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Case Report

Extra-glandular manifestations with Sjögren's Syndrome: Case Series in Bangladesh

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Abstract

Background: *Sjögren's syndrome (SS) is a chronic autoimmune disease that is mainly characterized by lymphocytic infiltration of the exocrine glands, leading to sicca symptoms. Extraglandular manifestations (EGMs) involving more than one organ system influence the prognosis. Though SS is gaining global recognition, information from Bangladesh remains limited. The present case series aims to contextualize the clinical spectrum of EGMs in SS patients within the setting of a Bangladeshi tertiary care.*

Methods: *We retrospectively analyzed patients with primary SS, based on the 2016 ACR/EULAR classification criteria, who were admitted to the rheumatology outpatient clinic of Shaheed Suhrawardy Medical College Hospital between July 2024 to December 2024. Clinical data were analyzed for demographic data, serologic features, and reported extra-glandular manifestations on renal, ocular, neurology, dental, endocrine and hematological systems.*

Results: *04 cases of Sjögren's syndrome were diagnosed with extra-glandular manifestations like non-eruption of teeth, central chorioretinopathy, thrombocytopenic purpura, renal tubular acidosis over 02 years. Sjögren's syndrome was diagnosed based on the 2016 ACR/EULAR classification criteria, including clinical features, Schirmer's test and autoantibodies. Biopsy was not performed.*

Conclusion: *SS extraglandular manifestations are still underdiagnosed in Bangladesh and can be one of the reasons for delay in diagnosis and less-than-ideal management. Our findings emphasize the need for systematic screening protocols and physician awareness of the multisystem nature of the disease. Prospective studies are necessary to ascertain the true prevalence and clinical impact of EGMs in the Bangladeshi population.*

Keywords: *Sjögren's syndrome, extraglandular manifestations, Bangladesh, autoimmune disease, case series.*

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Introduction

Sjögren's syndrome (SS), an autoimmune exocrinopathy of indeterminate cause, is considered one of the most common systemic inflammatory connective tissue disorders. The prevalence of primary Sjögren's syndrome (pSS) is reported from 0.04 to 0.66%[1,2], depending on the populations being studied and the classification criteria used. PSS most commonly affects females in the fourth and fifth decades of life, with a sex ratio of 9:1. The condition mainly affects the exocrine glands. It typically presents as chronic dryness of the eye and mouth, resulting in functional impairment of the salivary and lacrimal glands secondary to lymphocytic infiltration and consequent loss of glandular tissue. The development of extraglandular involvement and organ-specific autoimmune disease may complicate extraglandular organs.[3] As patients with pSS may involve many organs in addition to the exocrine glands, the clinical picture of pSS is divided into exocrine gland features and extraglandular manifestations (EGM),[4] the latter being experienced by a relatively considerable number of patients.[5-7]

We diagnosed 04 cases of Sjögren's syndrome with extra-glandular manifestations like non-eruption of teeth, central chorioretinopathy, thrombocytopenic purpura, renal tubular acidosis over 02 years.

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Case series

Particulars:

Features	Case 1	Case 2	Case 3	Case 4
Name	Shawpon Miah	Mrs.kohinoor	Mrs. Khadiza	Mrs.Chumki
Age	28 years	32 years	55 years	28 years
Sex	Male	Female	Female	Female
Occupation	Garment worker	Homemaker	Homemaker	Homemaker

Presenting complaints:

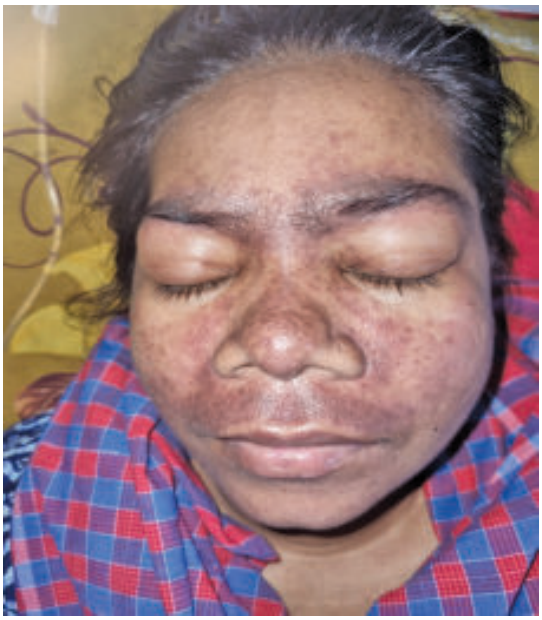
Case 1	Case 2	Case 3	Case 4
• Incomplete eruption of teeth since childhood • Fever for 02 months • Symmetrical Polyarthritis for 02 months • Alopecia, oral ulcer, Photosensitivity • Dry eyes, dry mouth, dysphagia	• Recurrent weakness of both upper and lower limbs over 01 year • Blurring of vision for 03 months • Inflammatory polyarthritis for 01 year, malar rash, alopecia, oral ulcer, weight loss	• Epistaxis and gum bleeding for 07 days • Multiple ecchymoses and purpura all over the body for 01 month • Dry eyes, dry mouth for 02 months • Known case of IHD, hypothyroidism	• Recurrent neurological weakness of all limbs over 02 years • Inflammatory polyarthritis • Photosensitive rashes, oral ulceration, Chronic hepatitis B

Examination findings:

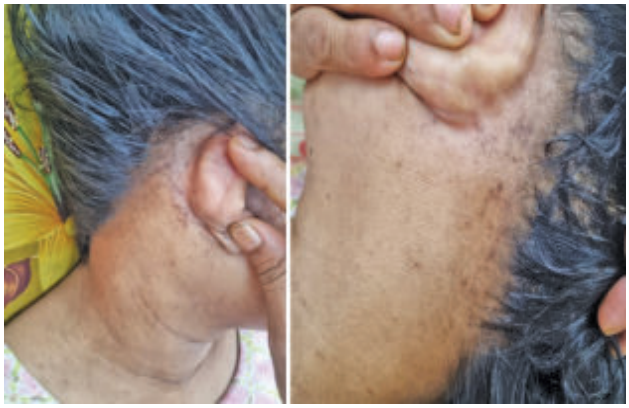
System	Case 1	Case 2	Case 3	Case 4
General	• Mildly anemic • Frontal hair loss • Bed side urine: trace protein	• Multiple erythematous maculopapular rashes over malar area, nose, upper lip, eyebrows • Moderate anemia • Alopecia	• Mildly anemic • Non-pitting edema • Multiple purpuric rashes all over the body	Nothing abnormalities detected
Nervous system	• Nothing abnormalities detected	• Muscle power: 4/5 (all limbs) • Antalgic gait	Nothing abnormalities detected	• Muscle power: 0/5(all limbs) • All the deep tendon reflexes were absent
Locomotor system	• Swelling and Grade 1 tenderness in PIP and DIP joints of both hands	• Swelling of PIP joints • Grade 2 tenderness • Antalgic gait	Nothing abnormalities detected	Nothing abnormalities detected
Alimentary system	• White plaque in the tongue • Multiple oral ulcer • Multiple missing teeth, Non-eruption of 2nd incisor teeth in lower jaw and partial eruption of upper left canine teeth. Caries on lower right 1st and 2nd molar and lower left 2nd molar teeth.	• Swelling of PIP joints • Grade 2 tenderness • Antalgic gait	Nothing abnormalities detected	Nothing abnormalities detected
Schirmer's test	Normal in both eyes (35mm)	Positive in both eyes (3mm in right eye, 2mm in left eye)	Positive in both eyes (3mm)	Normal in both eyes (15mm)

Case -1

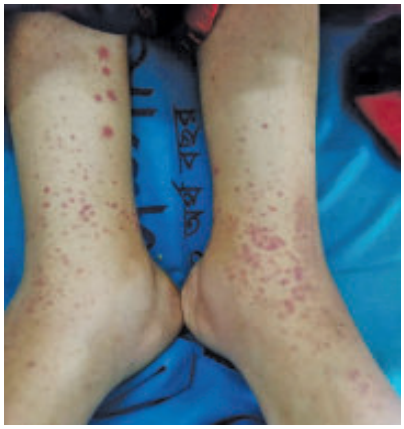
Case -2



Case -2



Case -3



Investigations

Case 1	Case 2	Case 3	Case 4
• OPG: missing tooth, periodontitis • ANA, Anti ds-DNA, Anti CCP, Anti SS-A/ Ro 52KD : positive	• Color fundus photography: central serous chorioretinopathy • ANA, Anti ds-DNA, Anti CCP, Anti SS-A : positive	• Platelet count: 5000/ cmm • Anti-TPO ab positive • ANA, SS-A/Ro60KD, SS-A/Ro52KD, SS-B/ La: positive	• Serum electrolytes: hypokalemia, hyperchloremia, metabolic acidosis • Urinary potassium excretion was high • ANA, anti SS-A positive

Final diagnosis

Case 1
SLE with RA overlap (Rhupus) with Primary Sjögren's syndrome [Rhupus: ACR/EULAR classification criteria met].

Case 2
SLE with RA overlap Syndrome (Rhupus) with secondary Sjögren's Syndrome with Central serous Chorioretinopathy (B/E)

Case 3
Thrombocytopenic Purpura (autoimmune) and Sjögren's Syndrome and Hashimoto's Thyroiditis,

Case 4
Primary Sjögren's Syndrome with Distal Renal Tubular Acidosis (Type 1) with Chronic Hepatitis B

Discussion:

Renal involvement, particularly distal renal tubular acidosis (dRTA), is a well-documented but under-recognized manifestation of SS, particularly in Asian populations, where it may be the presenting feature in young females with a prevalence of 5-10% in primary SS[8,9].

In patients with Primary Sjögren's Syndrome (pSS), the prevalence of autoimmune thrombocytopenia, specifically Immune Thrombocytopenic Purpura (ITP), is estimated to be around 12%. Some studies indicate that secondary ITP is common in pSS patients and may have distinct characteristics compared to pSS patients without ITP. While thrombocytopenia is not as common as other cytopenias, it can be a significant finding in pSS, and newly developed cytopenia can be a sign of lymphoma[10].

Rhupus, an overlap syndrome combining features of Rheumatoid Arthritis (RA) and Systemic Lupus Erythematosus (SLE), is rare in patients with Sjögren's Syndrome (SS). While estimates vary, rhupus is thought to affect a small percentage of individuals with rheumatic diseases, with some studies suggesting 0.5% to 2% of patients with rheumatic diseases may be affected.[11]

While central serous chorioretinopathy (CSCR) is not a common complication of Sjögren's syndrome, some studies suggest a possible association. It's essential to note that the prevalence of CSCR in Sjögren's syndrome is not well-defined, and ongoing research aims to understand this relationship better.[12]

Conclusion:

This case series illustrates that extraglandular manifestations of primary Sjögren's syndrome are diverse and potentially underrecognized. Clinicians in Bangladesh should consider routine EGM screening in suspected SS, and prospective multicenter studies are needed to establish the true prevalence and guide management in our population.

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