Abstract

Sjogren's syndrome is a chronic autoimmune inflammatory disorder with predilection for exocrine glandular and extra-glandular involvement . There are two forms of the disease depending on the presence of underlying connective tissue disease , primary and secondary . The latter being more common is associated with underlying connective tissue diseases . Extra-glandular renal involvement can present in the form of distal renal tubular acidosis giving rise to severe hypokalaemia.

A 32-year-old Sri Lankan female presented with bilateral symmetrical proximal muscle weakness. She was detected to have severe hypokalaemia with normal anion gap metabolic acidosis. A diagnosis of distal renal tubular acidosis due to primary sjogren's syndrome was made as the cause of hypokalaemia, based on background history of exertional dyspnoea with symptomatic anaemia, polyarthralgia and multiple dental caries and several specific investigations. There was a marked improvement of her weakness with potassium replacement, prednisolone and hydroxychloroquine.

Severe hypokalaemia is an emergency that must be excluded in a patient with myopathy. Identification of mechanism of hypokalaemia would direct to the

underlying cause. Hypokalemia due to distal renal tubular acidosis is a complication of primary sjogren 's syndrome which needs long term potassium replacement and immunosuppression with oral steroids, hydroxychloroquine orazathioprine.