Hypokalemic Periodic Paralysis and Renal tubular Acidosis as Presenting Manifestation of Primary Sjogren's Syndrome - A Case Report

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Abstract

Among wide list of differentials of periodic paralysis, hypokalemia is an important cause. However hypokalemia with respiratory failure as sole presenting feature of Sjogren's syndrome (SS) is rarely reported. We report a case of recurrent hypokalemic paralysis which presented with life threatening respiratory failure in second episode, hypokalemia was found due to distal renal tubular acidosis and on further investigation diagnosis of primary SS was made. In this article we emphasize need for detailed work-up of a case of recurrent hypokalemic periodic paralysis in order to identify the underlying etiology and do timely management in order to avoid future recurrence.

Keywords: Hypokalemic periodic paralysis, Sjogren's syndrome, Quadriparesis.

Introduction

Sjogren's syndrome (SS) is chronic progressive autoimmune mediated disorder having both glandular mainly exocrine and extra-glandular manifestations. It has predilection for middle aged females and can sometimes be life-threatening due to respiratory involvement[1]. One of the extraglandular manifestations is Renal Tubular Acidosis (RTA) [2]. Renal involvement in primary Sjogren's syndrome (pSS) is estimated between 18.45% and 67% [3]. It leads to hypokalemia due to underlying renal tubular acidosis. Although flaccid quadriparesis has a long list of differentials with hypokalemia as one of the cause, it is rarely reported as a presenting feature of underlying pSS.

Case Report

A thirty seven year old married female presented with history of dryness of eyes and mouth and gritty sensation in eyes for six months , history of quadriparesis followed by complete recovery five months back (no previous documentation was available with patient), two days history of acute onset rapidly progressive flaccid quadriparesis and respiratory failure for one day. Patient was on ventilator support on emergency arrival. Her BP was 110/80 mmHg, pulse rate 96/min and she was tachypneic with respiratory rate 38/min. She was immediately shifted to intensive care unit and continued on ventilator support. There was no h/o Fever, preceding respiratory illness, re-

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current abortions, joint pains, diabetes, alcohol abuse, high carbohydrate intake, diarrhea, diuretic use, vomiting, excessive exercise or sweating, drug intake or illicit drug abuse. On examination she was conscious, oriented, meningeal signs were negative. She had flaccid quadriparesis, diffuse hyporeflexia, bilateral absent plantars and weak neck flexors. There was no cranial nerve, bladder, bowel or sensory involvement, no eye signs. Her lab investigations were (Table 1), ABG revealed metabolic acidosis with pH 7.12 and normal anion gap. Hypokalemia was corrected with intravenous potassium chloride, for renal tubular acidosis sodium bicarbonate was given in dose of 0.5-2mmol/kg in four divided doses.

Patient showed gradual recovery and she was weaned off from ventilator on third day of admission. Her power increased to MRC grade 4/5 on seventh day of admission. Based on clinical presentation and laboratory features provisional diagnosis of Sjogren's syndrome was made according to SICCA 2012 criteria, subsequently patient was planned for salivary gland and renal biopsy, but patient refused.

So with diagnosis of pSS patient was discharged on low dose oral steroids, potassium chloride supplementation with regular potassium monitoring and was followed up after a month. Gradually steroids were tapered and Hydroxychloroquine was started in divided doses , since then patient was kept under follow up every two months for next 6 months and she was doing fine till last follow up.

Discussion

SS is an autoimmune disorder with progressive course and predominant exocrine gland involvement due to lymphocytic infiltration[4,5]. It has an estimated prevalence of 0.5-1%, female predominance and is further sub-classified into primary or secondary types[3]. One of the largest case series of 13 patients reported from India who presented as hypokalemic paralysis and later diagnosed as pSS, all patients were female with median age of 33.1+8.22 years [6].

Majority of patients presents with Sicca symptoms while others have extra glandular features amongst which renal involvement is well-recognized[7]. Most common renal involvement is related to tubular dysfunction and can manifest as Distal Renal Tubular Acidosis (dRTA) with reported frequency 25-40%, oth-

Table 1 : Laboratory investigations of patient

Lab Investigation	Result
CBC	Hb:7.6gm/dl , WBC:7.8× 10^3, Platelets:307K/μl, ESR:31mm/h
Serum Iron profile	Ferritin:33.38ng/ml , Iron:48$\frac{\mu g}{dl}$, $TIBC$: $\frac{292.5mcg}{dl}$, $UIBC$: 244.5$\mu g/dl$
Serum Biochemistry	RBS:108mg/dl, Urea:52.9mg/dl, Creatinine:1.3mg/dl
Liver function test	Total bilirubin:0.2mg/dl, AST:26.4IU/l, ALT:14IU/l, A:G ratio:0.86
Serum Electrolytes	Sodium:148mmol/L, Potassium:2mmmol/L, calcium:7.05mg/dl, Magnesium:3.45mmol/L, Phosphorous:2.15mmol/L
Urinanalysis	pH:6.8, Leucocytes : 2/3/hpf, RBC: Nil
Urinary Electrolytes	Sodium:44mEq/l, Chloride:71mEq/l, Phosphorous:10.6mg/dl, calcium:5.9mg/dl, 24HR protein:528mg
TT4,TSH,iPTH,Cholicalciferol	7.8$\mu g/dl$, 1.15, 213.40pg/ml, 11.9ng/ml
HBsAg, HCV, HIV	Non-reactive
ANA by IFA	1:3200 nuclear speckled 4+
APLA, anti-dsDNA, anti-Sm, RF, antiU1snRNP	Negative
SSA/Ro 60kdAb, SSA/Ro 52kdAb, SSB/La	59AU/ml, 78AU/ml, 26AU/ml
Schrimer's test at 5min	Left eye:2mm , Right eye:3mm
CXR , Serum ACE level	Normal, 34μl
USG KUB region	No evidence of nephrocalcinosis

ers include proximal tubular acidosis, tubular proteinuria, and nephrogenic diabetes insipidus [8]. dRTA may be presenting manifestation of autoimmune diseases in adults [9]. Correlation between RTA and high anti-SS A/Ro and anti-SS B/La level titres has been proposed as surrogate marker of disease progression in some studies [3,10].

Hypokalaemia is a late presenting feature of dRTA and develops due to mechanisms which include defective H+-K+ATPase and secondary hyperaldosteronism. Among types of hypokalaemia, secondary hypokalemia due to acquired cases like thyrotoxicosis, hyperaldosteronism, diabetic ketoacidosis, diarrhea, vomiting, drugs or RTA are more common [11]. Periodic paralysis secondary to hyokalemia as presenting feature of Sjogren's syndrome has been documented in <2% of cases [3].

The clinical presentation of hypokalemic quadriparesis with respiratory failure as the first manifestation of pSS is very atypical. Our patients presented with flaccid quadriparesis with respiratory involvement without any bladder, bowel or sensory involvement without any prior history of fever, joint pain, rashes, loose motions, high carbohydrate intake, prolonged exercise, recurrent abortions. Based on clinical presentation, differential diagnosis of GB syndrome and Hypokalemic Periodic Paralysis (HPP) was considered. Laboratory investigations suggested hypokalaemia and dRTA. NCS was done which was suggestive of conduction block in left common perineal and bilateral tibial nerves.

Literature regarding this entity is in form of case reports and case series due to rarity of this entity. Eighteen cases were described between 1966 and 2004 and till 2010 another 25 cases were reported. According to case series of hypokalaemia periodic paralysis on 31 cases only 10% were diagnosed with SS [12].

We report this case to highlight the fact that patients presenting with HPP can be due to early SS which needs to be diagnosed so as to prevent future hypokalemic attacks and complications .

Conclusion

A patient presenting with flaccid quadriparesis should be investigated in detail which should include good history taking, thorough neurological examination and lab work up so as to identify underlying etiology at an early stage and plan management in order to avoid recurrent episodes.

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