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(CASE REPORT)

Overlapping autoimmune diseases: Systemic lupus erythematosus with secondary Sjögren's syndrome

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Abstract

Systemic lupus erythematosus (SLE) and Sjogren's syndrome (SS) are two important autoimmune conditions. SLE is characterized by increased production of autoantibodies and the development of immune complexes, SLE is a highly diverse condition that can affect multiple organ systems. Its disease activity ranges from consistently mild to potentially life-threatening. Sjogren's syndrome arises due to the buildup of lymphocytes in the exocrine glands. The most common clinical manifestations are dry mouth (xerostomia) and dry eyes (xerophthalmia), caused by the involvement of the salivary and lacrimal glands. This paper discusses the case of a 45-year-old female diagnosed with SLE along with secondary Sjogren's syndrome & nephrotic range proteinuria, and additional comorbidities including diabetes, hypertension, and pulmonary tuberculosis. Presenting a detailed case can help healthcare professionals improve recognition of sSS in SLE patients, ensuring timely diagnosis and comprehensive management.

Keywords: Systemic lupus erythematosus; Sjogren's syndrome; Autoimmune disorder; Xerostomia; Xerophthalmia

1. Introduction

Systemic lupus erythematosus (SLE) is a chronic autoimmune disease marked by inflammation in multiple systems due to immune system dysregulation. The presentation of SLE can vary greatly, from mild skin issues to severe organ damage that can be life-threatening.[1] The global incidence of systemic lupus erythematosus (SLE) varies from 1.5 to 11 cases per 100,000 person-years, while its prevalence ranges from 13 to 7,713.5 cases per 100,000 individuals. Research consistently indicates that women are more commonly affected by this condition than men worldwide.[2] The exact cause of systemic lupus erythematosus (SLE) is unknown, but it involves genetic, environmental, hormonal, and immunological factors. Genetic links include HLA-DR2, HLA-DR3, and complement deficiencies (C1q, C2, C4). Environmental triggers such as UV radiation, Epstein-Barr virus, certain medications (e.g., hydralazine, procainamide), smoking, and toxins can contribute. Estrogen enhances B-cell activation, explaining the female predominance. Immune system dysregulation, particularly involving B cells and T cells, leads to the production of autoantibodies like anti-dsDNA and anti-Sm, further driving SLE pathogenesis. [1] The symptoms vary widely, affecting multiple organ systems. Constitutional symptoms such as fatigue, fever, and weight loss are common. Musculoskeletal involvement may present as arthritis and muscle pain. Dermatological signs include malar rash, discoid lesions, and photosensitivity. Renal complications like lupus nephritis can manifest as proteinuria and hematuria. Cardiopulmonary issues such as pleuritis and pericarditis are also seen. Additionally, neurological manifestations may include seizures, psychosis, and peripheral neuropathy, reflecting the systemic nature of the disease.[1] SLE diagnosis involves clinical assessment, ANA, anti-dsDNA, and anti-Sm testing, decreased C3/C4 levels, and renal biopsy for lupus nephritis.[1] The treatment primarily aims to control immune disturbances through the use of anti-inflammatory and immunosuppressive medications. These treatments include antimalarials, glucocorticoids (GCs), non-steroidal immunosuppressants, and targeted therapies. Targeted therapies specifically focus on reducing B-cell survival and activation. They achieve this either by directly

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depleting B-cells or inhibiting their function. This approach helps to reduce disease activity and prevent organ damage.[3]

Sjogren's syndrome (SS) is a chronic, systemic autoimmune disorder primarily characterized by lymphocytic infiltration and progressive destruction of exocrine glands, predominantly the salivary and lacrimal glands. This glandular dysfunction leads to classic sicca symptoms such as xerostomia (dry mouth) and xerophthalmia (dry eyes).

Sjogren's syndrome is classified into two types:

- Primary Sjogren's syndrome (pSS): Occurs independently without any associated autoimmune disease.
- Secondary Sjogren's syndrome (sSS): Develops in conjunction with other autoimmune conditions, such as systemic lupus erythematosus (SLE), rheumatoid arthritis (RA), or systemic sclerosis.

The exact etiology of Sjogren's syndrome remains unclear, but it is believed to arise from a combination of genetic, environmental, and immunological factors. Pathogenesis involves immune-mediated destruction of glandular tissue, driven by activated B cells, T cells, and pro-inflammatory cytokines. In addition to sicca symptoms, patients may experience fatigue, arthralgia, parotid gland enlargement, and systemic involvement affecting organs such as the lungs, kidneys, and nervous system. Notably, patients with SS have a higher risk of developing non-Hodgkin's lymphoma. Diagnosis is based on clinical presentation, autoantibody markers (e.g., anti-Ro/SSA, anti-La/SSB), glandular function tests (e.g., Schirmer's test), and labial salivary gland biopsy. Early diagnosis is crucial to prevent complications and improve quality of life. For dry eyes, treatment includes regular use of preservative-free artificial tears during the day and preservative-free gels or ointments at night. In severe cases, punctal occlusion with plugs or cauterization may be necessary; however, cauterization should follow a successful trial of plugs to prevent excessive tearing. Anti-inflammatory eye drops like cyclosporine (Restasis) can help boost tear production. Additionally, serum tears made from the patient's blood are beneficial and available at many eye centers. Patients may find relief through increased water intake, chewing gum, or saliva substitutes for dry mouth. Prescription medications such as pilocarpine (Salagen) or cevimeline (Evoxac) can stimulate saliva production. If yeast infections develop, antifungal treatments are recommended for nasal dryness, using humidifiers and nasal saline irrigation can help. Managing acid reflux with proton-pump inhibitors or H2 blockers may reduce associated symptoms. In cases of systemic organ involvement, corticosteroids are the first-line treatment. DMARDs like methotrexate and azathioprine are commonly used as steroid-sparing agents, although none are specifically approved for Sjogren's syndrome. Hydroxychloroquine is advised for inflammatory polyarthritis. For severe extra-glandular manifestations such as vasculitis, B cell depletion therapy may be beneficial. Conversely, TNF-alpha inhibitors have not shown effectiveness in treating Sjogren's syndrome.[4]

Systematic reviews and meta-analyses indicate that the prevalence of secondary Sjögren's syndrome (sSS) among patients with systemic lupus erythematosus (SLE) ranges between 14% and 17.8%. The pathophysiology linking SLE and secondary Sjögren's syndrome (sSS) involves shared immune dysregulation. B-cell hyperactivity drives autoantibody production, with anti-dsDNA in SLE and anti-Ro/SSA and anti-La/SSB in sSS. Dysfunctional CD4+ T cells release inflammatory cytokines like IL-6, IL-17, and IFN-γ, promoting chronic inflammation. Elevated type I interferons amplify B-cell activation and autoantibody production. Apoptotic cell debris exposes autoantigens, triggering immune responses. In sSS, lymphocytic infiltration damages exocrine glands, causing dryness. Genetic predisposition and environmental triggers further contribute to disease progression. These interconnected mechanisms result in overlapping systemic and glandular manifestations in SLE with secondary sSS. Presenting a detailed case can help healthcare professionals improve recognition of sSS in SLE patients, ensuring timely diagnosis and comprehensive management.[5] Highlighting key diagnostic markers (e.g., ANA, anti-dsDNA, anti-Ro/SSA, anti-La/SSB) and investigative approaches will provide insights for clinicians.

2. Case presentation

A 45 yrs old female patient was admitted to the hospital with a complaint of generalized body pains for one month, unable to walk for one month, not taking feeds for 1 month, altered sensorium in the form of irrelevant talk since 1 week & slurring of speech, fever for 6-8 months, low grade, increased during evening time, hair fall and weight loss associated with loss of appetite since 6 months. No complaints of cough & swelling of limbs (h/o + - 2months back).

- **Past medical history:** k/c/o DM since 6 yrs (on medication –T. Metformin 500mg & T. Glimiperide1mg), HTN since 6yrs (on medication T. Met XL 50mg) and k/c/o inflammatory poly arthropathy for 8 months, PTB on ATT (3 tabs/day) since1 month.
- **Personal history:** Takes mixed diet, non-alcoholic, non-smoker, Bowel& bladder- regular, decreased Sleep & appetite.

- **On Examination:** The patient was conscious, coherent, cooperative, and poorly built. The pulse rate was 82bpm, blood pressure was 110/70mmHg, spo2 was 97%, and respiratory system (RS) examination BAE+ clear indicated regular bilateral air entry with no abnormal breath sounds. Cardiovascular examination: Both heart sounds were heard with no murmur. There was no evidence of pallor, cyanosis, icterus, clubbing, or lymphadenopathy. In Neurological examination -No neurological deficits, Per-abdomen examination: Abdomen is soft and non-tender. Non-scaring alopecia is noted.
- **Laboratory findings:** Urine examination revealed the presence of proteins, glucose, ketones, bilirubin, and blood in the urine, which could be a UTI. A CECT abdomen and USG abdomen revealed grade 1 hepatic steatosis with mildly complex ascites with septations noted in the pelvis. UGIE findings are indicative of H. pylori-induced erosive duodenitis.

Routine blood investigations were as follows, Hemoglobin was 13 mg/dl at admission, MCV 83, Platelet count of 195000, and WBC of 7000. Sodium levels of 132meq/L Potassium – 5.7meq/L, chlorine levels: 1047meq/L, Creatinine of 0.75 and urea of 78. Total bilirubin was 1.1 mg/dl of which Direct was 0.51 mg/dl and Indirect was 0.59 mg/dl. Total protein was 6.8 with Albumin of 2.0 and globulin of 5. Urice acid levels were 11.77, Sr. Calcium: 6.2. HIV, HBsAg, and HCV was negative.

dsDNA ++, nucleosomes +, SS-A & SS-B were positive, histones +, Schirmer's test was positive for dry eyes (3 mm/5 minutes), C3 (44mg/dl) & C4(7mg/dl) levels are decreased, ESR levels(100mm/hr.) are significantly higher than normal. 24 hr. urine analysis- loss of 3gm of protein. These findings are suggestive of SLE with secondary Sjogren's syndrome with nephrotic range proteinuria.

Treatment: Initially, the patient was treated with Tab. Prednisolone 10mg, Tab. Hydroxychloroquine, ATT 3Tab/day, Tab. Pyridoxine 40 mg, Tab. Met-XL, Injection HAI according to GRBS, Injection pantoprazole 40mg, Tab. Pilocarpine 5mg.

3. Discussion

This case report describes a 45-year-old female diagnosed with Systemic Lupus Erythematosus (SLE) with secondary Sjögren's syndrome (sSS), presenting with a complex array of systemic symptoms. The patient's history of diabetes mellitus, hypertension, inflammatory polyarthropathy, and pulmonary tuberculosis (PTB) compounded her clinical presentation. Key symptoms such as generalized body pain, altered sensorium, hair loss, and weight loss indicated underlying systemic inflammation. Diagnostic findings, including positive dsDNA, nucleosomes, SS-A/SS-B, and histones, along with a positive Schirmer test, confirmed the presence of SLE with sSS. The significantly elevated ESR, reduced C3/C4 levels, and nephrotic range proteinuria indicated active disease with renal involvement. The presence of H. pylori-induced erosive duodenitis and grade 1 hepatic steatosis further complicated the case. Management focused on immunosuppression with prednisolone and hydroxychloroquine to control autoimmunity, while pilocarpine addressed sicca symptoms. Concurrent treatment for PTB, electrolyte management, and gastric protection was implemented to ensure comprehensive care. This case emphasizes the need for careful assessment and multidisciplinary management in patients with overlapping autoimmune conditions to prevent organ damage and improve clinical outcomes.

4. Conclusion

This case underscores the diagnostic complexity and clinical challenges associated with SLE with secondary Sjögren's syndrome, especially in the presence of multiple comorbidities such as diabetes mellitus, hypertension, and pulmonary tuberculosis. The patient's symptoms, including altered sensorium, systemic involvement, and nephrotic-range proteinuria, indicated severe disease progression. Positive serological markers (dsDNA, SS-A, SS-B), reduced complement levels, and sicca symptoms confirmed the diagnosis. Prompt initiation of immunosuppressive therapy alongside supportive management resulted in clinical improvement. Timely identification of SLE with secondary Sjögren's syndrome is crucial to prevent irreversible organ damage. Regular assessment of disease activity, renal function, and infection risk is vital for long-term management and relapse prevention.

Compliance with ethical standards

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Disclosure of conflict of interest

No conflict of interest to be disclosed.

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