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BRIEF REPORT

Life-threatening hypokalaemia and quadriparesis in a patient with ureterosigmoidostomy

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ABSTRACT

We report quadriparesis as a result of severe hypokalaemia and acidosis in a 50-year-old man who had undergone ureterosigmoidostomy for bladder extrophy 48 years earlier. Aggressive suppletion with intravenous potassium and bicarbonate combined with potassium-sparing diuretics and ACE inhibitors resulted in complete restoration of the serum potassium and resolution of the neurological symptoms. The underlying mechanism as well as the treatment of hypokalaemia and hyperchloraemic metabolic acidosis after ureterosigmoidostomy are briefly discussed.

INTRODUCTION

Various intestinal segments have been used as conduits to receive urine when the urinary bladder has been removed or is non-functioning. The anastomosis of one or both ureters into the sigmoid (ureterosigmoidostomy) almost always results in hyperchloraemic metabolic acidosis, hypokalaemia and other electrolyte abnormalities (e.g. hypomagnesaemia and hypocalcaemia). Nowadays, this diversion technique is rarely applied because of the high rate of metabolic complications associated with its use. Instead, isolated loops of ileum, jejunum or colon are taken as urine conduits. Since the introduction of the ileal conduit method, the incidence of electrolyte abnormalities has declined considerably.[1]

CASE REPORT

A 50-year-old man was admitted to the hospital for drainage of an abdominal wall abscess that developed 'spontaneously'. He had undergone bilateral ureterosigmoidostomy for bladder extrophy 48 years earlier. He was seen regularly at the outpatient clinic because of mild acidosis and hypokalaemia, for which he received sodium bicarbonate and potassium chloride. His serum potassium was normal (4.0 mmol/l). Following drainage of the abdominal wall abscess, his condition rapidly worsened and he developed generalised muscle weakness. On physical examination, he was alert and afebrile, although he was hyperventilating. The pulse was 72 beats per minute and blood pressure was 130/70 mmHg. There were no pulmonary or cardiac abnormalities. Neurological examination revealed generalised muscle weakness with absence of all deep tendon reflexes. The patient was unable to move his arms or legs or to lift his head from the bed. Fortunately, swallowing and breathing capacity were not disturbed. No abnormal cranial nerve findings or sensory loss were detected. Laboratory investigations revealed severe hypokalaemia (1.9 mmol/l) and severe metabolic acidosis (pH 7.07). The serum bicarbonate was 6 mmol/l, chloride 120 mmol/l, magnesium 0.60 mmol/l, sodium 143 mmol/l, creatinine 324 μmol/l and urea 22.5 mmol/l. The ECG showed hypokalaemic U waves and widening of the QRS complex. Chest X-ray was normal. Ultrasound examination of the upper abdomen showed a dilated collecting system of the right kidney. A diagnosis of hypokalaemic muscle weakness with severe metabolic acidosis causing Kussmaul respiration was made. The patient was treated with intravenous potassium up to 20 mmol/hour combined with bicarbonate, magnesium sulphate and three

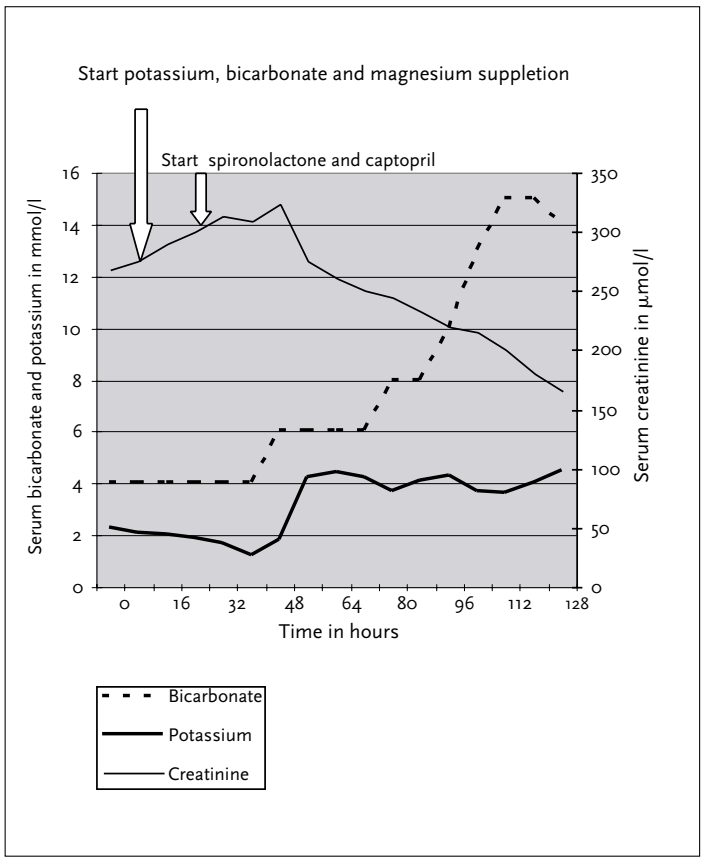

Figure 1

litres 0.9% sodium chloride. Despite this aggressive regime, the serum potassium decreased further to 1.2 mmol/l and the patient developed fever (39 °C). In addition, a combination of potassium-sparing diuretics, ACE inhibitors and antibiotics was administered. This resulted in an increase in the serum potassium to normal values within two days (see *figure 1*). Concomitant with the correction of serum potassium, the neurological symptoms dramatically improved. By then, it became apparent that blood cultures were positive for *Escherichia coli*. Serum bicarbonate normalised approximately four weeks later, as did renal function. Subsequently colonoscopy showed a small polypoid tumour in the right ureteric ostium. The mass was removed by means of snare polypectomy. Microscopical examination revealed adenomatous tissue, but no signs of malignancy.

DISCUSSION

Implantation of the ureters into the gut is commonly used for reconstruction of the urinary tract after extirpative surgery or bladder extrophy. A well-known problem of diversion of urine through intestinal segments is the development of metabolic complications. When the sigmoid is used, severe electrolyte abnormalities such as hyperchloraemic metabolic acidosis and hypokalaemia may occur. Other complications include recurrent infections and osteomalacia.

Chronic *hyperchloraemic metabolic acidosis* secondary to ureterosigmoidostomy was first described by Boyd *et al.*[2] in 1931 in a child who also developed rickets. Since then, numerous investigations have been carried out to elucidate the pathophysiology of this abnormality. Although the mechanism is not completely understood, the acidosis is explained as follows.[3,4] Electrolyte shifts will occur any time urine is in contact with the intestinal mucosa. Urinary ammonia plays a central role in these processes. Ammonium transport is regulated by Na/H antiporters, located at the luminal borders of intestinal cells. The weak acid NH_4 is exchanged for a proton, Na. The exchange of NH_4, however, is coupled with the exchange of serum bicarbonate for luminal chloride. The result is loss of bicarbonate on the one hand and absorption of ammonium chloride in the blood on the other. Furthermore, ammonium transport may also take place through potassium channels. As a result of this metabolic hyperchloraemic acidosis, a net loss of calcium from bones may occur, which can lead to bone demineralisation and kidney stones. Hypocalcaemia may provoke severe complications: tetany in the presence of severe acidosis has been reported in a patient with cloacal extrophy.[5]

Severe *hypokalaemia* is another important metabolic complication. Because urinary concentration of potassium is much higher than the serum concentration, it seems logical to assume that the rectosigmoid would absorb urine potassium, thereby preventing the developement of hypokalaemia. It has been shown, indeed, that the ileum is able to reabsorb potassium. Patients with an ileal conduit do not experience hypokalaemia and have normal potassium body stores.[6] The distal colon, however, is less likely to reabsorb potassium.[7,8] In one third of patients with uretrosigmoidostomy, hypokalaemia and depletion of total body potassium have, in fact, been observed.

The metabolic findings in our patient, however, cannot be fully explained by the passage of urine in the sigmoid. The hypokalaemia was new. Previous laboratory investigations had shown normal serum potassium and only mild acidosis. We suggest that infection and renal obstruction played a major role in this case. Several arguments lend support to this statement. Metabolic acidosis and hypokalaemia in patients with an ureterosigmoidostomy may deteriorate due to renal tubular dysfunction secondary to repeated episodes of pyelonephritis,[9,10] prolonged urine retention in the colon[11,12,13] and hypomagnesaemia.[14] Ascending urinary tract infection is common in patients with uretrosigmoidostomy. Acute pyelonephritis occurs in

10-17% of them and approximately 4% die of sepsis.[15] Hypomagnaesemia, which is often seen in patients with an ureterosigmoidostomy, initiates kaliuresis or causes existing kaliuresis to become worse. Although the pathogenesis is unclear, increased aldosterone activity and impaired distal chloride transport may have played a role, as shown earlier in patients with ileostomy and dehydration.[3] Both aspects, obstruction as well as bacteraemia, were present in our patient. On the one hand there was an *Escherichia coli* septicaemia and hypovolaemia due to an abdominal wall abscess. On the other hand, ultrasound and colonoscopy demonstrated obstruction of the right uretrosigmoidostomy. Taking this into account, it is plausible that infection and tubular dysfunction contributed to the severe hypokalaemia.

As to the therapeutic interventions, some measurements need attention. In stable patients severe deterioration of the electrolyte abnormalities can be avoided by daily administration of oral potassium and bicarbonate, restriction of chloride intake,[16] regular emptying of the colon (nightly insertion of a rectal tube),[17] and timely administration of antibiotics in case of renal infection.
In severely ill patients, as shown here, immediate aggressive suppletion might be life-saving. Bicarbonate and potassium should be administered both orally and intravenously. Correction of the acidosis without potassium replacement might be dangerous, due to progressive paralysis.[3] Hypokalaemia-induced muscle weakness may require mechanical ventilation of a patient.[18] Adequate drainage of the urine is needed in case of ongoing sepsis. In addition, we experienced benefit from treatment with potassium-sparing diuretics and ACE inhibitors. At the time we assumed that the latter treatment reduced the renal excretion of potassium. Afterwards, however, we could not find conclusive literature data on this topic and the exact influence of ACE inhibition remains favourable, but questionable.

Metabolic acidosis occurs far less commonly in patients with an ureteroileostomy, since rapid drainage of urine into the bag means a short contact time, which prevents significant changes in urinary composition.[3, 4, 8] Therefore, we considered constructing an ileal conduit in order to prevent further episodes of hypokalaemia and metabolic acidosis. The patient, however, recovered fully and was unwilling to undergo surgery.

CONCLUSION

We presented a patient with a ureterosigmoidostomy based acidosis and a severe, even life-threatening hypokalaemia developed over a short period of time. Such patients should be promptly treated with a number of (pharmaco) therapeutic measures.

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