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Hypokalemic Periodic Paralysis-A Rare Complication of a Rare Case

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Introduction: Hypokalemic Periodic Paralysis a relatively common disease of reversible weakness can be secondary to systemic cause. Here we describe multitude presentations of a systemic vasculitis (Sjogren' s syndrome) presenting as quadriparesis with respiratory failure. Primary Sjögren's syndrome (pSS) is an autoimmune disease characterized by a progressive lymphocytic infiltration of the exocrine glands with varying degrees of systemic involvement.

Case Report: Following a sumptuous wedding meal our 45 year old female presented with symmetric weakness of all four limbs with pain of three days duration with difficulty in breathing. On examination she was tachypnoeic, with normal cranial nerves, flaccid are flexic quadriplegia, sensory being normal and plantar flexor. Her investigations revealed electrolytes of 132/2.8/106 mEq/L, RFT-44/1.5mg/dl. First ABG showed metabolic acidosis with non compensated respiratory acidosis with pH of 6.6 and HCO3 16. ECG showed sagging of the ST segment, depression of the T wave and elevation of the U wave suggestive of Hypokalemia. After 2 hrs of correction, serum potassium was still 2.9, Thyroid profile normal. Anti TPO antibody normal. Repeat blood gases showed pH-7.11, pCO2-34.5, p02-100 and HCO3-12. After 24hrs of potassium correction patient showed improvement in weakness. However patient couldn't be weaned off due persistent acidosis. In view of persistent acidosis with hypokalemia possibility of "Renal Tubular Acidosis" was considered with urine pH-7.0(4.6- 8.0), urine potassium-20.9mEq/L (17-145), pH being >5.5-suggested type 1 RTA.

Subsequently she made a recovery both in terms of respiration and weaknesss. Past history interrogation revealed petichiae over trunk and extremities, since 3 years with painful parotidomegaly and dry itchy eyes. Her USG parotid and FNAC, revealed hypoechoic lesions throughout parenchyma and mononuclear inflammatory infiltrates respectively.

Her connective tissue workup showed, RA factor was positive, ESR being high at 90mm/hr and ANA was positivity with ssA, ssB, Ro52 being strongly positive. Lip and skin biopsy histopathological photographs further supported the diagnosis of "Sjogren's syndrome".

Discussion: Prevalence of pSS is 0.5-2%. Of pSS, having renal involvement is mostly for interstitial nephritis followed by RTA (5-6.6%). Reported incidence of hypokalemic paralysis in patients with type 1 renal tubular acidosis (RTA) is about 28-33%. Literature shows 18 cases of similar presentation between 1966 and 2004. Subsequently, approximately 13 more cases has been reported.

Conclusion: Hypokalemic paralysis is a well known, rare complication of severe distal RTA from any cause. Cases of pSS manifesting as hypokalemic paralysis caused by distal RTA have been rarely reported. A high index of suspicion for pSS should be kept in all patients with hypokalemic paralysis.