Shoshin beriberi mimicking central line sepsis in a child with short bowel syndrome

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Background: **Shoshin beriberi, cardiac failure secondary to a severe deficiency of the vitamin thiamine, can develop in patients following extensive intestinal resection or bypass; however, parenteral supplementation has largely eliminated this complication. Hemodynamic instability resulting from central line sepsis is a far more common complication in these parenteral nutrition-dependent patients. This case report details the diagnosis and treatment of shoshin beriberi in a patient with short bowel syndrome whose presentation mimicked central line sepsis.**

Methods: **A retrospective chart review was performed. Appropriate laboratory data were included.**

Results: **The patient was treated unsuccessfully with antibiotics and supportive measures. Resolution of symptoms was achieved only after the empiric administration of thiamine and folate.**

Conclusions: **This case highlights that life-threatening thiamine deficiency mimicking septic shock can develop in patients with short bowel syndrome, despite oral multivitamin administration. We recommend diligent monitoring of vitamin levels in any total parenteral alimentation dependent patient unable to receive the intravenous multivitamin complex, regardless of oral vitamin supplementation or clinical findings.**

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Introduction

Short-bowel syndrome is associated with malabsorption secondary to loss of intestinal length and requires long-term parenteral nutrition[1] and vitamin supplementation. The latter is a routine part of total parenteral nutrition (TPN). A common and significant source of morbidity for patients are catheter-related bloodstream infections, related to either intestinal bacterial translocation secondary to altered intestinal barrier function or a break in sterile technique during local catheter care.[2-4] When these patients present with signs and symptoms of sepsis, a catheter-related bloodstream infection is empirically treated and ruled out. On the other hand, shoshin beriberi, cardiac failure secondary to a severe deficiency of the vitamin thiamine, is rarely seen today in children in developed countries, although it has been reported with TPN-dependent patients during a shortage of the multivitamin infusion.[5,6] We present a case of a pediatric short gut patient who developed signs and symptoms of hyperdynamic shock with metabolic acidosis as a result of thiamine deficiency while on oral multivitamin supplementation.

Case report

The patient is a 28 month-old boy of normal height (91.4 cm, 50th percentile) and weight (12 kg, 20th percentile), with no significant past medical history, until he presented with an acute abdomen at 26 months of age. At his emergent laparotomy, he was found to have a midgut volvulus with necrotic bowel. He required an extensive small intestinal resection and right hemicolectomy, leaving him with 100 cm of the small intestine and no ileocecal valve. Six weeks into his hospitalization, he developed an allergy, which was manifested by hypotension and hives. It was believed to be secondary to the polysorbate vehicle of the multivitamin infusion. Thus, the multivitamins were removed from his TPN. He failed to desensitization attempts, so he was started on an oral multivitamin which did not contain polysorbate. He tolerated the oral vitamin administration, along with a small amount of elemental formula. Prior to discharge home, a

peripherally-inserted central line was replaced with a Broviac central venous catheter. His discharge weight was 13 kg.

Four days following discharge, the patient developed severe lethargy and fever, prompting acetaminophen administration and a return to our emergency room. He was noted to be somnolent, but when aroused, he complained of generalized abdominal pain. On physical examination, he was diaphoretic and listless; his abdomen was mildly distended, with minimal diffuse tenderness, no peritoneal signs and hypoactive bowel sounds. In the emergency room, vital signs showed him to be afebrile; however his blood pressure was 84/39 mmHg with a heart rate of 160 beats per minute. Laboratory evaluations were noteworthy for a white blood cell count of $2.7 \times 10^3/\mu L$, a serum lactate level of 9.6 mmol/L, a bicarbonate level of 11 mmol/L, as well as a C-reactive protein level of 1.7 mg/dL (Table). Random cortisol levels were within normal limits. The patient immediately underwent resuscitative measures and received intravenous broad-spectrum antibiotics (ceftriaxone, vancomycin and metronidazole). An abdominal ultrasound demonstrated some sludge in the gallbladder. There were no gallstones, pericholecystic fluid collections or dilated biliary ducts. He was admitted to the pediatric intensive care unit with a presumptive diagnosis of central line sepsis, acknowledging that the severe lactic acidosis was unusual for this diagnosis. Despite the broad-spectrum antibiotic therapy, the patient continued to deteriorate by developing worsening lactic acidosis (increasing to 10 mmol/L) and requiring intubation. Given that the patient had been on vitamin-free parenteral nutrition supplemented with enteral vitamins for 41 days, an empiric diagnosis was made of severe thiamine and/or folate deficiencies manifesting as shoshin beriberi. Serum folate and thiamine were drawn and sent to the laboratory [The folate level was later found to be low (2.8 ng/ml; normal 3.1-17.5 ng/ml). The thiamine sample was sent to an outside laboratory and lost]. 100 mg of thiamine and 1 mg of folate were empirically infused absence of the polysorbate carrier. Within several hours, the patient's blood pressure normalized, and the serum lactate levels decreased dramatically to 4.1 mmol/L, coupled with a normalized serum bicarbonate level of 25 mmol/L. In less than 24 hours, serum lactate levels were normal, the patient became hemodynamically stable, and was weaned off vasopressor support. He was subsequently weaned from his respirator, extubated and had a full recovery. His blood cultures were negative on final report. He was ultimately discharged home on oral multivitamins with careful follow-up to ensure maintenance of normal vitamin levels. Nineteen months have passed since elapsed. Periodic intravenous thiamine and folate supplementation have been necessary on two occasions during the past six months. On both occasions, the patient's parenteral nutrition was weaned off, and his gut was challenged with full nutritional requirements.

Table. Laboratory data on presentation

Test	Value	Normal range
White blood cell count	$2.7 \times 10^3/\mu L$	$6.0\text{-}17.0 \times 10^3/\mu L$
Lactate	9.6 mmol/L	0.5-1.6 mmol/L
Serum bicarbonate	11 mmol/L	22-29 mmol/L
C-reactive protein	1.7 mg/dL	<0.9 mg/dL
Hemoglobin	10.0 g/dL	11.5-13.5 g/dL
Albumin	3.1 g/dL	3.8-5.4 g/dL
Prealbumin	23 mg/dL	20-40 mg/dL

Discussion

Although the signs and symptoms of beriberi have been found in Japanese documents dating back as early as 808, its direct correlation to thiamine deficiency was not recognized until the early 1900s.[7] In Western culture, the most common causes of adult thiamine deficiency are related to chronic alcohol abuse, prolonged critical illness,[8-11] or previous bariatric surgery.[12] The vast majority of references in children document neuropathy are related to inadequate dietary supplementation.[13,14] The more common neuropathic symptoms such as weakness and pain in the limbs are the hallmarks of dry beriberi. The clinical spectrum of wet beriberi, however, ranges from the classical form to the more severe shoshin beriberi. The classical form of wet beriberi is a slowly developing, tenuous hyperdynamic state with peripheral edema that, if left untreated, may progress to fulminant shoshin beriberi with high output cardiac failure, severe lactic acidosis, peripheral circulatory collapse and shock.[11,15] Shoshin beriberi may also present acutely, as in our patient, without antecedent classic wet beriberi.[16]

Prior to arriving at the correct diagnosis, line removal and a possible exploratory laparotomy awaited our patient; however, his severe acidosis and benign abdominal exam did not fit with either a diagnosis of line sepsis or ischemic bowel. D-lactic acidosis secondary to bacterial overgrowth was also considered in the differential diagnosis, for it had been previously reported in a short gut patient who presented with severe metabolic acidosis and lethargy.[17] The treatment is typically enteral antibiotic therapy; however, intravenous antibiotics would also be indicated in someone with septic shock. Our patient failed to respond to this management.

Wet beriberi has been previously documented

in a short gut patient. Hiroi et al[18] reported a case of thiamine deficiency in a 4-year-old girl who had short bowel secondary to type IV intestinal atresia, leaving her with only 15 cm of the jejunum. She was on a regular diet without parenteral support. Because thiamine is a water soluble vitamin absorbed in the jejunum,[19] she eventually developed heart failure, but it took four years.

Shoshin beriberi related to parenteral nutrition in a 12-year-old girl diagnosed with ulcerative colitis has also been reported.[9] This patient differs from ours in that she was being supplemented with regular but insufficient amounts of intravenous thiamine. She was receiving less than half of the required daily dose (400 μg/1000 glucose Kcal/day). Our patient was administered appropriate oral supplementation (one chewable multivitamin containing 1.5 mg thiamine); however, he did not adequately absorb it, secondary to his shortened bowel length. This was not clinically apparent given his lack of diarrhea, and the fact that no formed vitamins were found in his stool. Therefore, we now recommend diligent monitoring of vitamin levels in any TPN-dependent patient unable to receive the intravenous multivitamin complex, regardless of oral vitamin supplementation or clinical findings.

Patients with short bowel syndrome who are dependent on parenteral nutrition are apt to present with signs of catheter-related sepsis, as this is the most common associated morbidity. Nevertheless, as these patients routinely rely on vitamin supplementation as a component of their nutritional support, thiamine and/or folate deficiency should be suspected if a short gut patient presents with refractory lactic acidosis in conjunction with a hyperdynamic circulation. The diagnosis of beriberi must be considered, and empiric intravenous thiamine and folate should be quickly administered to avoid possible death from cardiac collapse.

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