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Case Report

A CASE OF PRIMARY SJOGREN`S SYNDROME WITH DISTAL RENAL TUBULAR ACIDOSIS PRESENTING AS HYPOKALEMIC PARALYSIS

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ABSTRACT

Renal tubular acidosis secondary to autoimmune interstitial nephritis is quite common in patients with Sjogren`s syndrome. Here we present a case of 24 year old female who presented with Acute Hypokalemic Quadriparesis and was later diagnosed with distal RTA. Patient did not have features of xerostomia or xerophthalmia but was diagnosed to have Primary Sjogren`s syndrome fromserological findings,in this case renal involvement preceded subjective Sicca syndrome.Patient recovered after giving i.v potassium. Patient was treated with corticosteroids and is asymptomatic during the one year follow up period.

KEYWORDS

Hypokalemic paralysis, Renal tubular acidosis, Primary Sjogren`s syndrome.

INTRODUCTION

Acute Hypokalemic quadriparesis is a common neurological problem though most of them are due to gastrointestinal loss. Distal RTA causing Hypokalemic paralysis is a well-known but a rarely encountered complication of RTA. Further work up should always be done to rule out autoimmune disorders such as Sjogrens`s syndrome with primary renal involvement causing distal RTA. We report this case to emphasise the wide array of exocrine gland involvement in Sjogren`s syndrome.

CASE REPORT

A 25 year old woman came with history of sudden onset weakness of all four limbs. There is no history of gastrointestinal loss, recent vaccination, loss of consciousness, trauma to back, ingestion of diuretics or any toxins .She is a diagnosed case of primary hypothyroidism on the thyroxine replacement since 4 years. She is not a known case of diabetes or hypertension. She had similar episodes of weakness in the past diagnosed as hypokalemic paralysis. She also has recurrent episodes of pancreatitis and underwent Cholecystectomy with ERCP stenting in the first episode. There was no history of similar limb weakness in the family.

On examination: Vitals were normal. Neurological examination: Higher mental functions were normal. Cranial nerve examination was normal. Motor system examination revealed power of 2/5 in all four limbs with hypotonia. Superficial and deep reflexes were absent. Bilateral plantar reflex was equivocal .Sensory system was normal.

Preliminary blood investigations revealed hypokalemia with hyperchloremia and normal anion gap metabolic acidosis. Serum amylase and serum lipase were raised. Urine routine showed alkaline urine with increased specific gravity of urine with positive urinary anion gap. The findings were highly suggestive of distal RTA.

USG abdomen and pelvis: Revealed features of chronic pancreatitis. Also, small renal calculi and nephrocalcinosis.

CT scan: Features suggestive of chronic pancreatitis. Bilateral renal pyramidal calcifications (Figure 1).

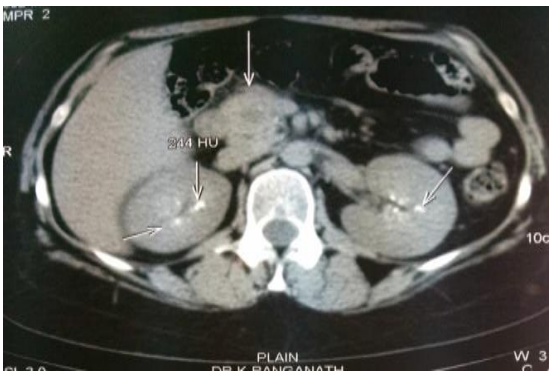

Figure 1. CT scan: Bilateral renal pyramidal calcifications

Patient was treated with iv potassium infusion and patient regained power of 5/5 in all four limbs within 6 hours. Patient was also given oral sodium bicarbonate tablet for the treatment of acidosis. Potassium citrate was started to

maintain the potassium> 3.5meq/l. However the patient had GI symptoms with potassium citrate and was replaced with oral potassium chloride. Patient was discharged with oral kcl solution and sodium bicarbonate tablets.

Investigations were done to find out the cause of distal RTA. ANA profile was done which showed anti SSA/ro and Anti SSB/la to be strongly positive (+++) suggestive of Sjogren`s Syndrome. In our case there was no subjective history of xerophthalmia or xerostomia during the course of illness and renal involvement had preceded the onset of clinical/ subjective SICCA syndrome. Schirmer`s test was negative. Serum electrophoresis showed Hypergammaglobulinemia of polyclonal origin (Figure 2).

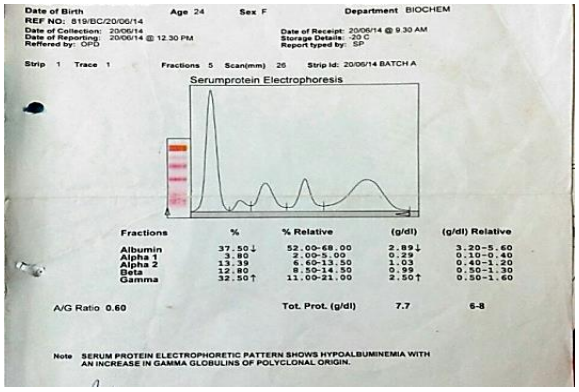

Figure 2. Serum electrophoresis: Hypergammaglobulinemia of polyclonal origin

So the final diagnosis of Primary Sjogren's syndrome with Hypokalemic Periodic Paralysis distal RTA with Autoimmune pancreatitis was made, Patient was discharged on Oral Prednisolone and KCl syrup 15 ml tid.

Patient was followed for 1 year she is Asymptomatic till date her Potassium level and Arterial blood gas levels are normal.Currently her Prednisolone tapered to 5 mg per day.

INVESTIGATIONS

Hb: 11.7gm/dl, TC: 7800/mm^3, ESR: 20mm/hr, PLATELET: 1.6lakh, Na: 140mEq/L, K: 2.2mEq/L, Mg: 2.0 mEq/L, Ca: 9.2mg/dl, LFT: WNL, RFT: WNL, ECG: FLATTENING OF T WAVES.

Thyroid Profile:

FT4: 1.03, TSH: 2.65.

SERUM AMYLASE: 35 IU/ml, SERUM LIPASE: 1000 IU/ml

SERUM ALDOSTERONE: NORMAL

ABG

PH: 7.25 URINE ROUTINE

PO2; 104 PH: 8.0

PCO2:21

URINE SPECIFIC GRAVITY: 1.002 (1.020-1.028)

HCO3; 12.0 Urine anion gap +7 (positive)

Cl^-: 118, Na^+: 140, K^+: 2.2, Serum Anion Gap: 10 (8-14)

DISCUSSION

Renal tubular acidosis is the main cause for normal anion gap acidosis. Hypokalemia though common in both is more severe and symptomatic in type 1 distal RTA.

Treatment of distal RTA involves oral intake of sodium bicarbonate along with potassium supplementation as potassium citrate which is given at the dose of 0.5-2.0meq/kg body weight in order to keep serum K+ levels normal and serumHCO3> 18meq/L.[1]

Renal involvement in Primary Sjögren's Syndrome may be seen in as much as 27% of cases[2] In a large case studies,[2] reduction in urinary concentrating capacity was the most common defect and was observed in 20% of cases. Prevalence of other renal defects included reduction in creatinine clearance (13%), frank distal RTA (5%), hypokalemia (7%), subnephrotic proteinuria (17%) and nephritic syndrome (3%).[2]

Sjögren's syndrome is chronic autoimmune disorder affecting many organs predominantly exocrine glands.[3] Sjögren's when occurs in isolation is known as primary Sjögren's Syndorme (pSS) as reported here. Predominantly seen in middle aged women (M:F=1:9). In addition to dry eyes and dry mouth pSS can also cause renal(renal tubular acidosis, nephritis), thyroid (hypothyrodisim or thyroditis), gastrointestinal (atrophic gastritis, pancreatitis), pulmonary and liver disease.

The pathophysiology is still unclear but renal biopsies have demonstrated tubulointerstitial nephritis with focal or diffuse interstitial lymphocytic infiltrates.[4] This inflammatory process disrupts the cellular architecture causing a secretory defect in the distal tubules.[5] It is thought that hypergammaglobulinemia might cause distal renal tubular dysfunction and the tubular pathology in SS is secondary to this dysfunction.[6] Our patient did have a significant increase in gammaglobulins; (Figure 2).

While the response of tubulointerstitial nephritis to steroid treatment is debatable, many studies have noted a remission of RTA by treating the primary disease with long term low dose glucocorticoid therapy.

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