The ESRF Research Journal for Undergraduate Medical Students

Case Report

Volume 3 Issue 2 (Jan-Jun) 2026

Submission: 28th December, 2025

Acceptance: 06th April, 2026

DOI: 10.5281/zenodo.19633815

Corresponding Author

Dr. Suchisubhra Roy
Intern,
Nil Ratan Sircar Medical College and Hospital
e-mail: suchisubhra-roy1206@gmail.com

Available from: https://esrfrjums.co.in/index.php/main/article/view/93

Published by: Eureka Scientech Research Foundation, Kolkata.

Online access: https://esrfrjums.co.in

Full-house Membranous Nephropathy in a Case of Primary Sjögren's Syndrome :
A Lion's Mane on a Lamb

Suchisubhra Roy[1], Tushar Modak[2], Subhajit Das[2], Angan Karmakar[3], Badal Kumar Sahu[3], Arijit Sinha[4]

[1]*Intern, Nil Ratan Sircar Medical College, Kolkata*

[2]*Post Graduate Trainee, Department of General Medicine, Nil Ratan Sircar Medical College and Hospital, Kolkata*

[3]*Assistant Professor, Department of General Medicine, Nil Ratan Sircar Medical College and Hospital, Kolkata*

[4]*Professor, Department of General Medicine, Nil Ratan Sircar Medical College and Hospital, Kolkata*

Abstract

Primary Sjögren's syndrome is a systemic autoimmune condition affecting primarily the exocrine glands, with an incidence rate of 6.92 per 100,000 person-years (95% CI 4.98 to 8.86). Initial presentation of this disease varies widely, depending on the level of systemic involvement. Renal involvement remains a concern in affected patients with a recent study showing almost a 5% rate of the same, although specifics remain difficult to characterise. This diversity of symptoms makes it difficult to finally diagnose patients with Sjögren's syndrome.

This case report emphasises the importance of a thorough clinical history and investigations in diagnosing primary Sjögren's syndrome. The initial presentation was non-specific generalised weakness due to hypokalaemia, while the renal biopsy in the end even pointed towards possible lupus nephritis further complicating the clinical picture. Such diverse and often overlapping manifestations underscore the diagnostic challenges associated with Sjögren's syndrome.

Keywords: Sjögren's syndrome, hypokalemia, renal tubular acidosis, full-house membranous nephropathy, anaemia of chronic disease

INTRODUCTION

Primary Sjögren's syndrome is a systemic autoimmune condition affecting primarily the exocrine glands, with an incidence rate of 6.92 per 100,000 person-years (95% CI 4.98 to 8.86).[1] Initial presentation of this disease varies widely, depending on the level of systemic involvement. Renal involvement remains a concern in affected patients with a recent study showing almost a 5% rate of the same, although specifics remain difficult to characterize.[2,3] This diversity of symptoms makes it difficult to finally diagnose patients with Sjögren's syndrome.

This case report emphasises the importance of a thorough clinical history and investigations in diagnosing primary Sjögren's syndrome. The initial presentation was non-specific generalised weakness due to hypokalaemia, while the renal biopsy in the end even pointed towards possible lupus nephritis further complicating the clinical picture. Such diverse and often overlapping manifestations underscore the diagnostic challenges associated with Sjögren's syndrome.

CASE PRESENTATION

A 31-year-old woman, with no prior history of hypertension or diabetes mellitus, presented to the outpatient department of General Medicine, at a tertiary care hospital in Kolkata on 8/10/2025, with complaints of progressive generalized weakness and fatiguability for the past four months. The weakness was insidious in onset, gradually progressive, and exacerbated by moderate exertion. There was no associated muscle tenderness, fasciculations, or proximal muscle wasting.

She additionally reported polyuria and polydipsia, characterized by the passage of large volumes of dilute urine, not accompanied by dysuria, hematuria, or flank pain There was no history of vomiting,

diarrhea, or use of diuretics or laxatives, thereby ruling out gastrointestinal or drug-induced potassium losses. The patient also noted dryness of mouth, difficulty swallowing dry food, and a gritty or foreign-body sensation in both eyes. There was no history of arthralgia, rash, photosensitivity, alopecia, oral ulcers, or parotid gland enlargement. Features suggestive of thyroid dysfunction or other connective tissue disorders were absent. Appetite and menstrual cycles were normal. There was no family history of autoimmune diseases.

The patient was moderately built and on general survey at admission, she was found to be afebrile, with no pallor, icterus, cyanosis, clubbing or pedal edema. Vitals were maintained at 110/70 mm Hg blood pressure, 78 beats per minute heart rate and a Sp02 of 99%. Systemic examination showed normal heart sounds with no murmur, bilaterally clear vesicular breath sounds along with a soft and non-tender abdomen with no organomegaly. Patient was alert and conscious with no focal neural deficits. ENT examination revealed dry, erythematous, sticky mucosa with reduced salivary pooling orally. Schirmer's test revealed reduced tear flow in at least one eye.

On admission, initial laboratory evaluation revealed a hemoglobin level of 7.6 g/dL and an elevated erythrocyte sedimentation rate (ESR) of 43 mm/hour. Total leukocyte count (6,700/µL) and platelet count (2.6×10^5/µL) were within normal limits. The patient was transfused with one unit of packed red blood cells, following which hemoglobin rose to 10.8 g/dL. Serum biochemistry showed a urea level of 26 mg/dL and a marginally elevated creatinine level of 1.3 mg/dL. Serum sodium was 137 mmol/L, while potassium was reduced to 2.8 mmol/L. Bicarbonate was decreased (13 mmol/L) with a corresponding elevation in chloride levels to 112 mmol/L, yielding an anion gap of 8 mEq/L, indicating a normal anion gap (hyperchloremic) metabolic acidosis. The total bilirubin level was within the normal range at 0.5 mg/dl. SGOT, SGPT and ALP levels were also normal at 26 IU/L, 29 IU/L and 86 IU/L respectively. Total protein was 8.1 g/dl with globulins slightly elevated at 4.3 g/dl. A lipid profile of the patient was normal except for a borderline low HDL (39 mg/dl). A complete autoimmune profile revealed positivity for ANA, Anti-Ro (SSA), Anti-La (SSB), Ant Ro 52 and JO 1, indicating Sjögren's to be a strong differential diagnosis, and negative for Anti-dsDNA, ANCA, Anti-CCP and myositis profiles. RF factor levels were slightly elevated at 79.1 IU/ml.

An ultrasonography of the whole abdomen demonstrated both kidneys to be normal in size and echotexture, with no evidence of nephrocalcinosis or hydronephrosis. A small collection of free fluid was noted in the pouch of Douglas. Ultrasonography of the neck revealed bilateral parotid glands to be mildly enlarged with diffuse heterogeneity of echotexture and the presence of multiple small hyperechoic foci. The submandibular glands also appeared mildly heterogeneous. Multiple small, discrete, oval-shaped lymph nodes were visualized in bilateral cervical regions (levels II–V), the largest measuring approximately 9–10 mm in short axis. The thyroid gland appeared normal in size, outline, and echotexture, with no focal nodules or calcifications. Overall, the features were suggestive of chronic sialadenitis, of likely autoimmune origin, associated with a mild reactive cervical lymphadenopathy. A PET-CT scan performed on 22/10/2025 revealed FDG-avid bilateral parotid glands and cervical lymph nodes, indicating granulomatous or inflammatory changes (Figure 2). Echocardiography showed no vascular abnormalities with a normal ejection fraction of 61%. Kidney needle biopsy reports indicated a "full house pattern" (Figure 1).

The clinical features of sicca and xerostomia and autoimmune profile allowed us to finally diagnose the patient with primary Sjögren's syndrome with distal (Type 1) renal tubular acidosis along with anemia of chronic disease, with hypokalaemia secondary to the distal renal tubular acidosis. Renal biopsy reports also allowed us to diagnose the patient with full-house nephropathy (FHN). Oral potassium replacement with Potassium Chloride syrup was done to manage hypokalemia, while normal anion gap metabolic acidosis (NAGMA) was treated with bicarbonate therapy and immunomodulation was done using hydroxychloroquine. Sicca symptoms showed improvement with oral pilocarpine tablets and oral hydration. Therapy was tolerated well by her and she showed biochemical and symptomatic improvements.

At the time of discharge, she was hemodynamically stable, afebrile, well-oriented and accepting of oral diet with normal urine output. Potassium levels were also stabilzed with no weakness in patient. Her discharge advice consisted of oral medications, namely, hydroxychloroquine 200mg, pilocarpine 5 mg, pantoprazole 40mg, and potassium chloride syrup 15ml. Additionally carboxymethylcellulose eye drop was prescribed to continue and she was encouraged to drink plenty of fluids. Immunosuppression with cyclophosphamide was advised to the patient to combat her decreasing renal function. Patient was asked to review at medicine or rheumatology outpatient department after 2 weeks with her serum electrolyte levels. Ophthalmology and dental consultations were advised for sicca symptoms and management and patient was asked to report to the emergency in case of warning signs like weakness, cramps or decreased urine output.

DISCUSSION

Sjögren's syndrome is a chronic autoimmune disease, best characterised by lymphocytic infiltration of the exocrine glands and profound B-cell hyperactivity. It is classically divided into two major types: primary when it presents by itself without any other underlying autoimmune disease and secondary when it presents in conjunction with some other autoimmune etiology like systemic lupus.[4] Sjögren's has become increasingly common as a rheumatological disease with its prevalence estimated to be about 0.5% and incidence at about 4/1000 people per year. Middle-aged women are more likely to be affected although it may present in any age group. However, females are almost 10 to 20 times more likely to be afflicted by this disease.[5]

Symptoms arise mainly due to two mechanisms - infiltration of primarily the salivary and lacrimal glands along with B lymphocyte hyperactivity which is manifested by hypergammaglobulinemia and serum antibodies against non-organ-specific antigens like immunoglobulins (rheumatoid factors) and extractable cellular

antigens (Ro52, Ro60 and La). This syndrome is initiated as result of combination of various endogenous and exogenous factors like genetic expression, environmental influences, hormones and immune dysregulation which act as triggers.[6] Symptoms in Sjögren's syndrome are mostly benign with oral symptoms including dryness, difficulty swallowing dry food, a burning mouth sensation and increased dental caries among others while ocular symptoms include burning and accumulation of secretions in thick strands at the inner canthi.

Initial presentation in our patient was atypical, with patient presenting with generalised weakness. On further clinical and laboratory work-up the raised ESR and reduced haemoglobin level of 7.6 g/dl with a normocytic and normochromic blood smear indicated severe anemia of chronic disease. Further history taking led us to the findings of xerostomia and sicca, which then prompted us to do a complete autoimmune evaluation of the patient which revealed anti-Ro (SSA), anti-La (SSB), and anti Ro 52 positive all indicating a diagnosis of Sjögren's syndrome.

At this point of our clinical work-up we were also concerned about the lower than 8 g/dl threshold of hemoglobin which pointed to a more sinister origin than a simple case of anemia of chronic disease. This prompted us to look into the renal and haematological systems and consequently order a renal and bone marrow biopsy. While the bone marrow biopsy report came out normal, the immunostaining pattern for the kidney biopsy came positive for IgA, IgG, IgM, C3 and C1q. Speculation for a diagnosis of SLE is a common mistake made in the presence of full-house membranous nephropathy; however, we must remind ourselves that renal biopsy reports cannot conclusively diagnose a patient with systemic lupus erythematosus (SLE) in the absence of any corresponding clinical features.[7,8] Consequently, the diagnosis of non-lupus "full-house" nephropathy was made. Non lupus FHN is a rare entity and in the absence of proper clinical examination we might even overestimate its pathogenicity by misdiagnosing a patient with conventional lupus nephritis and as a result SLE. It is therefore necessary to isolate the lion's mane from the lamb before diagnosing SLE based solely on renal biopsy reports. Our case report accordingly highlights the role of comprehensive history taking in a case of Sjögren's syndrome.

Renal involvement in Sjögren's syndrome as a spectrum consists of distal or proximal renal tubular acidosis, glomerulonephritis and acute or chronic tubulointerstitial nephritis. A vast majority of the hypokalaemia seen in pSS and even in our patient can actually be attributed to distal RTA.[9] A possible explanation for this could be the complete absence of vacuolar H(+)-ATPase pump in the collecting duct in patients with Sjögren's syndrome.[10]

Management of pSS has remained relatively unchanged over the years with treatment principally based on managing sicca and other systemic features.[11] Artificial tear drops are mainstay in treatment while systemic conventional immunomodulating drugs like hydroxychloroquine have shown modest benefits.[12] Certain recent randomised clinical trials have even shown marginal benefits to muscarine receptor agonists like pilocarpine and cevimeline for relief of sicca symptoms.[13] Additionally renal involvement in the disease needs to be managed accordingly with immunosuppression as and when needed.

CONCLUSION

Distal RTA and hypokalaemia are increasingly recurring features seen in a case of Sjögren's syndrome. Making the relevant connections are therefore important in a case of progressive generalised weakness with symptoms of sicca and hypokalaemia in order to arrive at the correct diagnosis of an autoimmune condition. Additionally, distinction between non lupus FHN and the more familiar lupus FHN must also be made in any autoimmune workup with the help of clinical and laboratory results.

CONFLICT OF INTEREST

None Declared

FUNDING

None Declared

FIGURES

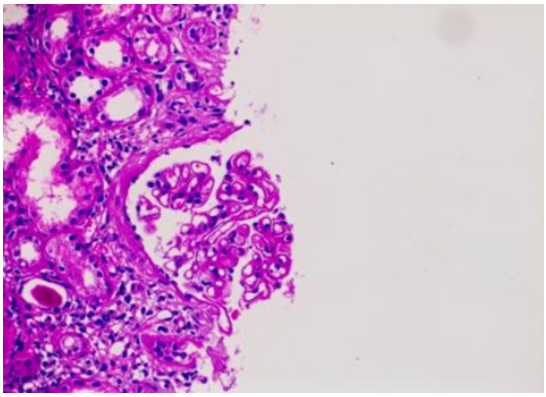

Figure 1: Microscopic section of H&E stained renal cortical parenchymal biopsy core

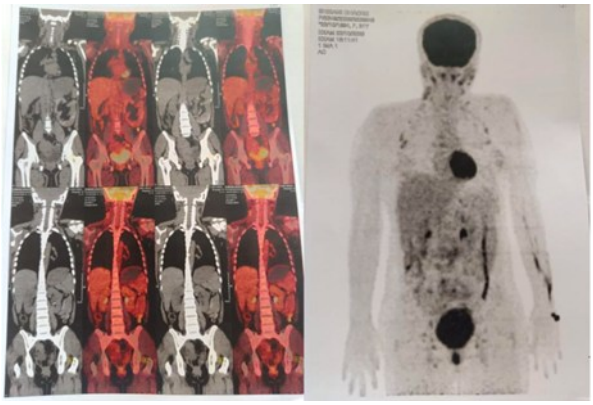

Figure 2: PET CT Scan showing FDG-avid bilateral parotid glands and cervical lymphnodes, suggestive of granulomatous/ inflammatory changes

REFERENCES

1. Qin B, Wang J, Yang Z, Yang M, Ma N, Huang F, et al. Epidemiology of primary Sjögren's syndrome: a systematic review and meta-analysis. Annals of the Rheumatic Diseases [Internet]. 2014 Jun 18;74(11):1983–9. Available from: https://doi.org/10.1136/annrheumdis-2014-205375.
2. Tejaswee B, Swapnil T, Pankaj S, Kumar BM, Krishna BG. Distal RTA with Hypokalemic Quadriparesis as the Initial Presentation of Primary Sjogren's Syndrome. Clinical Medical Reviews and Case Reports [Internet]. 2021 Jan 30;8(1). Available from: https://doi.org/10.23937/2378-3656/1410336.
3. Goules AV, Tatouli IP, Moutsopoulos HM, Tzioufas AG. Clinically Significant renal involvement in Primary Sjögren's Syndrome: Clinical presentation and outcome. Arthritis & Rheumatism [Internet]. 2013 Oct 28;65(11):2945–53. Available from: https://doi.org/10.1002/art.38100.
4. Shahane A, Patel R. The epidemiology of Sjögren's syndrome. Clinical Epidemiology [Internet]. 2014 Jul 1;6:247. Available from: https://pmc.ncbi.nlm.nih.gov/articles/PMC4122257/
5. Shahane A, Patel R. The epidemiology of Sjögren's syndrome. Clinical Epidemiology [Internet]. 2014 Jul 1;6:247. Available from: https://pmc.ncbi.nlm.nih.gov/articles/PMC4122257/
6. One year in review 2020: pathogenesis of primary Sjögren's syndrome [Internet]. PubMed. 2020. Available from: https://pubmed.ncbi.nlm.nih.gov/33025887/
7. Weening JJ, D'agati VD, Schwartz MM, Seshan SV, Alpers CE, Appel GB, et al. The classification of glomerulonephritis in systemic lupus erythematosus revisited. Kidney International [Internet]. 2004 Jan 8;65(2):521–30. Available from: https://doi.org/10.1111/j.1523-1755.2004.00443.x
8. Stokes MB, D'Agati VD. Full-house glomerular deposits: beware the sheep in wolf's clothing. Kidney International [Internet]. 2017 Dec 29;93(1):18–20. Available from: https://doi.org/10.1016/j.kint.2017.08.001.
9. Pertovaara M, Korpela M, Kouri T, Pasternack A. The occurrence of renal involvement in primary Sjögren's syndrome: a study of 78 patients. Lara D Veeken [Internet]. 1999 Nov 1;38 (11):1113–20. Available from: https://doi.org/10.1093/rheumatology/38.11.1113.
10. DeFranco PE, Haragsim L, Schmitz PG, Bastani B. Absence of vacuolar H(+)-ATPase pump in the collecting duct of a patient with hypokalemic distal renal tubular acidosis and Sjögren's syndrome. Journal of the American Society of Nephrology [Internet]. 1995 Aug 1;6(2):295–301. Available from: https://doi.org/10.1681/asn.v62295.
11. Ramos-Casals M, Brito-Zerón P, Sisó-Almirall A, Bosch X, Tzioufas AG. Topical and systemic medications for the treatment of primary Sjögren's syndrome. Nature Reviews Rheumatology [Internet]. 2012 May 1;8(7):399–411. Available from: https://pubmed.ncbi.nlm.nih.gov/22549247/
12. Vitali C, Minniti A, Pignataro F, Maglione W, Del Papa N. Management of Sjögren's Syndrome: Present issues and Future Perspectives. Frontiers in Medicine [Internet]. 2021 Jun 7;8:676885. Available from: https://doi.org/10.3389/fmed.2021.676885
13. Brito-Zerón P, Retamozo S, Kostov B, Baldini C, Bootsma H, De Vita S, et al. Efficacy and safety of topical and systemic medications: a systematic literature review informing the EULAR recommendations for the management of Sjögren's syndrome. RMD Open [Internet]. 2019 Oct 1;5(2):e001064. Available from: https://pubmed.ncbi.nlm.nih.gov/31749986/