Acute pernicious beriberi in a patient receiving parenteral nutrition

A case report

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Summary

Parenteral nutrition is being used increasingly in the treatment of the critically ill patient but it causes complications and metabolic derangement. A patient receiving parenteral nutrition in whom protracted vomiting from intestinal obstruction led to the development of acute cardiovascular beriberi (Shoshin) with severe metabolic acidosis — probably lactic — is described. The acidosis was refractory to bicarbonate infusion and inotropic support but the administration of intravenous thiamine 100 mg resulted in a dramatic recovery. Biochemical confirmation of thiamine deficiency was obtained by the measurement of an elevated thiamine pyrophosphate level (24.4%). The patient received thiamine 2.4 mg weekly, a dose that proved insufficient. Thiamine deficiency should be considered when patients receiving parenteral nutrition develop metabolic acidosis with a wide anion gap, even if vitamin supplementation appears adequate.

In the Western world beriberi is almost exclusively a disease of alcoholics. In southern Africa it has been described in hostel dwellers who consume large amounts of beer rich in carbohydrates but poor in thiamine.1 When there is reduced absorption of ingested thiamine or increased demand, as in fever, septicemia and thyrotoxicosis, an acute deficiency state may be precipitated. The development of acute cardiovascular beriberi in a young non-alcoholic woman, who presented with protracted vomiting due to intestinal obstruction and who required total parenteral nutrition (TPN), is reported.

Case report

A 25-year-old woman was admitted to hospital on 17 September 1986 for investigation of colicky abdominal pain and marked weight loss. The pain was epigastric, occurred 1 hour after meals and was relieved by vomiting. Examination revealed a pale, emaciated woman. She was pyrexial (temperature 38°C) and had finger clubbing. Except for the liver, which was 2 cm enlarged beneath the costal margin, the abdomen was normal. Small shotty lymph nodes were present in the tonsillar region. The Mantoux test was positive (15 mm). The haemoglobin value was 7.7 g/dl, white cell count 7.1 x 10^9/l and platelet count 657 x 10^9/l. Blood culture showed no growth. Upper gastro-intestinal tract endoscopy revealed bile contents in the duodenum but the examination was otherwise negative. Abdominal ultrasonography and peritoneoscopy revealed no abnormalities. Tuberculous therapy was administered and the patient’s temperature settled. She asked to go home while considering a liver biopsy. The patient was readmitted 6 days later complaining of vomiting and abdominal distension. Plain radiography of the abdomen revealed a single loop of bowel of normal calibre (colon cut-off sign) in the left upper quadrant. Apart from slight epigastric tenderness, abdominal examination was again unremarkable. A liver biopsy specimen showed minimal fatty change and mild siderosis. On 29 October a barium meal and follow-through examination revealed obstruction in the third part of the duodenum. Two days later she became ill with abdominal distension and was transferred to the alimentation unit in preparation for laparotomy.

Over the next 10 days the patient continued vomiting 1 litre of bile-stained fluid per day. Investigations revealed a severe metabolic alkalosis: serum sodium level 130 mmol/l; potassium 1.6 mmol/l; chloride 77 mmol/l; bicarbonate 40 mmol/l; and urea 3.5 mmol/l. The low serum potassium level required repeated intravenous supplementation and her alimentation regimen included Vitalipid (10 cc), Soluvit (10 cc), and Addamel (Kabi Vitrum (Saphar)) (10 cc) administration twice a week. Intralipid (Kabi Vitrum) 10% (500 cc) was given on a daily basis. All nutrients were mixed in a 3-l bag and administered over a 24-hour period. This provided 1 950 kcal (8.19 MJ) and 14.2 g nitrogen. The thiamine content in Soluvit is 1.2 mg/10 ml vial. On this regimen the patient’s condition appeared to improve and her metabolic alkalosis slowly resolved. During hyperalimentation she received regular blood transfusions (6 U packed cells in all).

Surgery was planned for 2 December but on the evening of 1 December the patient suddenly became ill with hypotension (systolic blood pressure 50 mmHg) and surface cold. The pulse was 100/min and of small amplitude; the urine output dropped to 20 ml/h. The serum sodium level was 119 mmol/l; potassium 2.6 mmol/l; chloride 97 mmol/l; bicarbonate 5.1 mmol/l and urea 7.4 mmol/l (anion gap 20 mmol/l). The Astrup test confirmed severe metabolic acidosis: pH 6.97, carbon dioxide pressure (Paco2) 1,77 kPa, oxygen pressure (Po2) 23.62 kPa, bicarbonate 2.9 mmol/l and base deficit —27 mmol/l.

Intravenous sodium bicarbonate, freeze-dried plasma 2 U, intravenous saline and 50% dextrose (2 x 40 ml) were used in resuscitation. The patient remained hypotensive and oliguric for the next 48 hours. Refractory metabolic acidosis (pH 7.11) and persistent hypoglycaemia (blood glucose 2.7 mmol/l) necessitated further administration of intravenous bicarbonate and dextrose. Puffiness of the face was noted together with signs of heart failure. Precordial T-wave inversion was present on ECG (Fig. 1). Differential diagnosis included cardiogenic shock and an acute adrenal crisis. The serum cortisol level was 1 134 nmol/l (normal 190-690 nmol/l). Digoxin (0.25 mg), intravenous furosemide (80 mg), dopamine infusion (5 μg/
The factors' Hyperalactic contributed to the Johannesburg al. 14. The level of red cell transketolase was 235 U/l (normal 75 - 93 U/l). Following incubation with thiamine pyrophosphate there was a 24,4% (normal < 15%) increase in enzyme activity. One hour later she became more alert and responsive with immediate improvement in her acidotic state (pH 7,26), and 8 hours later the acidosis was fully corrected: pH 7,46; Pco2 4,82 kPa; Po2 7,77 kPa; sodium bicarbonate level 26,9 mmol/l; and base deficit 2,2 mmol/l. The blood pressure rose to 110/90 mmHg and the urine output to 950 ml/min. The central venous pressure remained elevated at 16 cm H2O. During the next 5 days the patient remained hypotensive but polyuric (3 - 5 l urine/d). Dopamine and thiamine were continued until 11 December when the blood pressure stabilised at 110/85 mmHg. Her subsequent course was punctuated by an episode of sepsis which responded to antibiotic therapy. At surgery on 6 January 1987 3 inflammatory strictures of duodenum and proximal jejunum. Histological examination revealed non-caseating granulomas in the lamina propria of the lower jejunum. Although Crohn's disease was not entirely excluded, tuberculosis was thought to be the most likely aetiology. Strictureplasty was performed and postoperatively the patient made a steady recovery on antituberculosis therapy. The presence of hypoglycaemia was an additional complicating feature. Unbalanced administration of dextrose solution to the thiamine-deficient individual may precipitate Wernicke's encephalopathy because the carbohydrate load poses an even greater demand for thiamine. 12,13 Lack of awareness of this hazard prompted several repeated administrations of intravenous dextrose without thiamine with the development of dullness, confusion and a depressed mental state, signs of early Wernicke's encephalopathy. The severe metabolic acidosis was due mainly to lactate accumulation. Thiamine serves as a co-enzyme in the oxidative decarboxylation reaction in the conversion of pyruvate to acetyl co-enzyme A. Deficiency of thiamine leads to accumulation of pyruvate and lactate, which is reflected by severe metabolic acidosis with a large anion gap (> 20 mmol/l) and markedly raised serum lactate levels.

The persistent hypotension and acidosis which was refractory to inotropic support with dopamine and sodium bicarbonate has been well described in acute cardiovascular beriberi. The immediate response to intravenous thiamine characterised by a rise in blood pressure, mental alertness and subsequent diuresis constitute definitive evidence of beriberi. The elevated thiamine pyrophosphate level provided confirmatory evidence of thiamine deficiency. Since the assay is performed on red cells it is likely that repeated transfusions would invalidate the test; this may explain the raised level of transketolase in our patient. The subsequent periods of hypotension in this patient were probably related to excessive diuresis and hypovolaemia that follows therapy with thiamine. In addition there was also a phase of septicaemia, possibly intravascular coagulopathy, which was a contributory factor in the development of hypotension.

Our patient received thiamine 2,4 mg weekly, a dose that proved inadequate. Although Kishi et al.14 have shown that thiamine HCI 5 mg daily is sufficient replacement during TPN, it should be noted that patients on TPN probably have markedly increased thiamine requirements because of associated malnutrition, liver disease, alcoholism, major surgery and sepsis. We therefore endorse Anderson and Charles'15 recommendation that intravenous thiamine replacement therapy (100 mg) be instituted at the start of parenteral feeding. Thereafter daily maintenance dose (2,5 mg/1 000 Kcal) should be based on caloric intake.

Serial monitoring of serum bicarbonate and chloride levels will facilitate early detection of metabolic acidosis. In patients receiving TPN the condition is essentially an iatrogenic one related to the type and amount of caloric load.15 Hyperlacticaemia (as evidenced by metabolic acidosis with a wide anion gap) should prompt a search for thiamine deficiency and treatment should be instituted even if vitamin supplementation appears adequate.

REFERENCES

Discussion
Inadequate vitamin replacement therapy in patients on prolonged intravenous therapy may lead to thiamine deficiency manifesting as Wernicke's encephalopathy. Only recently has acute beriberi with metabolic acidosis (Shoshin) been described in patients receiving parenteral nutrition. This case emphasises the rapidity with which a state of thiamine depletion can develop during prolonged administration of intravenous fluids — even in patients receiving multivitamin supplements if these are inadequate. Studies on volunteers indicate that when thiamine is totally excluded from the diet a state of total body depletion develops within 18 days.11 Our patient became symptomatic 1 month after admission to the hyperalimentation unit. Several factors contributed to the development of her thiamine-deficient state. She was cachectic and probably had depleted thiamine stores before the institution of TPN. Inadequate intake was further compounded by persistent vomiting. In addition, absorption was probably compromised in the setting of upper intestinal obstruction. Finally, the sudden caloric load of TPN resulted in rapid utilisation of residual thiamine precipitating an acute deficiency state.

kg/min) and dexamethasone (0,5 mg twice daily) were added to the therapeutic regimen.

The patient remained acidotic, confused, hypotensive and oliguric until the administration of intravenous thiamine HCI (100 mg). One hour later she became more alert and responsive with immediate improvement in her acidotic state (pH 7,26), and 8 hours later the acidosis was fully corrected: pH 7,46; Pco2 4,82 kPa; Po2 7,77 kPa; sodium bicarbonate level 26,9 mmol/l; and base deficit 2,2 mmol/l. The blood pressure rose to 110/90 mmHg and the urine output to 950 ml/min. The central venous pressure remained elevated at 16 cm H2O. During the next 5 days the patient remained hypotensive but polyuric (3 - 5 l urine/d). Dopamine and thiamine were continued until 11 December when the blood pressure stabilised at 110/85 mmHg. Her subsequent course was punctuated by an episode of sepsis which responded to antibiotic therapy.

At surgery on 6 January 1987 3 inflammatory strictures of duodenum and proximal jejunum. Histological examination revealed non-caseating granulomas in the lamina propria of the lower jejunum. Although Crohn's disease was not entirely excluded, tuberculosis was thought to be the most likely aetiology. Strictureplasty was performed and postoperatively the patient made a steady recovery on antituberculosis therapy.

Thiamine status was determined by the anthrone method. The level of red cell transketolase was 235 U/l (normal 75 - 93 U/l). Following incubation with thiamine pyrophosphate there was a 24,4% (normal < 15%) increase in enzyme activity. One day after therapy with thiamine was started the thiamine pyrophosphate effect fell to 2,6%.

Retroperitoneal lymphangioma

A case report

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Summary

Retroperitoneal cystic lymphangiomas are rare benign tumours. Ultrasonography, computed tomography, lymphography or fine-needle percutaneous aspiration may be used to make the diagnosis pre-operatively. Complete excision of the cyst without rupture is the preferred treatment. However, in complicated cases a conservative surgical approach is mandatory.


Lymphangiomas are a congenital malformation of the lymphatic system, with less than 1% arising in the retroperitoneal space. The first case of retroperitoneal cystic lymphangioma was reported by Gaudier and Gorse in 1913. True retroperitoneal lymphangioma fulfilling the criteria for pathological diagnosis are relatively rare and are usually found incidentally during surgery, autopsy or lymphography. A case of retroperitoneal lymphangioma presenting in adult life that was diagnosed pre-operatively is reported.

Case report

A 58-year-old man was admitted to hospital complaining of progressive abdominal distension and a palpable mass in the left side of the abdomen of several months’ duration. Physical examination revealed a distended abdomen with engorgement of the superficial veins and diminished bowel sounds. A large, moderately tender, non-mobile, soft, smooth mass was noted over the left hypochondrium; this extended into the left lower quadrant. There was no hepatomegaly or lymphoedema of the extremities. The external genitalia were normal. Upon digital examination the mass could not be palpated in the pelvis. The remainder of the physical examination was normal. Routine laboratory values were within normal limits.

Radiography of the kidney/bladder/ureter area showed a huge mass occupying the entire left abdomen, displacing the small intestine to the right. Ultrasonography of the abdomen revealed that the mass was cystic in nature with multiple septa and an irregular margin. Computed tomography (CT) of the abdomen demonstrated a huge retroperitoneal mass in the left abdomen, which was homogeneous and had an attenuation value of 9. The spleen and left kidney were elevated and displaced anteriorly by the mass (Fig. 1). The wall and septa of the mass were slightly and uniformly thickened and enhanced following the administration of contrast medium (Fig. 2). Percutaneous aspiration and drainage of the mass was performed under ultrasonographic guidance. An F8 pigtail tube was inserted into the mass and about 1 500 ml of yellowish milky chylous fluid was drained. An antegrade study, performed by injecting contrast medium into the cyst through the pigtail tube and then taking plain X-ray films, revealed no communication between the cyst and other organs.

On laboratory investigation the aspirates were bacteriologically and cytologically negative. The acid-fast bacillus stain was negative and the biochemical results were: serum amylase level 104 U/l, protein 3 610 mg/dl, glucose 237 mg/dl; fat particles were identified. Bipedal lymphangiography showed interruption of lymphatic vessels over the para-aortic channels of the L3 level with an oblique course to the left retroperitoneal space (Fig. 3).

Upon surgical exploration a huge multilocular cystic mass posterior and medial to the left kidney was found extending down to the pelvis and confined in the retroperitoneal space. As much as possible of the mass was excised and intraperi-