Unexplained lactic acidosis and multiple organ failure resolved completely with thiamine: Shoshin beriberi.

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Introduction

Beriberi is a syndrome caused by thiamine (vitamin B1) deficiency. It presents with non-specific features that may mimic other disease processes and can be classified into dry or wet beriberi. Shoshin beriberi is a fulminant form of wet beriberi that presents with lactic acidosis, circulatory collapse and multiple organ failure.[1] We present a case of multiple organ failure with profound circulatory collapse, renal failure and refractory lactic acidosis that improved markedly following intravenous thiamine replacement, diagnostic of Shoshin beriberi

Case description

A 29-year-old male with a background of haemochromatosis, folate deficient anaemia and peripheral neuropathy presented with acute renal failure. On day five he was admitted to critical care for haemofiltration due to oliguria and fluid overload. His condition deteriorated markedly with refractory hypotension and lactic acidosis. He was commenced on noradrenaline and vasopressin which reached maximal doses, as well as high dose steroids. Antibiotics were started empirically. Imaging was unremarkable and septic screening investigations were negative.

Collateral history from relatives revealed history of alcohol excess (50 units per week) and poor diet consisting mainly of pasta, crisps and tea. They reported an overall decline in health over the previous year due to fatigue.

A diagnosis of beriberi was suspected and intravenous thiamine supplementation was administered. Within several hours haemodynamic parameters improved and lactic acidosis resolved. Vasopressors were weaned off at 36 hours and haemofiltration was discontinued at 72 hours. Renal function improved to normal and he was stepped down to the ward on day 5 and subsequently discharged home on lifelong oral thiamine supplementation.

Serum vitamin panel had been taken prior to thiamine administration, but results were only available after discharge. They revealed global deficiencies with vitamin B1 263ng.g Hb^{-1} (275-675ng.g Hb^{-1}), vitamin A 0.5μmol.l^{-1} (1-3μmol.l^{-1}), vitamin C 2μmol.l^{-1} (15-90μmol.l^{-1}), zinc 9.1 μmol.l^{-1} (11-18μmol.l^{-1}). Iron studies showed iron 3μmol.l^{-1} (12-31μmol.l^{-1}) and transferrin saturations 4.8% (0-50%).

Discussion

Beriberi is a syndrome due to thiamine deficiency. Beriberi typically occurs due to inadequate thiamine intake, such as diets largely consisting of milled white cereals and wheat flour or in chronic alcoholism. Other aetiologies include total parenteral nutrition, malabsorption syndromes, diarrhoea or hyperemesis gravidarum resulting in increased thiamine losses, or disease states with increased utilisation such as pregnancy, hyperthyroidism, refeeding syndrome and critical illness.[2] Thiamine plays an essential role in the Krebs cycle. Deficiency will cause disruption to aerobic metabolism leading to cellular dysfunction and acidosis.[1] Thiamine is found in the diet and body stores are small with deficiency occurring within weeks if inadequate intake.[3] Beriberi involves the nervous system or the cardiovascular system and is typically classified into dry and wet. Rarer forms are Shoshin beriberi and gastrointestinal beriberi. Dry beriberi is characterised by peripheral neuropathy and encephalopathy. Wet beriberi is characterised by heart failure.[1] Shoshin beriberi is a fulminant form of wet beriberi, and has been described as “a rapidly curable haemodynamic disaster.”[4] It is characterised by circulatory collapse and refractory lactic acidosis that can deteriorate rapidly in absence of prompt thiamine replacement, leading to multiple organ failure and death.[1]

Suspected diagnosis should be made based on clinical history and presentation. Risk factors may not be obvious but consider Shoshin beriberi in a patient with unexplained lactic acidosis and cardiovascular compromise. If Shoshin beriberi is suspected, laboratory assays can be sent but these are not always routinely available and most importantly, results should not be awaited for prior to administration of treatment. Furthermore, formal diagnosis is made clinically based on several features: characteristic history or common aetiology of thiamine deficiency, exclusion of other causes of heart failure, additional signs of thiamine deficiency, and therapeutic response to thiamine administration.[5] The administration of thiamine is both diagnostic and therapeutic. Intravenous thiamine will improve haemodynamics within minutes to hours of administration in Shoshin beriberi.[1] It is important to give thiamine before glucose infusion, as without thiamine, pyruvate cannot enter the Krebs cycle and is converted to lactate further exacerbating lactic acidosis.[2]

Supportive measures were commenced rapidly but with little improvement which ultimately may have been fatal. However, within 24 hours of thiamine administration this patient had dramatically improved and eventually made full recovery.

Laboratory results

	Day 0 ICU	Day 1 ICU (6 hours on RRT)	Day 2 ICU (12 hours post thiamine, still on RRT)	Day 3 ICU (36 hours post thiamine, still on RRT)
Urea (mmol.l^{-1})	31.1	26.7	14.7	7.8
Creatinine (umol.l^{-1})	319	312	192	125
Hydrogen ion concentration (nmol.l^{-1})	34	39	37	38
Base excess (mmol.l^{-1})	-4.8	-10.4	-2.4	0.1
Bicarbonate (mmol.l^{-1})	16	13	21	23
Lactate (mmol.l^{-1})	4.1	6.6	1.7	1.5

Conclusion

While Shoshin beriberi remains rare, it has a wide array of aetiologies which are commonly seen. Often diagnosis may be delayed and patients require critical care admission with invasive treatments that can be associated with significant risk, when the correct treatment is relatively simple and inexpensive to administer. Improving awareness of this disease as well as the non-specificity in its presentation will lead to more prompt recognition and treatment of beriberi.

References

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