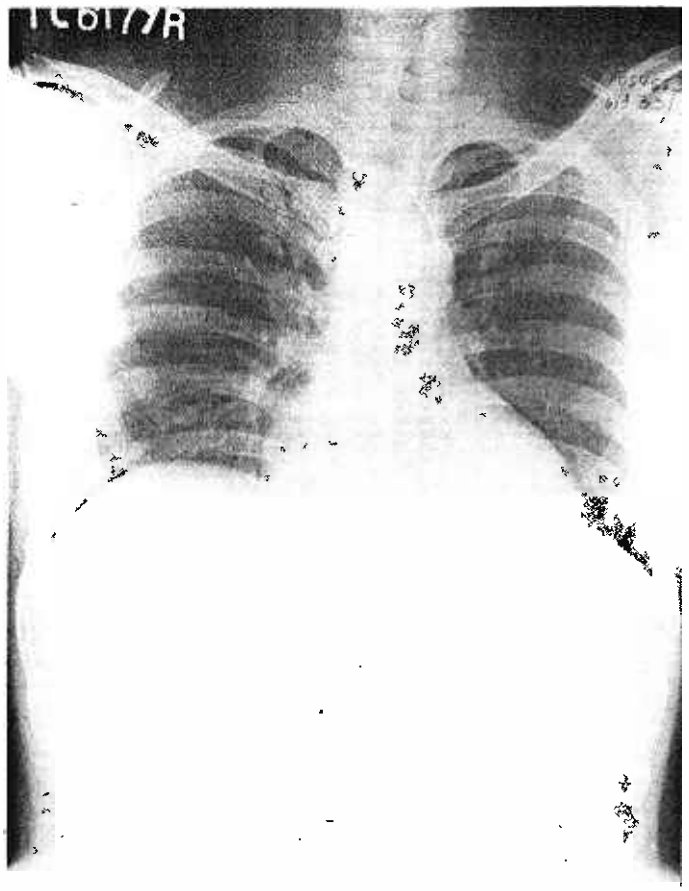

Fig. 3.

glucose, oxygen, digoxin and diuretics.

He made remarkable progress and the following day was found to be comfortable, non dyspnoeic and with a blood pressure of 130/70. Arterial blood gases showed pH 7.49, Standard Bicarbonate 27, pCO_2 34 and pO_2 46 with the patient breathing room air. The blood sugar was 78 mg%. Apart from developing transient withdrawal symptoms in the form of visual hallucinations, the eventual recovery was uneventful and an X-ray chest done on 16th March, 1976 (Fig. 3) was normal. Repeat blood gases done six days later were normal.

DISCUSSION

In thiamine deficiency carbohydrate metabolism is abnormal and pyruvates and alphaketo acids accumulate in the blood. The peripheral vessels dilate, the vascular resistance falls, the cardiac output rises and a state of hyperkinetic circulation is produced (Akbarian *et al*, 1966).

A diagnosis of cardiac beri-beri can be made if the following criteria are met: (1) signs of hyperkinetic biventricular failure (2) evidence of peripheral neuropathy or other vitamin deficiency signs (3) biochemical evidence of low erythrocyte transketolase and raised blood pyruvate levels (4) exclusion of other causes of cardiac failure in a patient with a poor nutritional history plus chronic ethanol consumption and (5) response to thiamine therapy (Blankenhorn M.A., 1945, Wagner P.I., 1965, Sauberlich H.E., 1967, Tanphaichitr *et al*, 1970).

Both the patients described had most of the features listed above except for the high-output state. The estimation of erythrocyte transketolase has been used recently as a possible aid in the diagnosis of thiamine deficiency (Akbarian *et al*, 1966, Sauberlich H. E., 1967). We were unable to measure the transketolase levels. A raised serum pyruvic acid is non-specific but when taken in conjunction with the other findings it helps to substantiate the diagnosis (Wagner P.I., 1965). Several toxic agents inhibit pyruvate oxidation and can lead to the development of high blood pyruvate levels. There was no historical or clinical evidence to implicate any of these agents here. Metabolic acidosis occurs in thiamine deficiency. It is due to raised blood pyruvates and alpha keto acids plus lactic acidemia due to poor tissue perfusion and hypoxia (Sauberlich H.E., 1967). The acidosis seen in these two cases was exceptionally severe, a feature which has not been stressed enough in the past. Metabolic acidosis due to other causes such as diabetic ketosis, uraemia, salicylate poisoning and methyl alcohol intoxication had to be considered. The history, clinical state and biochemical investigations did not substantiate any of these conditions. Uraemia was probably partly responsible for the acidosis in the 1st case.

Other causes of cardiac failure were considered and excluded.

The cause of the shock is unclear. Possibly several factors in combination are responsible in producing it. Thiamine deficiency and acidosis cause myocardial damage and impair cardiac contractility (Fishman, A.P., 1971, Sukumalchantra Y. *et al*, 1976). Acidosis is known to constrict pulmonary vessels in animals and has a similar action in humans when hypoxia is present (Enson Y. *et al*, 1964). A significant fall in the blood flow to the left heart could occur as a consequence with resultant fall in the cardiac output. Akbarian *et al* (1966) postulated that low blood pressure could occur in cardiac beri-beri due to the marked peripheral vasodilatation. Clinically that is usually associated with a hyperkinetic state and as such this is unlikely to be the cause of the very low blood pressures in the two patients described.

The treatment for cardiac beri-beri is to give intra-venous thiamine immediately. The response is quite dramatic but it may take anywhere from 24 to 48 hours before the abnormal metabolism is corrected. Diuretics, digoxin and vasotonic agents are not the drugs of choice per se for the treatment of cardiac beri-beri but they may be used, if necessary, in acutely ill patients such as these two in the initial stages just as they are employed in other hyperkinetic cardiac failures till the basic abnormality is corrected (Blankenhorn M.A., 1945, Akbarian *et al*, 1966).

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