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Case Report

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

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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 quadriparesis 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 quadriparesis 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

in deglutition and polyuria. There was no significant family history. Other systemic examination was normal. Urgent ABG done at emergency suggested a severe hypokalemia with a normal anion gap hyperchloremic metabolic acidosis. ECG done at emergency suggested T inversion in V1-V6. A diagnostic evaluation workup for hypokalemia was sent and patient was started on IV and oral potassium replacement followed by bicarbonate replacement.

Meanwhile blood investigations revealed Hemoglobin of 8.3gm/dl, platelet count of 180000/mm^3, Total Leucocyte Count 5600/mm^3. Kidney function test and electrolytes revealed a serum potassium of 2.7 meq/L sodium 147 meq/L calcium 5.8 meq/L magnesium 2 meq/L phosphorous 1.4 meq/L creatinine 0.5 mg% and Urea 24 mg%. Liver function test was normal. Thyroid function test was normal. Urine examination did not reveal albuminura or any active urinary sediment but showed glucosuria 2+. 24 hour urine sample showed a excretion of potassium of 96meq/L and Urinary protein (24hours) was 1.1grams indicating amino-acid/proteinuria. Viral markers including HIV were non reactive. Spot urinary potassium/creatinine ratio was 1466 meq/g of creatinine indicating a renal loss of potassium. Ultrasound abdomen did not reveal any significant abnormality.

On the basis of above investigations and keeping a possibility of Renal tubular acidosis , a further workup for etiology of RTA and the type of RTA was performed. Persistent hypokalemia with hypocalcemia, hypophosphatemia, normal anion gap metabolic acidosis and glucosuria, proteinura with absence of renal calculi suggested a strong possibiltiy of Proximal RTA in our patient. Detailed history was suggestive of features of Sjogrens syndrome. However to further classify the type of RTA, fractional excretion of phosphorous was calculated which came as 42.86% indicating renal loss of phosphorous. Meanwhile patient was evaluated on the lines of Sjogrens syndrome and a positive Schirmers test along with ANA positivity- 1:320 titre with speckled pattern and anti-Ro, Anti- La positivity was documented which favoured the diagnosis of Sjogrens syndrome.

Patient was advised salivary gland biopsy and renal biopsy, however consent for the same was not provided by the patient.

Patient improved with potassium supplementation and was then started on potassium citrate along with phosphorus, calcium and bicarbonate replacement. Patient was started on Oral prednisolone 1mg/kg of body weight and was tapered after 6 weeks to a maintenance dose of 5mg alternate day. On follow up patient is doing better, with a normal potassium, calcium, bicarbonate and phosphorous levels maintained on Oral prednisolone 5mg alternate day along with potassium citrate and Vit D and calcium supplementation.

DISCUSSION

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 reabsorptive 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 bicarrbonate 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 demonstarting 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 acidifification 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 quadriparesis 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 possibilty 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 methotraxate and potassium citrate supplementation.

Conflicts of Interest: None

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