



Hypokalemic paralysis in systemic sclerosis - A clinical challenge

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General Note

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ABSTRACT

A forty year old female presented with weakness of all the four limbs since two days. On evaluation it was found that she was hypokalemic which she had for last four months. She was further investigated for persistent hypokalemia. On clinical examination she was found to have sclerodactyly which prompted to consider the diagnosis of systemic sclerosis. The diagnosis was further

supported by presence of antinuclear antibody and anti Scl 70 antibody. Urinary loss of potassium due to distal renal tubular acidosis was causing hypokalemia. The patient was managed with potassium supplementation.

Keywords: distal renal tubular acidosis, quadriplegia, urinary loss of potassium, scleroderma

1. INTRODUCTION

Systemic sclerosis also known as scleroderma is immune mediated rheumatic disorder characterized by fibrosis of internal organs and skin and characterized by vasculopathy. Despite patients of diffuse cutaneous systemic sclerosis having evidence of augmented survival systemic sclerosis still has the highest mortality rates when compared with other rheumatic diseases (Nihtyanova et al., 2010; Tyndall et al., 2010). Systemic sclerosis is designated as an orphan disease having highly unmet medical needs (Denton et al., 2015; Khanna et al., 2015; Chung et al., 2012). It is uncommon and associated with uncertain outcomes causing great reduction in quality of life with development of manifestations that may prove to be potentially lethal. It is a complex disease with estimation of affected cases being 1 in 10,000 people (Bossini-Castillo et al., 2015).

Distal Renal Tubular Acidosis (dRTA) is characteristic of impaired normal urinary acidification in presence of normal GFR (Glomerular Filtration Rate) with distal term implying disturbed acidification by connecting tubules and collecting duct which form the distal part of nephron (Both et al., 2014). dRTA is known to cause hypokalemia occurring because of failure in H/K ATPase pump (Mustaqeem & Arif, 2019). dRTA has a known association with autoimmune disorders like Sjogren's syndrome, Systemic lupus erythematosus, Multiple sclerosis, Rheumatoid Arthritis, Primary biliary cirrhosis and hepatitis (Weiner, 2018).

2. CASE REPORT

A 40 year female presented to us with complaints of vomiting for one month and sudden onset quadriplegia from 2 days. Patient was experiencing no difficulty in breathing or swallowing. She was diagnosed with scrub typhus 4 months back and her reports revealed hypokalemia during the treatment in another hospital, which has persisted since the episode and patient was on continuous treatment for the same in another hospital, though she was known to suffer from hypokalemia patient did not have any episode of limb weakness in the past (figure 1).

Management

Intravenous potassium chloride replacement and intravenous fluids given followed by oral potassium replacement.

Potassium levels started improving {3.0meq/dl (day2)-4.3meq/dl (day 4)}

Patient improved symptomatically, vomiting subsided and quadriplegia resolved.

3. DISCUSSION

Patients of systemic sclerosis are usually classified into 2 major subsets, first subset being diffuse cutaneous systemic sclerosis which includes patients with proximal limb or trunk involvement with systemic sclerosis, high frequency of severe lung fibrosis, short history of Raynaud's phenomenon, increased risk of renal crisis and cardiac involvement. The second major subset is limited cutaneous systemic sclerosis in which patients have distal skin sclerosis, pulmonary arterial hypertension and severe gut diseases with a long history of Raynaud's phenomenon. Another syndrome associated with systemic sclerosis as reported in literature is overlap syndrome where systemic sclerosis may be present along with clinical and investigational feature of another autoimmune rheumatoid disorder (Denton & Khanna, 2017). Most patients of this syndrome are of subset of limited cutaneous systemic sclerosis (Focharoen et al., 2016; Penn et al., 2007).

Distal renal tubular acidosis (dRTA) clinically presents with a triad of hyperchloremic metabolic acidosis, hypokalemia and inability to reduce urinary pH to less than 5.5 (Agarwal et al., 1993). Under the influence of aldosterone, distal tubule normally generate new bicarbonates however because of damaged alpha intercalated cells of distal tube, there is no new generation of bicarbonates in turn leading to no hydrogen ions thereby raising pH of urine. There is failure of H/K ATPase pump which is associated with hypokalemia (Mustaqeem & Arif, 2019). This impairment in H/K ATPase proton pump leads to defect in acidification, hyperchloremic acidosis and loss of urinary potassium (Misra et al., 2003). Among patients of chronic metabolic acidosis, reduced bone mineral density is an expected outcome (Somnuek et al., 2001). Suppression of bone formation and resorption with low bone mineral density is observed in patients of distal renal tubular acidosis.

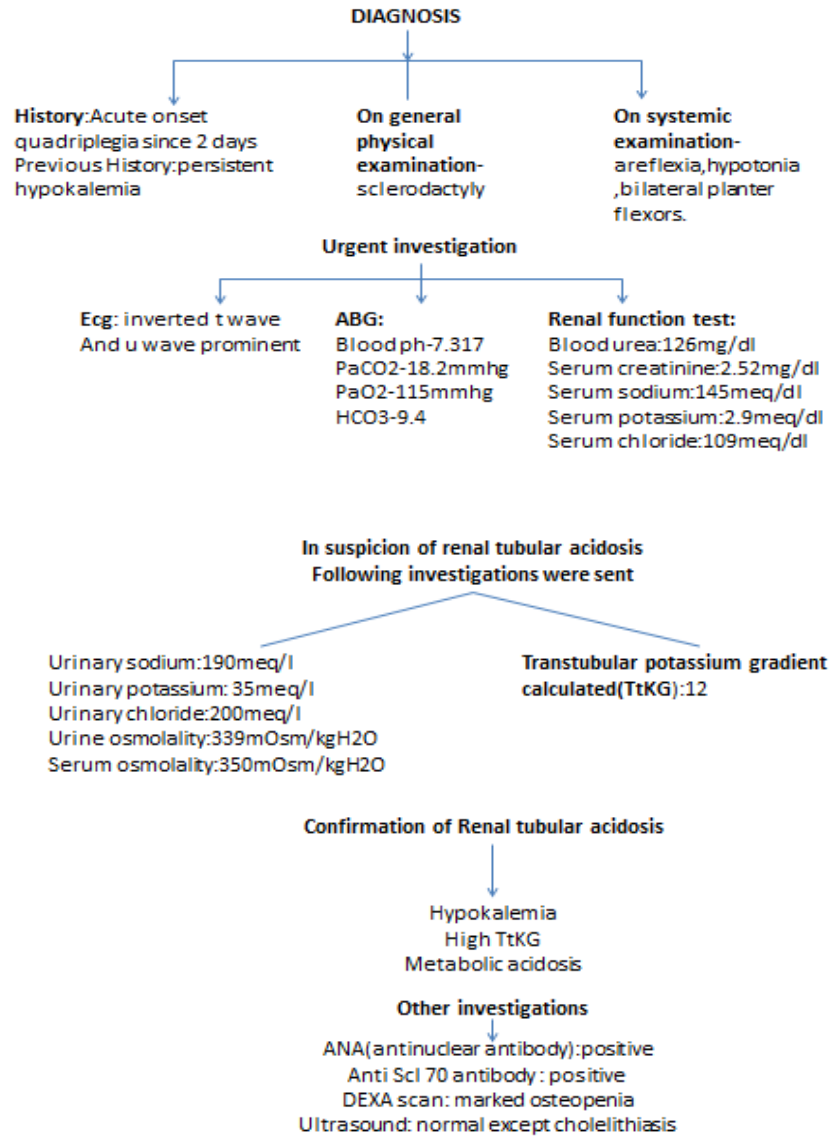


Figure 1 sclerodactyly

4. CONCLUSION

Our patient had a history of hypokalemia for almost four months before developing quadriplegia, but she was undiagnosed, underlining the fact that as these cases are very uncommon in general practice and us physicians are bound to miss out on these less common autoimmune disorders highlighting that sharing our experience of these cases becomes much more important so that these clinically challenging diseases can be detected early and treated, improving the patients quality of life.

Patient Consent

Written consent was obtained from the patient to publish as case report

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Conflicts of Interest: The authors declare no conflict of interest.

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