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(CASE REPORT)

A case report on Sjogren syndrome with autoimmune liver disorder

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Abstract

Background: Primary Sjogren's syndrome (pSS) is a chronic systemic and autoimmune disorder characterized by inflammation and dysfunction of exocrine glands. Liver involvement was the first extraglandular manifestations to be reported in patients with pSS. Tubulointerstitial nephritis (TIN) is the main renal involvement associated with pSS and manifest as distal renal tubular acidosis (RTA), and others, of which RTA is the main clinical presentation. RTA has been reported in 4.3 to 9% of pSS patients. In this case, patient experiences RTA.

Case Presentation: A 19 year old female patient was presented with complaints of abdominal pain, fatigue for 2 months and loose stools. She had history of autoimmune hepatitis and decompensated liver cirrhosis with portal hypertension (autoimmune), bilateral peroneal axonal neuropathy, grade II hepatic encephalopathy resolved and recurrent pustular lesion for past 2 years which rupture spontaneously and heals with scary. She also presented with recurrent hypokalemia which was corrected. No history of similar complaints among the family members.

Conclusion: This case explains the autoimmune liver disorder with newly diagnosed primary sjogren's syndrome associated with recurrent hypokalemia.

Keywords: Sjogren's syndrome; Autoimmune liver cirrhosis; Distal RTA; Hypokalemia; Peroneal neuropathy

1. Introduction

Sjogren's syndrome is a chronic, systemic, autoimmune disease with inflammation and dysfunction of exocrine glands resulting sicca symptoms. About 65% of pSS patients can experience extra glandular features including pulmonary, gastrointestinal, hematologic and neurologic disorders [1]. Primary SS (pSS) is differentiated from secondary SS by the non-appearence of other autoimmune diseases, like rheumatoid arthritis, systemic sclerosis, and systemic lupus erythematosus [2]. Renal involvement is a rare complication of Pss which occurs in 10% of patients but this can be managed appropriately. Tubulointerstitial nephritis (TIN) is the most common renal complication of primary sjogren's syndrome (pSS) [3]. This complication responds to steroid therapy alone. The pathogenesis of SS is unknown, but several etiological mechanisms have been proposed.

The association between liver disease and pSS was considered as surprisingly common [4] that is seen in many case studies. Hypokalemic paralysis is another initial symptom in seven percent of patients with Sjogren's syndrome [5].

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2. Case report

This is a case of 19 year old female patient presented with complaints of abdominal pain, fatigue for 2 months and loose stools. She had a history of autoimmune hepatitis and decompensated liver cirrhosis with portal hypertension (autoimmune), bilateral peroneal axonal neuropathy and grade II hepatic encephalopathy resolved. She also had a history of recurrent pustular lesion for past 2 years which rupture spontaneously and heals with scary. And also recurrent hypokalemia was noted. No history of similar complaints in the family members. On physical examination she was conscious, oriented, afebrile, mild pallor and also presented with sicca symptoms (dryness of mouth and eyes).

General examination revealed conscious, oriented, afebrile and mild pallor. Hematological examinations was found to be normal except ESR (67 mm/hr). LFT showed serum protein and serum globulin was elevated. RFT results revealed declined potassium (hypokalemia- 2.6 mmol/L) and elevated creatinine levels (1.8 mg/dl). Urine routine examination showed pus and epithelial cells were slightly elevated. USG abdomen and pelvis conveyed chronic liver disease with splenomegaly and bilateral renal parenchyma changes with medullary nephrocalcinosis suspected. Patient had persistent hypokalemia, raised creatinine levels and worsening renal function. Nephrology opinion was taken and diagnosed as Autoimmune CLD with autoimmune nephritis then suggested to continue the same medications and monitor RFT daily. She had active lesions over gluteal area. Dermatology consultation was sought and was diagnosed as Hidradenitis Sappurativa and advised to start medications for 10 days. Rheumatology consultation was sought in view of anti RO positivity and diagnosed as primary Sjogren with distal RTA.

Initially the patient was started with IV antibiotics like Inj. MONOCEF (CEFTRIAXONE 1gm IV BD), Inj. METROGYL (METRONIDAZOLE 500mg IV BD), oral antibiotics like T. RIFAGUT (RIFAXIMIN, 550mg BD), multivitamins (C. BECOSULES B COMPLEX FORTE WITH VITAMIN C + ZINC) and proton pump inhibitors (Inj. PANTOP PANTOPRAZOLE 40 mg BD) and potassium correction. Patient continued her own medication (T. WYSOLONE PREDNISOLONE 5mg OD, T. MMF MYCOPHENOLATE MOFETIL 500 mg BD). Nephrology opinion was sought for raised creatinine and declined potassium levels with Syp. POTKLOR (POTASSIUM CHLORIDE) 15 ml TDS. PEROCLIN GEL (CLINDAMYCIN + BENZOYL PEROXIDE) 2.5% and T- BACT OINTMENT (MUPIROCIN) L/A OD was used to treat pustular lesion in the body. On 4th day of hospital stay, patient was symptomatically improved and hence discharged with medications such as T. METROGYL (METRONIDAZOLE) 400mg PO BD for 3days, T. RIFAGUT (RIFAXIMIN) 400mg BD for 5days, T. RABIUM (RABEPRAZOLE) 20mg OD, PEROCLIN GEL for L/A OD, T BACT OINTMENT for L/A OD. Discharge recommendations include salt restricted and protein rich diet, avoid hepatotoxic medications.

3. Discussion

Primary sjogren syndrome is an autoimmune disease affects females than males. The difficulty in diagnosis is reflected on continuous review attempts of the 7 classification criteria that have been created in the past 25 years [6]. Renal imvolvement in pSS include TIN and glomerulopathy. The pathogenesis of renal involvement in pSS remains unclear. Tubulointerstitial nephritis can manifest as dRTA which was the common renal manifestation. dRTA is characterized by inability to acidify the urine maximally in the presence of systemic acidosis. Hypokalemia is the most common electrolyte abnormality in patients with dRTA. In many case studies, hypokalemic paralysis was present due to distal RTA in pSS patients.

Treatment of pSS is directed towards the particular areas of the body that are involved and complications. pSS can be treated with skin creams, immunosuppressants and corticosteroids. Already the patient was taking corticosteroid and immunosuppressant. These are the best option to prevent the symptoms of pSS and autoimmune liver disorders. There is no cure or remittive management for this condition. At present, only prevention is available. There is a chance of getting recovered from liver disorder which is liver transplantation which is advised to the patient. Patient was followed up and further recommended for the transplantation.

4. Conclusion

This autoimmune disorder results from lymphocytic infiltration of lacrimal and salivary glands. Corticosteroids and immunosuppressants are the best treatment option to prevent the symptoms a complications of Primary Sjogren's syndrome and autoimmune liver disorders. There is no cure or remitive management, therefore liver transplantation is another option, but chance of failure is more. The patients with these condition will suffer more severe disease and death incidence. More attention should be needed and should provide a better symptomatic treatment during -clinical practice.

Compliance with ethical standards

Disclosure of conflict of interest

There is no conflict of interests regarding the publication of this paper.

Statement of informed consent

Informed consent was obtained from all individual participants included in the study.

References

[1] Garza-Alpirez A, Arana-Guajardo AC, Esquivel-Valerio JA, et, al. Hypokalemic paralysis due to primary Sjogren syndrome: case report and review of the literature. Case reports in rheumatology. 2017.

[2] Manthorpe R, Asmussen K, Oxholm P. Primary Sjogren's syndrome: diagnostic criteria, clinical features and disease activity. J Rheumatol 1997; 24(50):811.

[3] Yang ML, Kuo MC, Ou TT, Chen HC. Primary Sjogren syndrome with minimal change disease- a case report. The Kaohsiung journal of medical sciences. 2011: 27(5):190-4.

[4] Whaley K, Goudie RB, Williamson J, et al. Liver disease in Sjogren's syndrome and rheumatoid arthritis. Lancet. 1970; 1:861-63.

[5] H. Ren, W. M. Wang, X. N. Chen, et al. Renal involvement and followup of 130 patients with primary Sjogren's syndrome. The Journal of rheumatology. 2008; 35(2):278–84.

[6] Baldini C, et. al. Classification criteria for Sjogren's syndrome: a critical review. Jounal ofautoimmunity. 2012; 39(1-2): 9-14.