CASE REPORT

Primary presentation with acute flaccid quadriparesis in Sjogren's syndrome sans sicca

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SUMMARY

We report the case of a 40-year-old housewife, who presented with vomiting since past 5 days and weakness of all four limbs since 1 day. Clinical examination confirmed the presence of flaccid quadriparesis with preserved tendon reflexes. Routine laboratory parameters showed severe hypokalaemia. On further evaluation she was diagnosed to have type 1 renal tubular acidosis secondary to Sjogren's syndrome. Sicca symptoms were conspicuous by their absence.

BACKGROUND

Renal tubular acidosis (RTA) is an important cause of renal potassium wasting, and should always be suspected when hypokalaemia coexists with metabolic acidosis. The two forms of RTA associated with hypokalaemia, that is, types 1 and 2, differ in the site of tubular disease, and the presence of associated defects in glucose, amino acid and phosphate reabsorption. Type 1 RTA, also known as distal RTA owing to the portion of tubule affected, typically occurs as an isolated defect. It can be distinguished from type 2 RTA by challenge infusion with sodium bicarbonate. Other features suggestive of type 1 RTA include relatively more severe acidosis with a disproportionately higher urinary pH, and a higher urinary sodium concentration. Type 1 RTA is also frequently associated with nephrocalcinosis and symptomatic nephrolithiasis, leading the patient to seek medical help. Hypokalaemia in type 1 RTA is often mild and asymptomatic but can occasionally be severe. Once type 1 RTA has been diagnosed, evaluation for an underlying disorder is mandatory. Sjogren's syndrome is an important aetiology in adults, once drug-induced RTA has been excluded.

As this case report amply demonstrates, secondary type 1 RTA can develop without pathognomic sicca symptoms of Sjogren's syndrome. Maintaining a high suspicion for possible autoimmune disease is therefore essential in adults diagnosed with type 1 RTA.

CASE PRESENTATION

A 40-year-old woman presented with sudden onset of weakness of all four limbs since 1 day. She also reported recurrent vomiting in the 5 days preceding development of weakness. There were no associated sensory disturbances and no symptoms suggestive of cranial nerve dysfunction. Bowel and bladder function was preserved. She did not report any neck injury or any similar episodes of weakness in the past. Her medical history was uneventful. She denied any history of substance abuse.

Retrospective questioning failed to elicit any symptoms of ocular or oral dryness.

At admission, the patient was haemodynamically stable. General physical examination was unremarkable. Neurological examination confirmed the presence of flaccid quadriparesis. Motor power was 2/5 in all four limbs. Plantars were bilaterally flexor and deep tendon reflexes were preserved. Sensory examination was normal.

INVESTIGATIONS

Routine laboratory tests showed severe hypokalaemia (serum potassium 1.5 mEq/l). Other serum electrolytes were within normal limits (serum sodium 130 mEq/l, serum calcium 9.3 mEq/l). Blood glucose, renal (serum creatine 1.1 mg/dl) and blood counts were all normal. Liver function tests showed hyperglobulinaemia (5.9 g/dl). Arterial blood gas analysis revealed severe metabolic acidosis (pH 7.17, pCO₂ 22, HCO₃ 7.7 mEq/l). Serum cortisol and thyroid function tests were normal (cortisol 27.39, thyroid stimulating hormone 1.45 μ IU/ml). Serum chloride was elevated (113 mEq/l), yielding a normal anion gap (9.3 mEq/l). HIV serology was negative.

ECG showed diffusely flattened T waves with prominent U waves (figure 1). Urinalysis showed proteinuria; 24 h urine protein was 351 mg. Urinary pH measured prior to correction of acidosis was 8.0. Transtubular potassium gradient was significantly elevated (41.07), confirming renal potassium wasting. Urinary anion gap was positive (7.8 mEq/l), consistent with RTA. Further evaluation of RTA with sodium bicarbonate infusion was not attempted considering the risk of exacerbating hypokalaemia. However, elevated urinary pH in the presence of severe metabolic acidosis, and elevated urinary sodium (83 mEq/l) were strongly suggestive of type 1 RTA.

Considering this diagnosis, the patient was screened for autoimmune disorders and tested strongly positive for antibodies to Ro/SSA antigen.

Schirmer's test was positive in both eyes. Abdominal ultrasonography ruled out nephrocalcinosis and nephrolithiasis. Renal biopsy was performed and showed healthy glomeruli and focal tubular atrophy with patchy interstitial lymphoplasmacytic infiltrate and fibrosis, consistent with chronic tubulointerstitial nephritis (figure 2). Immunofluorescent staining was negative.

DIFFERENTIAL DIAGNOSIS

- Hypokalaemic quadriparesis owing to renal tubular acidosis, secondary to Sjogren's syndrome without sicca symptoms.

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Figure 1 ECG (12 lead) showing diffusely flattened T waves with prominent U waves (black arrow heads).

TREATMENT

The patient was treated with parenteral and oral potassium supplementation (80 mEq through either route daily). Acidosis was gradually corrected with parenteral sodium bicarbonate (50 mEq intravenous every 8 h). She was also initiated on corticosteroid therapy with prednisolone (50 mg orally once a day) and a maintenance regimen of oral sodium bicarbonate (1300 mg orally every 8 h; equivalent to 48 mEq of bicarbonate) and potassium citrate 15 ml orally every 8 h; equivalent to 60 mEq of potassium).

OUTCOME AND FOLLOW-UP

Motor power recovered rapidly with potassium correction, and serial blood gas analyses showed improvement in metabolic acidosis. The patient was subsequently discharged. She remains asymptomatic on follow-up 6 months after the index illness, with no recurrences of quadriparesis.

DISCUSSION

Type 1 RTA is a well-known complication of Sjogren's syndrome, occurring in up to 11.3% of all cases,¹ although the incidence of primary presentation with RTA is much lower. A large retrospective study by Chen *et al*,¹ for instance, found six cases of primary presentation out of 250 patients with Sjogren's syndrome over a period of 11 years. Moreover, severe hypokalaemia with muscle weakness was noted in only three of those patients.

Figure 2 Renal histopathology showing healthy glomeruli and focal tubular atrophy with patchy interstitial lymphoplasmacytic infiltrate and fibrosis, consistent with chronic tubulointerstitial nephritis (H&E staining, magnification $\times 20$).

Vaidya *et al*² described a young woman with hypokalaemic periodic paralysis subsequently diagnosed to have RTA with hypokalaemia. Unfortunately, the diagnosis of underlying Sjogren's syndrome was only made when the patient developed sicca symptoms. Primary presentation with RTA was diagnosed retrospectively. Singhvi *et al* outlined a case similar to ours, wherein a woman presenting with flaccid quadriparesis and incipient respiratory paralysis, was diagnosed with type 1 RTA owing to Sjogren's syndrome.³ However, there was no clear reason in that instance for sudden manifestation of hypokalaemic symptoms. In comparison, our patient developed symptomatic hypokalaemia after protracted vomiting, which likely unmasked the underlying hypokalaemia by contributing to it. Another report by Khandelwal *et al*,⁴ describes two young women with quadriparesis, later diagnosed with Sjogren's syndrome and type 1 RTA. Interestingly, quadriparesis responded well to potassium correction alone and acidosis in both cases was detected much later. The authors did not offer any explanation for this phenomenon. In our instance, supplementation of both potassium and bicarbonate produced a rapid recovery from symptoms.

Palkar *et al* reported another case of RTA with severe hypokalaemia owing to underlying Sjogren's syndrome, unmasked by fever and dehydration.⁵ In that instance, the patient developed bulbar paralysis and cardiac dysrhythmia in addition to quadriparesis. Hyperglobulinaemia which was seen in this patient and our patient- has independently been linked with RTA; non-specific auto-immunity being the ostensible mechanism. The severe manifestation of hypokalaemia in the patient described by Palkar *et al* when compared with our patient is notable. The possibility of preexisting cardiovascular and/or cerebrovascular disease is reasonable, considering her older age and postmenopausal status. The close clinical similarities of this disorder to acute inflammatory demyelinating polyneuropathy were highlighted in a report by Agrawal *et al*.⁶ The patient in question rapidly developed respiratory failure, requiring invasive mechanical ventilation before a diagnosis of RTA with hypokalaemia owing to Sjogren's syndrome was made.

In contrast with the afore-mentioned cases where Sjogren's syndrome was confirmed by salivary gland biopsy, we opted to perform a kidney biopsy. It showed features of chronic interstitial nephritis, a pattern observed in up to 71% of cases of primary Sjogren's syndrome.⁷ Indeed, this histopathological pattern has been recommended as an additional diagnostic criterion for Sjogren's syndrome, and we believe that it vindicates our diagnosis. The renal-predominant presentation of our

patient also made it more reasonable to biopsy the kidney instead of the salivary glands.

Learning points

- ▶ Hypokalaemia of renal tubular acidosis (RTA) can be sufficiently severe to produce flaccid quadriplegia.
- ▶ Presence of metabolic acidosis in hypokalaemic patients is an important clue to the possibility of RTA.
- ▶ Sjogren's syndrome can present with RTA in the absence of sicca symptoms.
- ▶ Demonstration of chronic tubulointerstitial nephritis in renal biopsy supports a diagnosis of underlying Sjogren's syndrome in patients with RTA.

Contributors SD and KN have performed data acquisition and prepared the manuscript. GV edited the manuscript and performed review of literature. VA and MH provided clinical care to the patient and edited the manuscript and review of literature.

Competing interests None.

Patient consent Obtained.

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