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Case of 50 Years Old Female with Quadriplegia and Acidosis

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Abstract

Background: Hypokalemic Periodic Paralysis (HPP) is a rare muscle disorder characterized by episodic muscle weakness that can lead to respiratory failure. This disorder is a common manifestation of renal tubular acidosis. Renal tubular acidosis can occur associated with various systemic disorders such as Sjögren's syndrome and thyroid disorders.

Discussion: HPP is a rare presentation in distal renal tubular acidosis secondary to Sjögren's syndrome.

Conclusion: The patient was diagnosed with HPP, Sjögren's syndrome and renal tubular acidosis based on hypokalemia, metabolic acidosis, acidic urine and positive Ana profile.

Case Presentation

A 58-year-old woman complained of weakness in all extremities. Patient is a known case of hypothyroidism with history of similar complaints in past. Motor strength 2 in all four extremities. The ECG examination showed sinus bradycardia and u waves characteristic of hypokalemia. Laboratory examination found hypokalemia, metabolic acidosis with a high anion gap of 19.6 meq/L, urine pH 5.0, FT4

6.9 ng/dL, TSH 25.03 IU/mL. Anti TPO 77.7 IU/mL, ANA profile strong positive SS-A (Ro), Ro-52, SS-B (La) which indicates Sjogren's syndrome.

Introduction

Hypokalemic periodic paralysis(HPP) is the most common periodic paralysis, a rare channelopathy manifested by episodic flaccid weakness secondary to abnormal sarcolemmal excitability [1]. It is a neuromuscular disorder characterized by periodic skeletal muscle weakness that can cause respiratory muscle failure and even death [1]. The prevalence of HPP is 1 in 100,000 cases [2].

Muscle weakness depends on changes in serum potassium level (<3.5 mmol/dL) which can be due to primary or secondary causes. Primary causes are generally autosomal dominant, while secondary causes include diuretic use, loss from the gastrointestinal tract, renal tubular acidosis (RTA), primary hyperaldosteronism, Barter's syndrome, hyperthyroidism, and hypothyroidism and certain autoimmune disorders [3].

In this paper we report a case of HPP who presented with muscular weakness and eventually diagnosed to have RTA secondary to Sjogren's syndrome.

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Case Report

A 50 year old female, presented with sudden onset weakness in all four limbs and trunk since that morning. Patient couldn't move any of her limbs, neck or trunk. The weakness sudden in onset, symmetrical, equal in all four limbs, non progressive with complete loss of power. No history of stiffness, thinning of affected limbs. No history of altered sensorium, involuntary movements, visual disturbances, respiratory or bulbar weakness. No history of fasciculations, flexor spasms or sensory complaints. No history of trauma/fall or injury to the spine.

Patient also complained of non passage of urine since that morning. She was unable to feel fullness of bladder or the sensation to void. She also gave history of 2 episodes of vomiting the previous day.

Patient was previously admitted with similar complaints 3 years back, for which she was admitted in a local hospital and improved on receiving treatment of which details were unavailable. She was a known case of hypothyroidism and was currently on Tab Thyronorm 75 mcg OD.

Vitals on admission- Pulse rate- 55 bpm BP- 110/70 mmhg

Saturation- 100 % at room air

DAY 1

On admission, patient was vitally stable except she had bradycardia. On catheterisation around 600ml of urine output was obtained.

General physical examination- Pallor- Present

Icterus- Absent Clubbing- Absent Cyanosis- Absent

Lymphadenopathy- Absent Edema- Absent

Tongue- Dry

CNS examination revealed- Higher mental functions- Normal Tone- Flaccid

Power- Bilateral 0/5 Reflexes- Bilateral 2+ Plantars- Bilateral mute

Sensations- Fine touch, crude touch, temperature, vibration and joint position sense intact. No cerebellar signs.

Her lab values showed severe hypokalemia for which potassium correction was started. ABG analysis revealed metabolic acidosis with compensatory respiratory alkalosis with high anion gap.

Ph- 7.28

Pco2- 29.00

Po2- 90.40

So2- 95.1

Be- -11.8

Be(ecf)- -13.4

Bb- 36.1

Hco3- 13.4

DAY 2

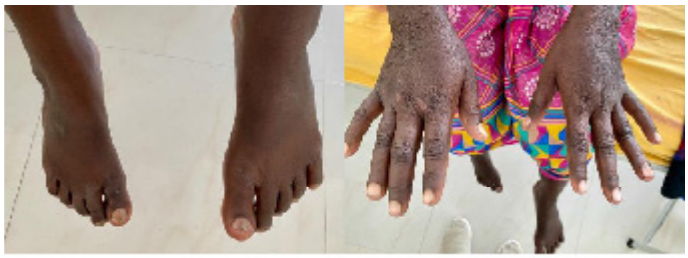
Serum potassium levels repeated and correction continued in view of low levels.

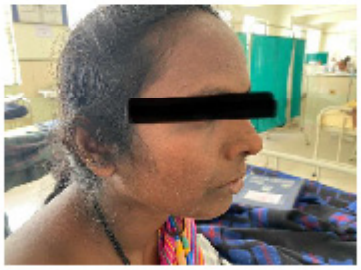
Her serum TSH levels were found to be 15.5 mcIU/ml for which the dose of oral. Thyroxine was increased from 75mcg to 100 mcg OD.

Her urine examination showed pH of more than 5.5 with normal urine osmolality and urine potassium levels of more than 20meq/l.

DAY 3

Detailed clinical examination was done to find any systemic cause for the apparent condition. It was observed that the patient had hyperpigmentation, her skin over hands and feet appeared taut and stretched. There were no features suggestive of polyarthritis, raynaud's syndrome or dysphagia. Patient also appeared dehydrated. Possibility of endocrine/ collagen vascular disorder was considered and serum cortisol levels and Ana profile were sent in view of presence of hypothyroidism and skin changes.





Date	7/8/22	9/8/22	11/8/22	13/8/22	15/8/22	17/8/22
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	MHA WITH NEUTROPHILI A					
Urea	35mg/dl					
Creatinine	0.9mg/dl		0.65mg/dl			
Totalbilirubin	0.8mg/dl					
Sgot	29 IU/L					
Sgpt	21 IU/L					
ALP	62 IU/l					
Total proteins	7.7g/dl					
Albumin	3.9g/dl					
A:G	1.1					
Sodium	139meq/l		136meq/l			
Potassium	1.4meq/l	3.55meq/l	3.56meq/l	3.25meq/l	3.71meq/l	3.73meq/l
Chloride	103meq/l					
Bicarbonate	13.4meq/l		18meq/l			
Uric acid	4.1mg/dl					
ESR	10mm 1st hr					
Urine ph	>5.5					
Urine routine and microscopy	Albumin-1+ Normal microscopy					
Urine osmolality	329mosm/kg					

Date	**7/8/22**	**9/8/22**	**11/8/22**	**13/8/22**	**15/8/22**	**17/8/22**
Urine potassium	>20meq/l					
Serum osmolality	290mosm/kg					
Total Calcium	8.2mg/dl					
Magnesium	2.7mg/dl					
Free t3	0.6ng/dl					
Free t4	6.9ng/dl					
TSH	15.03mIU/L					
Ana profile	Strongly positive for SSA, RO 52 Positive for SSB					
S. Cortisol		8 am- 6.18mcg/dl 4 pm- 5.48mcg/dl				
Lipid profile		Total cholesterol- 154mg/dl HDL- 44mg/dl LDL- 80mg/dl Triglycerides- 148mg/dl				

DAY 4

ANA profile was strongly positive for anti- SSA (Ro), Ro 52, SS-B (La). Schirmer's test revealed dry eyes.

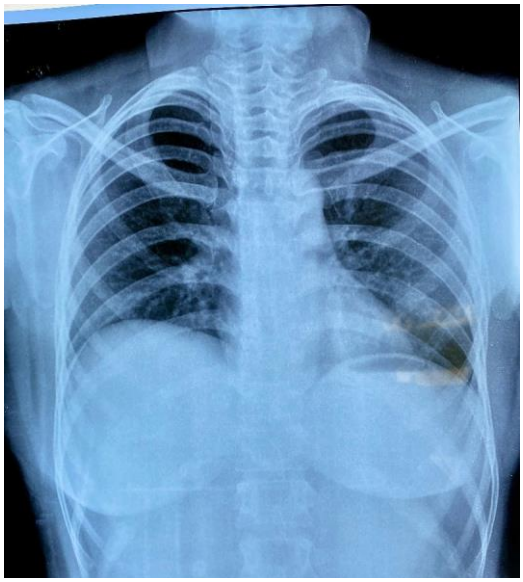
In view of persistent acidosis in spite of symptomatic improvement, diagnosis of renal tubular acidosis secondary to Sjögren's syndrome was made.

Discussion

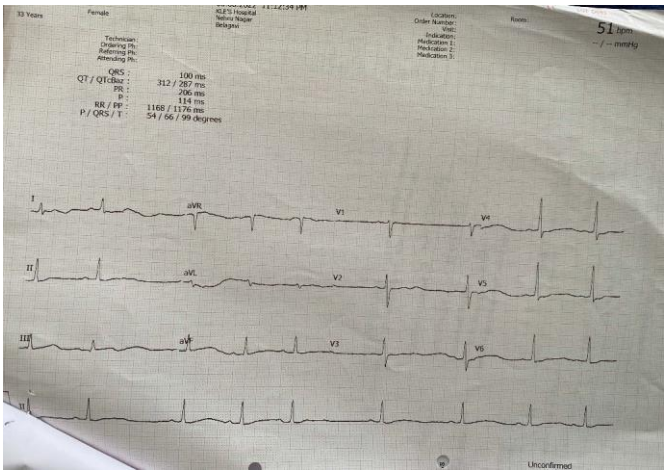
HPP is a heterogeneous disease characterized by attacks of skeletal muscle paralysis secondary to periodic drops in serum potassium levels. Hypokalemia may be secondary to potassium deficit as well as abnormal intracellular potassium shift [4].

Renal tubular acidosis is a transport defect characterised by inability of the kidneys to excrete acid (H+) and reabsorb bicarbonate (HCO3-) with clinical presentation of metabolic acidosis with normal anion gap, hyperchloremia and impaired urine acidification. Causes can be primary or secondary [5].

Distal Renal Tubular Acidosis (DRTA) is one of the rare secondary causes of hypokalemic periodic paralysis. Distal renal tubular acidosis is characterized by reduced acid secretion in



Chest x ray appeared to be normal.



Ecg showing sinus bradycardia with subtle u waves.

the urine, while proximal renal tubular acidosis is caused by impaired reabsorption of bicarbonate (HCO3-). Distal renal tubular acidosis is the most common type of renal tubular acidosis.

Distal renal tubular acidosis can be caused by primary or secondary causes. The most common "acquired" secondary causes of distal renal tubular acidosis are autoimmune diseases such as Sjogren's syndrome, systemic lupus erythematosus, rheumatoid arthritis, hypothyroidism, hyperthyroidism, Hashimoto thyroiditis. Renal tubular acidosis is very rarely reported in patients with thyroid disorders such as hypothyroidism.

Hypothyroidism can cause renal tubular acidosis through a defect in the Na-K ATPase pump in the cortical and/or tubular medulla so that the function of the Na–K ATPase pump is reduced, causing decreased H+ elimination.

Sjögren's syndrome is a systemic autoimmune disorder characterized by a unique set of signs and symptoms pre- dominantly caused by a cell-mediated autoimmunity against exocrine glands [6].

This patient had oral symptoms (dry mouth), and positive Anti-SSA (Ro) and Anti-SSB (La) and RO-52 autoantibodies, supporting the diagnosis of Sjogren's syndrome. In this patient, several exami- nations to exclude the possibility of SLE, rheumatoid arthritis.

The prevalence of renal tubular acidosis in patients with Sjogren's syndrome is 4.5-9%, generally occurring in middle age and only two- thirds are symptomatic.

Renal system involvement in Sjogren's syndrome is one of the extra-glandular manifestations and occurs in <10% of patients. Although dRTA is common in Sjö gren's syndrome, it is usually asymptomatic and in most cases it remains undetected. Hypokalemia is the most common electrolyte abnormality in patients with dRTA. The causes of hypokalemia include decreased distal tubular Na+ delivery, secondary hyperaldosteronism, defective H+-K+ ATPase, and bicarbonaturia [7].

The pathogenesis of distal renal tubular acidosis in patients with Sjogren's syndrome is not fully understood. Several studies indicate the absence or disruption of the H + -ATPase pump and the presence of autoantibodies against intercalated cells in the collecting duct and attack carbonic anhydrase (CA) in the distal nephron [8]. In addition, the presence of interstitial infiltration of lymphocytes and plasma cells that invade the tubular and epithelial membranes causes structural changes that lead to a secretory defect in the distal tubule [9].

Conclusion

Despite lack of a more comprehensive evaluation, the biochemical findings of renal potassium loss in association with HCMA were supportive of the diagnosis of distal Renal Tubular Acidosis (RTA) in our patient. Further detailed clinical examination of the patient revealed skin changes, dry mouth, which prompted us to evaluate for the possibility of Sjö gren's syndrome.

The aim of this case report was to highlight that although DRTA rarely causes HPP, it should be remembered in differential diagnosis of patients with HPP who also have hyperchloremic metabolic acidosis, hypokalemia, and a positive urinary anion gap.

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