CASE REPORT

Hypokalemic rhabdomyolysis: an unusual presentation of Sjogren's syndrome

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SUMMARY

Hypokalaemic rhabdomyolysis represents a medical emergency requiring rapid diagnosis and appropriate aetiological treatment. Renal tubular acidosis is a common cause of hypokalemia which can be idiopathic or secondary to systemic disorders such as Sjogren's syndrome. It can remain asymptomatic or manifest with metabolic abnormalities including hypokalemia paralysis, hypocalcaemia and hyperchloremic metabolic acidosis. Rhabdomyolysis presenting with severe hypokalemia as the first manifestation of Sjogren's syndrome is rare. We report a case of a 59-year-old woman who presented to our department with severe weakness of all limbs. Laboratory examination demonstrated hypokalemic rhabdomyolysis caused by distal renal tubular acidosis. Investigations revealed Sjogren's syndrome as the underlying cause of the metabolic disorders.

BACKGROUND

Sjogren's syndrome is a multifaceted autoimmune disease which may extend from an exocrinopathy to a systemic disorder including the involvement of visceral organs. Thus, there are several clinical scenarios in which the diagnosis of Sjogren's syndrome should be suspected.¹ Renal impairment is one of the extraglandular manifestations of the disease. It is usually subclinical and develops during the course of the disease over. However, in several cases, significant clinical renal involvement caused by tubulointerstitial nephropathy was described as the first symptoms of Sjogren's syndrome.^{2–3} Nephrogenic diabetes insipidus, hypokalemic paralysis, renal calculi and osteomalacia are the main complications of distal renal tubular acidosis (dRTA).⁴ In this case, we describe an unusual presentation of Sjogren's syndrome revealed by hypokalemia-induced rhabdomyolysis.

CASE PRESENTATION

A 59-year-old woman, without any medical history, presented with four limb muscle pain and generalised weakness since 10 days. She denied any acute infections, trauma, drug intake or intoxication. She did not have digestive symptoms. The family history was negative for renal and muscle disease. On the day of admission, she noted an inability to walk unassisted and profound fatigue. Physical examination revealed tenderness in the limb muscles and generalised weakness that was marked most proximally. Blood pressure was 110/60 mm Hg and pulse rate was 86/min. The cardiovascular, respiratory and abdominal examinations

were normal. The remainder of the physical examination was normal.

INVESTIGATIONS

Laboratory examination revealed low serum potassium (2 mmol/L), elevated creatinine phosphokinase (1200 IU/L) and mild renal insufficiency (creatinine clearance 40 mL/min). The serum myoglobin level was 750 UI/L. Serum sodium was 148 mmol/L and chloride was 124 mmol/L. The ECG showed sinus rhythm. No U-waves were seen. Additional tests showed that pH was 7.28, PCO₂ 27, PO₂ 110 mm Hg and bicarbonate 14 mEq/L. The serum calcium level was 2.14 mmol/L. Alkaline urine was observed (pH 6.5). No glucose or proteins were found in the urine. The thyroid function tests were normal. The patient was diagnosed as suffering from hypokalaemic rhabdomyolysis complicating distal renal tubular acidosis. As the patient had symptoms of xerostomia, xerophthalmia, high value of erythrocyte sedimentation (130 mm) with hypergammaglobulinemia at 22 g/L, Sjogren's syndrome was suspected. Schirmer's test was abnormal. Antinuclear antibodies were present in a titre of 1/6400. Anti-SSA and anti-SSB were positive. A labial salivary gland biopsy showed focal aggregates of lymphocytes corresponding to Chisholm stage 4 lesions. The diagnosis of hypokalaemic rhabdomyolysis caused by renal involvement of Sjogren's syndrome was retained.

TREATMENT

The patient was treated with potassium supplements and alkalinisation.

OUTCOME AND FOLLOW-UP

Her symptoms resolved over a 2-week period with normalisation of laboratory test abnormalities.

DISCUSSION

This case illustrates an unusual presentation of Sjogren's syndrome. The patient developed hypokalemic myopathy as a result of renal involvement. Renal tubular acidosis is considered to be due to lymphocytic tubulointerstitial infiltrates. It occurs in almost 25–40% of patients with Sjogren's syndrome.^{5–6} Hypokalemia is the most common electrolyte abnormality, occurring in 28–53% of patient with dRTA.⁷ It is associated with hyperchloremic metabolic acidosis and alkaline urine. The clinical spectrum of renal tubular acidosis may be silent to life-threatening.⁴ In Sjogren's syndrome, severe symptomatic hypokalemia with periodic paralysis and respiratory arrest has been reported in only a few cases.^{8–9} Potassium

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deficiency can induce muscle ischaemia and, in case of severe depletion, muscle necrosis and rhabdomyolysis. The main clinical manifestations of hypokalemic rhabdomyolysis are muscle pain, cramps, fatigability and generalised weakness.¹⁰ These symptoms may precede the sicca symptoms and reveal a previously undiagnosed disease process.⁸ Evidence of salivary and lachrymal gland involvement, characteristic lymphocytic infiltration on salivary gland biopsy and the presence of Ro/SSA or La/SSB autoantibodies establish the diagnosis of Sjögren's syndrome.¹¹ Treatment consists of potassium replacement and alkali therapy with bicarbonate. Systemic steroids can be used for severe dRTA.¹²

Learning points

- Management of patients with severe hypokalemia includes a vigorous search for the underlying aetiology.
- Physicians should bear in mind Sjögren's syndrome as a differential diagnosis among patients with hypokalemia-induced rhabdomyolysis.
- Renal tubular acidosis due to renal involvement must be overlooked even in the absence of eyes and mouth dryness.

Competing interests None.

Patient consent Obtained.

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