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Case Report

Hypokalemic Paralysis Due to Renal Tubular Acidosis as the Initial Presentation of Sjogren's Syndrome

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Abstract

We report a 45-year-old female with an initial presentation with proximal renal tubular acidosis (RTA) after initial presentation with hypokalaemic paralysis and metabolic acidosis. Serological investigations eventually exhibited positive anti-nuclear antibodies (ANA), anti-Sjögren's syndrome related antigen A (SS-A), and anti-Sjögren's syndrome related antigen B (SS-B) antibodies, thus indicating the diagnosis of Sjögren's syndrome. This case is one of its kind as proximal RTA was the presenting clinical manifestation of Sjögren's syndrome. We hope that a diagnosis of Sjögren's syndrome will be considered in in patients with seemingly idiopathic RTA.

Keywords: Renal Tubular Acidosis (RTA); Sjögren's Syndrome; COVID-19

Case Report

A 45-year-old female presented to the emergency medicine department at Himalayan Hospital, Jolly Grant with chief complaints of generalized weakness of all four limbs and fatigue since the last 3 days. Her complaints were not preceded by any additional symptoms, were insidious in onset and gradually progressive until she found herself unable to stand up on her own.

She denied any history of fever, rash, sore throat, myalgia, vomiting, diarrhea, burning micturition or vaginal discharge. She gave no history of trauma, backache, neck pain, night sweats, lymphadenopathy, weight loss or soreness of muscles. She had no respiratory complaints nor any speech or swallowing difficulty. She was able to move her neck and facial muscles freely. There was no change in voice or any pain or paraesthesia.

Prior to this episode, the patient had reported no health related complaints and denied any recent swelling, breathlessness, or fluctuation of weight. The patient gave no history of COVID-19 infection or immunization.

She reported no muscle soreness, fatiguability, fasciculations, ptosis, diurnal variation in weakness or reduction of muscle mass. She gave no history of any comorbidities, similar complaints in the past or any relevant family history of such nature.

On examination, the patient was conscious and alert. She was normal in overall appearance.

The patient's blood pressure was 110/60 mmHg, respiratory rate 23/min and heart rate 74/min, regular. Her oxygen saturation was normal and she appeared to be in no distress. General examination findings were unremarkable and her cardiovascular, respiratory and abdominal examination rendered no findings.

The weakness was flaccid and involved the proximal muscle groups of bilateral shoulders and hips as well as distal extremities. Muscle strength was 3/5 in her upper and lower limbs, deep tendon reflexes were 2+ in both upper and lower limbs, no sensory deficit was found and her plantar reflexes were normal. The muscles were non tender and did not exhibit any fasciculations or reduction in girth.

The patient's hemogram, renal and hepatic functions were normal. Her chest radiograph, abdominal ultrasound were normal as well. Thyroid function tests and CPK levels were found to be normal. Her electrolytes were- sodium 140.9 mEq/l (normal 135-145 mEq/l), potassium 1.2 mEq/l (normal 3.5-5.5 mEq/l), magnesium 2.17 mg/dl (normal 1.8-2.4 mg/dl) and normal calcium and chloride levels.

A diagnosis of hypokalaemic paralysis was made and an intravenous infusion of potassium chloride was initiated. The patient exhibited marked improvement with potassium replacement and was shifted to oral potassium on recovery of the paresis.

Further investigations were conducted to determine cause for the patient's hypokalaemia. Blood gas investigations revealed metabolic acidosis (pH = 7.34), pCO2 of 26.8 mm Hg (normal 35-45 mm Hg), pO2 of 116.1 mm Hg and bicarbonate of 11.9 mEq/l (normal 22-26 mEq/l). Serum anion gap was calculated as 12 (normal 3-12). This picture was consistent with normal anion gap metabolic acidosis with a tendency for hypokalaemia. Urinalysis showed urine pH of 5.4 and a raised TTKG of 6 confirmed renal losses of potassium. The urine potassium/creatinine ratio was calculated and determined to be 1.8 further confirming renal losses.

Most causes of normal anion gap metabolic acidosis were ruled out based on history and laboratory investigations and a diagnosis of proximal RTA was determined to be the cause for the patient's hypokalaemia.

Elaborate history was drawn to determine cause for the patient's proximal RTA. There was no history of nephrolithiasis, haematuria, thrombotic events, movement disorders, ocular complaints, heavy metal exposure, long term medication or drug abuse. She denied any history of dry eyes or mouth, stiffness of joints or difficulty in swallowing.

However, the patient was investigated for possible rheumatological illnesses and an auto-antibody panel was sent. Following a positive antinuclear antibody test, an elaborate autoantibody screen reported the following results: serum SS-A(Ro) levels and SS-B(La) levels were raised.

The test report was suggestive of Sjogren's syndrome, however due to the absence of any other complaints, a negative Schirmer's test as well the patient's refusal for biopsy- the diagnosis remained probable at best.

All other antibody tests were negative. Viral hepatitis titres were negative for hepatitis B and hepatitis C, thyroid function test was normal.

The patient's final diagnosis was determined to be Hypokalaemic Paralysis attributed to Proximal RTA due to asymptomatic Sjogren's Syndrome.

Discussion and Conclusion

Weakness is a frequent and non-specific presenting complaint in many cases. Although the differential diagnosis for the complaint is elaborate, the focus may be scaled down considerably with an extensive history followed by examination [1].

Focal deficits may often be attributed to strokes or tumours whilst generalised weakness may be further sub-divided based on flaccid or spastic presentations. Further still, site on onset and progression of generalised neuromuscular weakness may also render important clues [1].

Flaccid paralysis involving all limbs with no other specific symptom often point towards metabolic causes which may be easily identified on laboratory investigation. Hypokalemic paralysis is one such readily reversible cause which on identification rapidly improves the patient's prognosis and thus must be looked for [2].

However, determining causation for the patient's hypokalaemia is where one must exhibit due diligence as this finding may be indicative of far more sinister aetiologies [2].

In our patient, evaluation of the patient's hypokalaemia rendered a diagnosis of Proximal RTA which is not as uncommon as we'd like to believe [3]. Proximal RTA is typified by a reduced rate of

bicarbonate reabsorption in the proximal tubule whilst maintaining normal transport of remaining solutes. This impairment is initially characterized as a decrease in the renal threshold for bicarbonate reabsorption with eventual metabolic acidosis [4,5].

Isolated proximal RTA is primarily autosomal recessive or sporadic. The autosomal recessive type is associated with striking features such as severe growth and mental retardation as well as ophthalmic abnormalities such as glaucoma, cataracts and band keratopathy. Sporadic isolated proximal RTA is generally diagnosed in infancy and is generally temporary; additionally it is non-familial. Hereditary forms of proximal RTA require lifelong therapy with alkali replacement [6,7].

The acquired causes of proximal RTA include multiple myeloma, amyloidosis, paroxysmal nocturnal haemoglobinuria, kidney transplantation, drugs such as ifosfamide and antiretroviral drugs as well as heavy metals like cadmium and lead [8-11].

None of which, matched our patient's history or laboratory findings.

However, another cause for proximal RTA albeit a rare one is Sjogren's Syndrome [12]. Keeping in mind, the patient's age as well as several cases reported in literature suggesting RTA as the first clinical presentation of Sjogren's Syndrome, the patient's autoantibody panel as sent and found to be positive for anti-SS-A and anti-SS-B antibodies [13-15].

Sjogren's syndrome is an autoimmune condition whose pathogenesis involves lymphocytic infiltration of exocrine glands such as salivary and lacrimal glands, resulting in typical symptoms of xerostomia (dry mouth) and xerophthalmia (dry eyes). The same immune process may also affects organs such as the skin, gastrointestinal tract, kidneys as well as lungs [12].

Although sicca symptoms are the initial presentation of Sjogren's syndrome, they may not always present. A number of case reports have indicated that the presenting features may be diverse and easily missed. Complaints such as muscle weakness, hypokalaemic paralysis secondary to RTA as well as pathological fractures have been frequently reported. Thus implying that symptoms besides sicca symptoms may present as initial symptoms of Sjogren's syndrome. In our case particularly, symptoms secondary to RTA in the absence of sicca symptoms, should incite suspicion towards the discovery of Sjogren's syndrome [5,13-17].

Sjogren's poses a significant diagnostic challenge to clinicians as it is, particularly when the initial presentation differs from the exocrine manifestation of dry eyes and mouth. The "American-European Consensus Classification Criteria" places due emphasis on the presence of these symptoms for the final diagnosis of the same. It requires four of six criteria, including: "ocular or oral symptoms, objective ocular or oral signs, histopathology from a lip biopsy and the presence of autoantibodies" [18].

Our patient only met one criteria(positive anti-SS-A and anti-SS-B antibodies) barring her refusal for the biopsy. However given her young age, possibility exists that she may meet the complete criteria in the future.

This case report highlights the importance of casting a wider net for determining aetiology of RTA. Sjogren's Syndrome commonly presents with distal RTA and there are plenty of reports in literature to attest to the same. Proximal RTA is rare unto itself in Sjogren's; however the same presenting with hypokalaemic paralysis as the first presenting feature of Sjogren's Syndrome makes this case report the first of its kind.

As this patient is yet develop the debilitating symptoms of Sjogren's Syndrome, early identification and management may vastly improve her prognosis in the future. Especially in terms of renal dysfunction which is a major contributor to mortality in these patients. The patient will be monitored for early identification of Sjogren's Syndrome as early diagnosis may allow for better control over progression and eventual outcome of the disease [16,19,20].

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