Pediatr Nephrol (2012) 27:2213–2214
DOI 10.1007/s00467-012-2166-0

CLINICAL QUIZ

Renal tubular dysfunction and lactic acidosis: Questions

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Received: 5 March 2012 / Accepted: 13 March 2012 / Published online: 22 May 2012

Keywords Renal tubular acidosis · Lactic acidosis · Acute lymphoblastic leukaemia

Case presentation

A 10-year-old girl presented with a 3-week history of lethargy and weight loss with associated polyuria and polydypsia. There was no significant past medical or family history. On examination she was appropriately grown for age, with no organomegaly or lymphadenopathy, but was noted to be peripherally mottled. Serum electrolyte and venous blood gas levels indicated marked metabolic acidosis (pH 7.29, bicarbonate 12 mmol/L) and hypokalaemia (potassium 1.8 mmol/L). Lactic acid was also elevated (6.4 mmol/L; normal <2 mmol/L). Urinary potassium was 9.9 mmol/L and urinary pH was 6.0 on admission. Calcium, magnesium and phosphate levels were within normal limits, and the results of a full blood examination were normal.

The answers to these questions can be found at doi:10.1007/s00467-012-2180-2.

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She was admitted to the ward and her hypokalaemia corrected using oral supplementation with a mixture of oral potassium chloride and potassium citrate. The serum phosphate decreased to 0.73 mmol/L, requiring supplementation. Glycosuria was noted on dipstick, and there was associated aminoaciduria. The spot calcium:creatinine ratio was elevated at 1.30 mmol/mmol (<0.7 mmol/L). Ultrasound examination identified a right kidney measuring 12.1 cm and a left kidney measuring 13.2 cm, both greater than the 97th centile for age, with preserved corticomedullary differentiation; Doppler waveforms were normal. There was no nephrocalcinosis. Despite appropriate bicarbonate supplementation, the patient remained severely acidotic with bicarbonate levels of between 7 and 13 mmol/L and there was a persistent elevation of lactate of between 5 and 10 mmol/L.

The initial diagnosis was considered to be secondary to a combination of proximal renal tubular acidosis with features of distal tubular dysfunction. An underlying mitochondrial disorder was proposed due to the persistently elevated lactic acidosis. Other causes suggested as differential diagnoses of the tubular acidosis, including heavy metal poisoning, were considered but did not explain the persistent lactic acidosis. Initial haematological findings were normal with no associated organomegaly and a malignant cause was thought to be unlikely. Our patient underwent skin, liver and muscle biopsies for the suspected mitochondrial disorder.

One month post-initial presentation, our patient progressively developed anaemia with a decrease in haemoglobin from 116 to 75 g/L. The total white cell count was slightly depressed at 3.9×10^9/L, but the neutrophil and lymphocyte counts were normal at 1.5×10^9/L and 2.6×10^9/L, respectively. The platelet count was also within normal range. Bone marrow examination revealed that blasts accounted for 38 % of all nucleated cells and that these had the morphology of

lymphoblasts. Flow cytometry of bone marrow confirmed an abnormal population of B lineage cells expressing CD19 and CD38, consistent with precursor B-cell acute lymphoblastic leukaemia. There were no malignant cells in the cerebro-spinal fluid. Our patient was commenced on standard induction chemotherapy with prednisolone, asparaginase and vincristine.

Questions

1. What is the expected progress of the tubular acidosis after treatment with chemotherapy?
2. What features of our patient's initial presentation were suggestive of malignancy?