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A Curious Case of Quadriplegia

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Abstract:-

➢ ***Background:***

Hypokalemic periodic paralysis (HPP) is an uncommon disorder of the muscle marked by intermittent weakness of muscles , which, in severe cases, can progress to respiratory failure. It is frequently associated with renal tubular acidosis, a condition that can manifest in connection with systemic disorders like Sjögren's syndrome and thyroid disorders.

➢ ***Case Presentation:***

A 58-years-old woman presented to the hospital with weakness in all extremities. The patient exhibited motor strength of 0/5 in all limbs. The ECG revealed sinus bradycardia and hypokalemia changes. Laboratory tests showed hypokalemia, metabolic acidosis with a high anion gap, urine pH of more than 5.5, FT4 levels at 6.9 ng/dL, TSH levels at 25.03 IU/mL, Anti-TPO antibodies positive, and a positive SS-A (Ro), Ro-52, SS-B (La) in the ANA profile, indicative of Sjogren's syndrome. Patient has previously experienced similar issues and also has a confirmed diagnosis of hypothyroidism.

➢ ***Discussion:***

The patient received diagnoses of hypokalemic periodic paralysis(HPP), Sjögren's syndrome, and renal tubular acidosis due to findings of hypokalemia, metabolic acidosis, elevated urine pH, and a positive ANA profile.

➢ ***Conclusion:***

Distal renal tubular acidosis secondary to Sjögren's syndrome can rarely present as HPP.

I. INTRODUCTION

Hypokalemic periodic paralysis (HPP) is the most prevalent form of periodic paralysis, characterized by intermittent episodes of weak and flaccid muscles due to abnormal sarcolemmal excitability, often associated with channelopathies.(1). It's a neuromuscular disorder marked by intermittent skeletal muscle weakness, which can progress to respiratory muscle failure and potentially fatal outcomes. The prevalence is 1 in 100,000 cases (1).

Muscle weakness is affected by changes in serum potassium levels, that can result from primary or secondary causes. Primary causes usually show an autosomal dominant inheritance pattern, while secondary causes include factors like diuretic use, gastrointestinal losses, renal tubular acidosis (RTA), primary hyperaldosteronism, Barter's syndrome, hyperthyroidism, hypothyroidism, and certain autoimmune disorders.[2]

In this article, we outline a case involving hypokalemic periodic paralysis in a patient who initially exhibited generalized weakness and was later diagnosed with renal tubular acidosis (RTA) secondary to Sjogren's syndrome.

II. CASE

A 50-year-old female presented with abrupt onset weakness in all four limbs and trunk, unable to move any of these areas. The weakness was sudden, symmetrical, equal in all limbs, non-progressive, and accompanied by complete loss of power. There was no history of muscle wasting, altered consciousness, involuntary movements, visual issues, respiratory or bulbar weakness, fasciculations, flexor spasms, sensory problems, or spinal trauma.

The patient also reported not passing urine since that morning and lacked the sensation of bladder fullness or the urge to urinate. Additionally, she experienced two episodes of vomiting the day before.

Three years prior, the patient had a similar admission for which treatment details were not accessible. She had a known history of hypothyroidism and was currently prescribed thyroxine supplements.

➢ *Vitals on Admission*

- HR- 56 bpm
- BP- 112/72 mmhg
- Spo2- 99 % at RA

➢ *Day 1-*

Upon admission, the patient had bradycardia. A catheterization procedure yielded around 600 ml of urine output. General physical examination revealed presence of pallor and a dry tongue. Other signs such as icterus, cyanosis, clubbing, edema, lymphadenopathy were not noted.

The CNS examination revealed normal higher mental functions, generalized hypotonia, bilateral power of 0/5, DTR of 2+, mute plantar responses, intact sensations including touch, touch, temperature, vibration, and joint position sense. The patient's laboratory values indicated severe hypokalemia, necessitating potassium correction. ABG analysis showed metabolic acidosis with compensatory respiratory alkalosis and a high anion gap.

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➢ *Day 2*

The patient's serum potassium levels were monitored repeatedly, and potassium correction was continued due to persistently low levels. Her serum TSH levels were 14.6 mcIU/ml, prompting an increase in the dose of oral Thyroxine.

Urine examination revealed a pH of more than 5.5, normal urine osm, and urine K^+ levels exceeding 20 meq/l.

➢ *Day 3*

The patient underwent a thorough clinical examination to explore any systemic causes for her condition. Hyperpigmentation, taut and stretched skin over the hands and feet, dehydration, and the absence of features indicating polyarthritis, Raynaud's syndrome, or dysphagia were noted. The potential for an endocrine or collagen vascular disorder was taken into account., prompting the evaluation of serum cortisol levels and ANA profile due to the presence of hypothyroidism and skin changes.

Fig 1: Skin Changes Over Hands

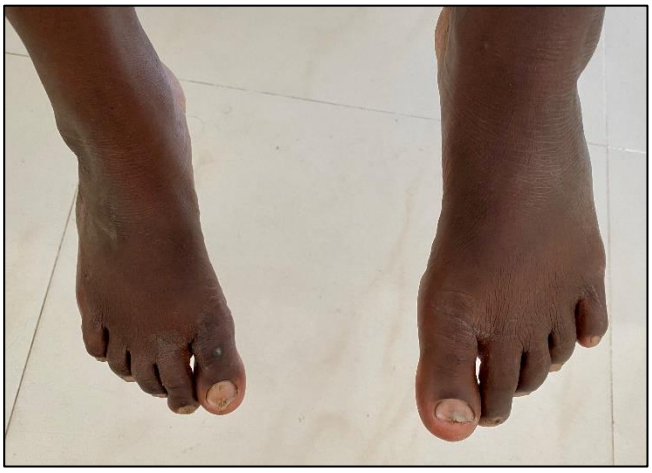

Fig 2: Skin Changes Over Feet

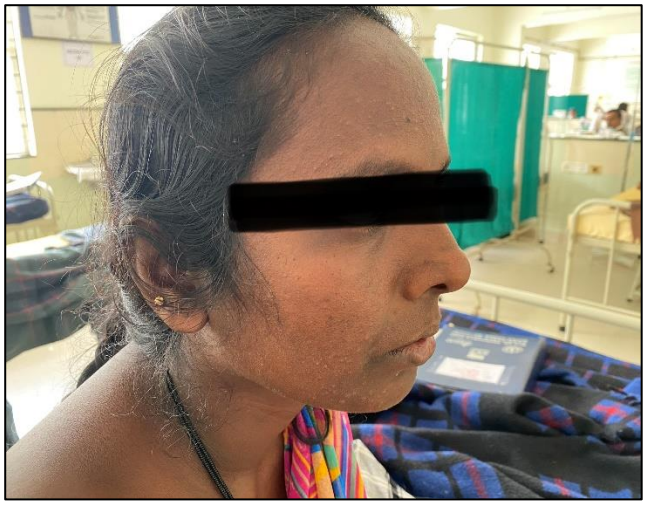

Fig 3: Pigmentation and Skin Changes Over Face

Table 1: Blood and Urine Investigations

Date	Day 1	Day 3	Day 5	Day 7	Day 8	Day 9
Hb	11.2g/dl					
Pcv	33.5%					
Rbc	4.26mil/mm3					
Platelets	413,000cells/mm3					
WBC count	9600 cells/mm3					
Neutrophils/lymphocytes	85 15					
Peripheral smear	Microcytic hypochromic anemia with neutrophilia					
Urea	32mg/dl					
Creatinine	0.98mg/dl		0.77mg/dl			
Total bilirubin	0.7mg/dl					
Sgot	30 IU/L					
Sgpt	25 IU/L					
ALP	58 IU/l					
Total proteins	7.6g/dl					
Albumin	3.6g/dl					
Sodium	137meq/l		135meq/l			
Potassium	1.43meq/l	3.57meq/l	3.58meq/l	3.26meq/l	3.79meq/l	3.74meq/l
Chloride	113meq/l					
Bicarbonate	14.4meq/l		19meq/l			
Uric acid	4.2mg/dl					

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ESR	10mm 1st hr					
Urine ph	>5.5					
Urine examination	Albumin-1+					
Urine osm	330mosm/kg					
Urine potassium	>21meq/l					
Serum osmolality	289mosm/kg					
Total Calcium	8.21mg/dl					
Magnesium	2.76mg/dl					
Free t3	0.5ng/dl					
Free t4	6.7ng/dl					
TSH	14.6mIU/L					
Ana profile	SSA + SSB + RO 52 +					

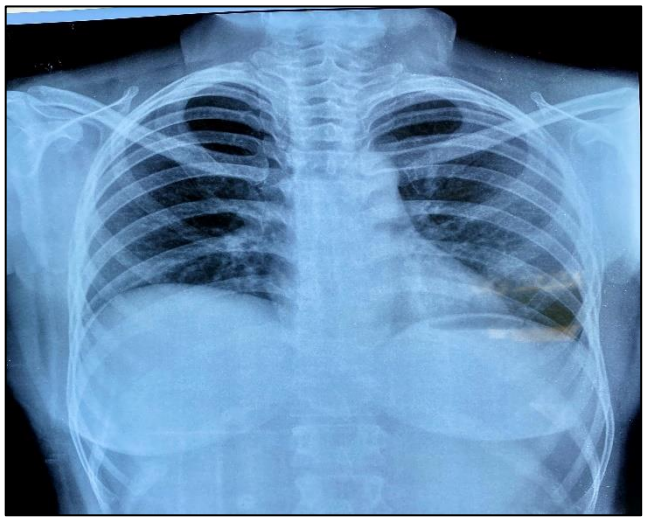

Fig 4: Chest X-ray PA View

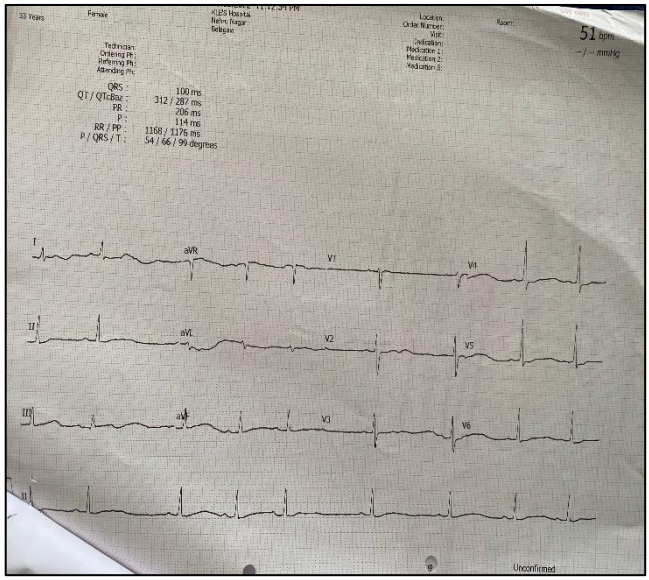

Fig 5: ECG

The patient also underwent a chest x-ray, which was normal, and an ECG showed sinus bradycardia with subtle u waves.

➢ *Day 4*

The patient's ANA profile showed a strong positive result for anti-SSA (Ro), Ro 52, and SS-B (La). Dry eyes were detected during the Schirmer's test. Despite symptomatic improvement, the patient continued to experience persistent acidosis, leading to a diagnosis of Renal tubular acidosis secondary to Sjögren's syndrome.

III. DISCUSSION

This female patient presented with HPP due to distal RTA secondary to Sjögren's syndrome as the first and most prominent manifestation.

Hypokalemic periodic paralysis (HPP) is a heterogeneous condition characterized by episodes of skeletal muscle paralysis associated with periodic reductions in serum potassium levels. Hypokalemia can occur due to either a deficiency of potassium or abnormal shifts of potassium within cells.[3]

Hypokalemic periodic paralysis (HPP) can be triggered by the rare condition known as distal renal tubular acidosis (DRTA). In DRTA, there is a reduced ability to excrete acid in the urine, distinguishing it from proximal renal tubular acidosis, where there is impaired bicarbonate reabsorption. Among the types of renal tubular acidosis, distal renal tubular acidosis is the most common.

DRTA can be caused by primary or secondary factors, with autoimmune conditions like Sjogren's syndrome, systemic lupus erythematosus, and rheumatoid arthritis being common secondary acquired causes. Additionally, renal tubular acidosis can uncommonly be associated with thyroid disorders such as hypothyroidism.

Sjögren's syndrome is a systemic autoimmune disorder characterized by a unique set of signs and symptoms primarily caused by cell-mediated autoimmunity targeting exocrine glands [4]

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In the case study, the patient had dry mouth and tested positive for Anti-SSA (Ro), Anti-SSB (La), and RO-52 autoantibodies, supporting the diagnosis of Sjogren's syndrome. Several examinations were conducted to rule out SLE and rheumatoid arthritis

In patients with Sjogren's syndrome, the prevalence of renal tubular acidosis is estimated to be 4.5–9%, typically appearing in middle age, with only two-thirds experiencing symptoms. Renal involvement is an extra-glandular manifestation in less than 10% of Sjogren's syndrome cases. Although distal renal tubular acidosis (dRTA) is common in Sjogren's syndrome, it is often asymptomatic, and in most cases, it goes undetected. Hypokalemia is the primary electrolyte abnormality in dRTA patients, attributed to factors such as secondary hyperaldosteronism, defective H+-K+ ATPase, and bicarbonaturia. [5]

The pathogenesis of dRTA in Sjögren's syndrome involves interstitial nephritis with high levels of anti-carbonic anhydrase antibodies. These antibodies affect the function of carbonic anhydrase in cortical collecting ducts, leading to an acidification defect. This defect results from a lack of intact H+-ATPase pumps in intercalated cells. [6]

IV. CONCLUSION

The biochemical indications of renal potassium loss associated with hyperchloremic metabolic acidosis supported the diagnosis of distal renal tubular acidosis in the patient, despite the absence of a more comprehensive evaluation. A subsequent detailed clinical examination revealed skin changes and a dry mouth, prompting further investigation for the potential presence of Sjogren's syndrome.

An important consideration is the rare occurrence of hypokalemic periodic paralysis (HPP) secondary to hypothyroidism, a condition reported in only a few cases globally. This possibility is also being explored in the present case, given the abnormal thyroid profile.

This case report highlights the significance of considering distal renal tubular acidosis (DRTA) in the differential diagnosis of patients with hypokalemic periodic paralysis who present with hyperchloremic metabolic acidosis, hypokalemia, and a positive urinary anion gap, even though DRTA seldom leads to HPP.

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