Sjogren’s Syndrome Presenting with Quadriparesis Due to Proximal Renal Tubular Acidosis

Ankit Grover¹, Sanjay Kumar², Smita Gupta³*, Pranav Mehta⁴, Vineet Bansal⁵

¹Senior Resident, Department of General Medicine, Shri Ram Murti Smarak Institute of Medical Sciences, Bareilly, Uttar Pradesh, India
²Associate Professor and Nephrologist, Department of General Medicine, Shri Ram Murti Smarak Institute of Medical Sciences, Bareilly, Uttar Pradesh, India
³Professor And Head, Department of General Medicine, Shri Ram Murti Smarak Institute of Medical Sciences, Bareilly, Uttar Pradesh, India
⁴Senior Resident, Department of General Medicine, Shri Ram Murti Smarak Institute of Medical Sciences, Bareilly, Uttar Pradesh, India
⁵Junior Resident, Department of General Medicine, Shri Ram Murti Smarak Institute of Medical Sciences, Bareilly, Uttar Pradesh, India

*Corresponding Author
Smita Gupta

Abstract: Renal Tubular acidosis (RTA) is a tubular acidification disorder characterised by severe electrolyte disturbances mainly hypokalemia and normal anion gap acidosis out of proportion to GFR. Sjogrens syndrome though a rare disorder usually causes distal RTA however few studies have reported cases of Proximal RTA as well. Here we report a case of Acute onset quadriplegia in a young female which was attributed to Hypokalemia. Further evaluation revealed that RTA probably was proximal in origin due to Sjogrens Syndrome.

Keywords: Sjogrens syndrome, Renal Tubular acidosis (RTA), tissue disorder.

INTRODUCTION

Renal Tubular acidosis (RTA) is a tubular acidification disorder characterised by severe electrolyte disturbances mainly hypokalemia and normal anion gap acidosis out of proportion to GFR. On clinical and pathophysiologic grounds, RTA has been separated into three main categories: proximal RTA or type 2; distal RTA or type 1; and hyperkalemic RTA or type 4. Each may occur in a varied number of hereditary or acquired etiologies [1].

Sjogrens syndrome is a autoimmune connective tissue disorder causing xerostomia, xerophthalmia with systemic manifestations as polyarthritis, renal tubular acidosis, Interstitial nephritis, interstitial lung disease and salivary gland lymphocytic infiltration [2]. Sjogrens syndrome though a rare disorder usually causes distal RTA however few studies have reported cases of Proximal RTA as well. Here we report a case of Acute onset quadriplegia in a young female which was attributed to Hypokalemia. Further evaluation revealed that RTA probably was proximal in origin due to Sjogrens Syndrome.

CASE REPORT

A 28 year old female patient from a remote village of Rampur presented to our Emergency room, with history of acute onset quadriparesis without any respiratory, bulbar or cranial nerve involvement. She had also suffered 3-4 similar episodes in the past which recovered with potassium chloride supplementation without any residual deficits. However no evaluation was done before. On admission patient was conscious, oriented, power 0/5 in all 4 limbs without any respiratory involvement, mute plantars, absent deep tendon reflexes and hypotonia. There was No history of any drug abuse or trauma. Detailed history suggested history of similar episodes in the past, dryness of mouth and eyes, difficulty...
Renal Tubular acidosis occurs either due to H+ secretory defect or a HCO3- reabsorptive defect. It may occur in patients with a normal renal function or in patients of chronic kidney disease [3, 4].

Three types of RTA are commonly encountered in clinical practice. Types 1 RTA is caused due to H+ secretory defect in distal tubule whereas Type 2 RTA is caused by defects in proximal tubule in HCO3- absorption, and type 4 RTA is characterized by abnormal excretion of acid and K+ in the collecting duct, leading to hyperkalemic acidosis. Type 3 RTA is rare and has features of both distal and proximal RTA [5].

In distal (type 1) RTA the nephrons lack the ability to secrete H ions and hence acidify the urine normally during spontaneous or induced metabolic acidosis [2]. Inherited forms include autosomal-dominant, autosomal-recessive, or X-linked of which mutations in the basolateral chloride-bicarbonate exchanger has been identified as the most common form of inheritance. Acquired causes include hypergammaglobulinemic states, Sjogren syndrome, SLE, chronic active hepatitis, thyroiditis, Graves’ disease, tubulointerstitial diseases include leprosy, chronic pyelonephritis, obstructive uropathy; and genetic diseases like Ehler Danlos syndrome, hereditary eliptocytosis, sickle cell disease [6]. In distal RTA, a tendency for nephrocalcinosis due to hypercalciuria, and hypocitraturia [6]. Severely depressed plasma bicarbonate levels with a corresponding inappropriate urinary pH >5.5 differentiates from type 2 RTA [7].

Proximal type 2 RTA is characterized defective reabsorption of bicarbonate in the proximal tubule usually without defects in the transport of other solutes. This leads to increased delivery of bicarbonate to distal nephron which has less reabsorbative capacity for HCO3- leading to urinary loss of bicarbonate [8, 9]. This leads to consequent systemic acidosis. However the urine pH remains acidic inspite of filtered bicarbonate after reaching a steady chronic state wherein the bicarbonate levels falls below the reabsorptive threshold and the filtered bicarbonate gets reabsorbed at this point. Persistent acidosis causes a fluid depleted state leading to activation of the Renin- Angiotensin aldosterone system causing increased aldosterone secretion and consequent loss of potassium in the urine.

A diagnosis of Proximal RTA should be suspected in patients with persistent hypokalemia with acidic urine and a normal anion gap metabolic acidosis. A common association with Fanconi’s syndrome in patients presenting with
hypocalcemia, hypophosphatemia, hypouricemia and euglycemic glycosuria should be suspected. A definitive diagnosis of Proximal RTA lies in demonstrating a high filtered bicarbonate load in the urine by increased fractional excretion of bicarbonate.

The etiology of proximal RTA varies from inherited congenital disorders, paraproteinemias, autoimmune connective tissue disorders and drugs related. Proximal RTA in association with Fanconi syndrome can occur following exposure to some medications, including tenofovir sodium valproate and topiramate. Topiramate is a carbonic anhydrase inhibitor that can cause simultaneous defects in both proximal and distal acidification mechanisms, presenting as type 3 RTA [10]. The most common cause of Proximal RTA is usually Multiple Myeloma presenting with anemia, hypercalcemia, hyper gammaglobulinemia, raised ESR, renal dysfunction and bone lesions. In all adults of Proximal RTA a primary diagnosis of multiple myeloma should be kept in mind unless an alternative diagnosis is found out.

A unique presentation of our case was its manifestation as acute onset quadripareis with severe hypokalemia which on evaluation was attributed to renal tubular acidosis. On further workup we documented the etiology of renal tubular acidosis as Sjogrens Syndrome. Sjogrens syndrome is usually associated with Distal RTA however very few case reports have demonstrated its association with Proximal RTA. Although common cause of proximal RTA includes paraproteinemias likely multiple myeloma, but the absence of hypercalcemia, renal failure, bone pain, normal globulin levels, normal xray skull, age and gender excluded the possibility of multiple myeloma in our patient.

A definitive diagnosis of Sjogrens syndrome and Interstitial nephritis in our patient required minor salivary gland biopsy and renal biopsy however the same was refused by the patient. A preliminary diagnosis of Sjogrens syndrome was thus made in view of autoimmune panel and the electrolyte abnormalities. Patient responded to Oral Potassium citrate solution, oral bicarbonate replacement along with oral prednisolone and methotrexate. With over 6 months of follow up, patient is presently doing well and maintained on prednisolone 5mg alternate day along with methotrexate and potassium citrate supplementation.

Conflicts of Interest: None

REFERENCES
