Pediatric Rheumatology

Poster presentation

Hypokalemic paralysis revealing Sjögren's syndrome (case report)

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Case report

A 16-year old girl presented with rapid onset of progressing muscular weakness, dysphagia, dysphonia and significant wasting. She lost 13 kg during the last year. On examination she was dystrophic (BMI 15,7) and had clinical signs of severe myopathy. Laboratory findings confirmed myopathy (CPK 106,4 ukat/L, ALT 0,96 ukat/l, AST 2,86 ukat/L, myoglobin 1582 ug/L), inflammatory markers were elevated (ESR 60/92, CRP 37 mg/L). There was marked hypokalemia (S-K 1,8 mmol/l) suggesting hypokalemic paralysis. Diagnosis of distal renal tubular acidosis (dRTA) was based on confirmation of hyperchloremic metabolic acidosis (S-Cl 120 mmol/l, pH 7,31, BE-10) with normal serum anion gap, severe hypokalemia, high urinary pH (pH 7,5) and positive urinary anion gap. Other signs of renal tubular impairment were obvious (high urinary beta-2-microglobulin 213 mg/l, glomerulotubular proteinuria 1,01 g/24 h). Positive autoimmune tests (high positivity of rheumatoid factor IgG, IgA, IgM, positive ANA/IF, ENA SS-A/Ro, SS-B/La, elevation of circulating immunocomplexes and IgG) and mildly reduced values of sialometric measurements revealed primary Sjögren's syndrome (SS) as the underlying cause of dRTA. The renal biopsy confirmed chronic tubulo-interstitial nephritis compatible with this diagnosis. Full recovery of muscle weakness and laboratory findings of hypokalemia and acidosis followed potassium and alkali replacement. Corticosteroids were administered with subsequent addition of cyclosporine A because of disease activity. The girl is in longterm remission.

Conclusion

We report the patient with severe hypokalemia and subsequent hypokalemic paralysis. The cause of hypokalemia was dRTA as the manifestation of renal impairment in primary Sjögren's syndrome.