

Case Report

A male presenting with polyuria: a case of primary Sjögren syndromeSatharasinghe DS¹, Atukorale SH², Weerawansa P³, Senanayake HMS³¹Postgraduate Institute of Medicine, University of Colombo, Sri Lanka²Department of Rheumatology, Teaching Hospital, Anuradhapura, Sri Lanka³Department of Medicine, Faculty of Medicine and Allied Sciences, Rajarata University of Sri Lanka, Sri Lanka**Abstract**

Sjögren syndrome is a chronic autoimmune disorder with lymphocytic infiltration of exocrine glands causing oral and ocular dryness and well recognized systemic manifestations. Although primary Sjögren syndrome (pSS) is common in women, it is not well recognized in men. This is the first reported case of pSS in a Sri Lankan male to the best of our knowledge.

A 58-year-old man presented with polyuria. He has had sicca symptoms for six months and polyuria was secondary to the increased water intake to relieve oral dryness. Examination revealed enlarged tender bilateral salivary glands. After excluding secondary causes, he was diagnosed with pSS. The treatment commenced with steroids, azathioprine, and symptomatic therapy.

Primary Sjögren syndrome is an overlooked diagnosis, especially in men, leading to increased morbidity given its multi-organ involvement. Recognition and supportive therapy can help to improve the quality of life of patients.

Key words: *Primary Sjögren Syndrome, sicca symptoms, polyuria, polydipsia*

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Funding: None

Competing interests: None

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Cite this article as: Satharasinghe et al. A male presenting with polyuria: a case of primary Sjögren syndrome. *Anuradhapura Medical Journal* 2020; 14 (1): 13-16
DOI: <http://doi.org/10.4038/amj.v14i1.7659>

Sjögren syndrome (SS) is an autoimmune disorder and it has the highest prevalence in females among all rheumatological conditions. We report a male patient diagnosed with primary Sjögren syndrome (pSS) and negative anti-Ro/La antibody contrary to the common serological pattern seen in males.

Patient information

A 58-year-old man was admitted for evaluation of polyuria and polydipsia for six months. During ward stay, his measured urine output was not sufficient to describe as polyuria. On further inquiry, he revealed an exhaustive history of dry mouth leading to psychogenic polydipsia. He had a recurrent sensation of grittiness in the eye and lethargy, malaise, and weight loss of 5kg over the last four months.

He had no history of head or neck irradiation therapy and denied sexual promiscuity. He had no shortness of breath, numbness, or urinary symptoms. He had no joint pain, stiffness, skin rashes, hair loss, mouth ulcers, Raynaud's phenomenon, or skin thickening.

On examination, his pulse rate was 80 bpm, and the blood pressure was 110/80 mmHg. His BMI was 17 kg/m². He had poor dental hygiene and enlarged, tender, firm salivary glands. The rest of the examination was normal.

His white blood cell count was $9.1 \times 10^9/L$, haemoglobin 10g/dL, platelet count $245 \times 10^9/L$, serum creatinine $81 \mu\text{mol/L}$, and ESR 40mm in 1st hour. His chest X-ray, c-reactive protein level, urine full report, blood picture, serum potassium, serum calcium, and arterial blood gas analysis were normal. Liver enzymes, liver function tests, alkaline phosphatase, and gamma-glutamyl transferase were also normal. Fasting blood sugar, glycosylated haemoglobin (HbA1c), thyroid-stimulating hormone, and serum cortisol levels were normal. Schirmer's test was positive, as the strip was wet, longer than 5mm after 5 minutes. (Figure: 1)

Anti-nuclear antibodies (ANA) were positive (1/160) with a nucleolar pattern. Rh factor was positive with a titre of 237 IU/mL. Anti Ro/La antibodies were negative. He underwent salivary gland biopsy of lip and the histology showed lobules of salivary acini infiltrated by lymphoid cells. Serum protein electrophoresis and C3, C4 complement levels were normal.



Figure: 1: Schirmer's test of the patient

Hepatitis C antibodies were negative and screening for HIV was negative. Ultrasound scan of the abdomen and pelvis revealed no intra-abdominal lymphadenopathy. Due to the unavailability of facilities, IgG4 panel could not be performed.

A thorough evaluation to exclude mimics of SS, including Hepatitis C, HIV, and lymphoma was carried out before diagnosing pSS and the patient fulfilled the 2016 American College of Rheumatology/ European League Against Rheumatism (ACR/EULAR) criteria for diagnosing pSS(1). A multi-disciplinary management approach was offered to the patient with the involvement of a general physician, consultant rheumatologist, consultant ophthalmologist, consultant orthodontist, and a nutritionist.

He was started on oral prednisolone 1mg/kg daily with azathioprine 70mg daily dose. Artificial tears were prescribed. Intervention on gradually improving his dental hygiene was introduced. Three months later, as the eye symptoms showed poor resolution, artificial tear application was optimized and, insertion of punctual plugs or lacrimal duct ligation were discussed as options for the future.

Over 10 months follow up, his quality of life improved, allowing him to attend his routine family affairs and resume his job. Educating him about the disease and setting achievable targets with frequent reviews helped the patient.

Discussion

SS is a chronic autoimmune disorder causing mononuclear infiltration of the salivary and lacrimal glands leading to sicca syndrome where the typical manifestations include oral and ocular dryness. Although its primary involvement is in these glands, SS is a multi-systemic disease with well-recognized involvement of pulmonary, renal, gastrointestinal, and neurological systems(2).

This patient having sicca symptoms for months, failed to communicate his symptoms, rather sought medical attention for polyuria caused by excessive fluid intake to relieve the oral dryness symptoms. This led to an undue delay and efforts to investigate him for the cause of polyuria and polydipsia. Literature describes many instances when the diagnosis of SS was delayed due to symptoms originating from non-exocrine organ involvement(3,4).

The diagnosis of SS is clinical. The 2016 ACR/EULAR criteria(1) developed for research purposes encompassing clinical features, histological features of salivary glands and serological criteria, were fulfilled by this patient. The diagnosis of pSS was made after the exclusion of associated autoimmune disorders such as rheumatoid arthritis, systemic lupus erythematosus, and systemic sclerosis(5).

Primary Sjögren syndrome is an autoimmune disease with a female preponderance ranging from a ratio of one to between 10 and 20(6). Although males have a poor prognosis, other features such as negative anti -Ro/La autoantibodies, normal gamma-globulin levels and normal complement levels predict a good prognosis in this patient.

Research on SS is still in its infant stage in terms of therapeutics. There is no therapeutics available to reverse the pathology and the treatment is targeted at symptom relief. Although systemic therapy has a place in SS management, there is no available research evidence to guide systemic therapy. Hence the treatment is decided on ad hoc basis depending on the disease activity. Patients with 'clinical European league against rheumatism Sjögren syndrome disease activity index' (clinESSDAI) score ≥ 1 are defined as having active systemic disease, and organ damage. This patient had a clinESSDAI score of 5.

A systematic review and meta-analysis of cohort studies revealed that pSS is not associated with an increase in all-cause mortality as compared with the general population. However, a subset of patients with extra-glandular involvement, vasculitis, hypocomplementemia, and cryoglobulinemia may be at increased risk of mortality and require close follow-up(7).

Conclusion

Despite being a rare illness in men, pSS has to be considered when patients present with polyuria and nonspecific symptoms, as they may not reveal sicca symptoms unless probed.

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