doi:10.17659/01.2024.0018

Journal of Case Reports 2024;14(3):65-67

Severe Thrombocytopenia: An Uncommon Presentation of Primary Sjögren's Syndrome

Dawlat Sany[1], Wadah Ahmed[2], Heba Mohamed Abdelfattah[3]

[1]Consultant Internal Medicine and Nephrology, Kalba Hospital, Sharjah, EHS; [2]Specialist Internal Medicine, Kalba Hospital, Sharjah, EHS; [3]Consultant Clinical Pathologist, Pure Health Group Company, Sharjah, UAE.

Corresponding Author:
Dr Dawlat Sany
Email: dawlat.hussein@ehs.gov.ae

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Received : March 21, 2024
Accepted : June 4, 2024
Published : August 5, 2024

Abstract

Background: Primary Sjögren's syndrome (pSS) is a chronic autoimmune disorder typically presenting with dry eyes and mouth. Hematological abnormalities are common, but severe thrombocytopenia is rare and poses diagnostic challenges. *Case Report*: A 50-year-old female with chronic eye dryness post-LASIK presented with acute petechial rash, ecchymosis, minimal epistaxis, and persistent menorrhagia. She also reported occasional dysphagia, chronic heartburn, and pain in her neck, back, and knees. Examination showed no significant findings apart from peticheal rash. Laboratory tests revealed microcytic, hypochromic anemia and severe thrombocytopenia (3,000/mm³). Immunological tests were positive for ANA, ENAs, anti-SSA, and anti-SSB, suggesting pSS. A Schirmer test and conjunctival fluorescein staining supported this. Bone marrow biopsy and flow cytometry ruled out other hematological disorders. Treatment with methylprednisolone and IVIg, followed by oral prednisolone, improved hemoglobin and platelet counts. At discharge, platelet count was 160,000/mm³, maintaining around 200,000/mm³ after three months. *Conclusion*: Severe thrombocytopenia can be an initial manifestation of pSS, even without typical exocrine gland symptoms. Recognizing this can lead to earlier diagnosis and treatment, improving outcomes in pSS-related thrombocytopenia.

Keywords: Autoimmune Disorders, Bone Marrow, Methylprednisolone, Pain, Thrombocytopenia.

Introduction

Primary Sjögren's syndrome (pSS) is a chronic autoimmune disorder primarily characterized by lymphocytic infiltration of exocrine glands, leading to symptoms such as dry eyes and dry mouth. Although the classic presentation involves sicca symptoms, pSS can manifest with a wide range of systemic and extra-glandular features [1,2]. Hematological abnormalities are relatively common in pSS, but severe thrombocytopenia is an uncommon and atypical presentation. Thrombocytopenia in pSS can complicate the clinical course and pose significant diagnostic and therapeutic challenges. Its presence may indicate an underlying hematological or autoimmune disorder, necessitating a comprehensive evaluation to exclude other potential causes such as malignancies, bone marrow disorders, or other autoimmune conditions.

We hereby report a 50-year-old female patient who presented with severe thrombocytopenia, diffuse petechial rash, and minimal epistaxis. Further investigation led to a diagnosis of primary Sjögren's syndrome, highlighting the importance of considering pSS in patients with atypical hematological presentations.

Case Report

A 50-year-old female patient, with a history of chronic eye dryness since undergoing LASIK

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surgery 11 years ago, was admitted with an acute onset of a diffuse petechial rash, ecchymosis, minimal epistaxis, and persistent menorrhagia over the past two years. She also reported occasional dysphagia to some solids, chronic intermittent heartburn, and pain in her neck, upper back, and knees over the past few months. The patient did not report weight loss, fever, peripheral joint swelling, or significant oral dryness. On examination, there were no signs of clinical synovitis, dactylitis, parotid swelling, annular erythema, erythema multiforme, erythema nodosum, lupus-specific rashes, vasculitis rashes, psoriasis, sclerodactyly, Raynaud's phenomenon, or major organ involvement.

Laboratory tests revealed microcytic, hypochromic anemia and severe thrombocytopenia (3,000/mm³). Immunological profile tests showed positive antinuclear antibodies (ANA), antibodies to extractable nuclear antigens (ENAs), anti-SSA, anti-SSB, and rheumatoid factor. These results suggested a diagnosis of Sjögren's syndrome with an atypical hematological presentation. A Schirmer test was positive, and mid-conjunctival fluorescein staining was observed in both eyes. A bone marrow biopsy showed mildly hypocellular marrow with mature trilineage hematopoiesis, with no diagnostic morphology of lymphoproliferative, myelodysplastic/myeloproliferative neoplasms, or plasma cell neoplasms. Flow cytometry revealed no definitive involvement by acute leukemia or significant increases in blast cells.

The diagnosis of primary Sjögren's syndrome with severe thrombocytopenia was confirmed. The patient was treated with methylprednisolone 500 mg/day for three days and intravenous immunoglobulin 0.4 g/kg/day for five days, followed by oral prednisolone 60 mg/day. This treatment led to a progressive improvement in bicytopenia, with a gradual rise in hemoglobin to 9.3 g/dL and an increase in platelets to 160,000/mm³ at discharge. The patient was discharged on oral prednisolone 60 mg/day for two weeks, with a gradual taper over six weeks. At a follow-up three months post-discharge, the platelet count was maintained at approximately 200,000/mm³.

Discussion

Hematologic abnormalities are uncommon in primary Sjögren's syndrome (pSS) patients; however, approximately one-third of pSS patients experience cytopenias such as leukopenia, anemia, and immune thrombocytopenia (ITP) because its degree is usually mild and asymptomatic, often disregarded in daily [3]. Anemia is more commonly observed in pSS, occurring in 10-19% of patients [4]. While leukopenia and anemia are the most prevalent hematological complications in pSS, cases involving thrombocytopenia are relatively rare [5]. Severe ITP increases the risk of hemorrhagic events and can be fatal. The pathogenesis of ITP in pSS involves auto-antibodies leading to peripheral platelet destruction, splenic sequestration, and reduced platelet production due to autoantibodies targeting megakaryocytes [6]. Some patients may be missed because they lack the typical dryness in their eyes and mouths [7].

In 2015, the European League Against Rheumatism (EULAR) promoted an international collaborative study group (EULAR-SS Task Force) aimed at developing consensual recommendations for a homogeneous approach to pSS patients with systemic involvement. According to these guidelines, patients with thrombocytopenia associated with positive Ro/SS-A and/or La/SS-B auto-antibodies should be considered for a possible diagnosis of SS [8]. First-line management for SS-related ITP includes corticosteroids and immunoglobulin [9]. However, long-term remission is rare, with relapse rates as high as 70-80% [10]. Recurrent patients may require second-line treatments such as cyclosporin A, azathioprine, and rituximab, and may ultimately need a splenectomy [11].

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Conclusion

Thrombocytopenia may present at the onset of pSS development without any involvement of the exocrine glands. Increased awareness of pSS-associated thrombocytopenia can lead to earlier and more accurate diagnosis and treatment, especially in severe cases with hemorrhagic manifestations.

Contributors: DS: manuscript writing, patient management; WA, HMA: manuscript editing, patient management. DS will act as a study guarantor. All authors approved the final version of this manuscript and are responsible for all aspects of this study.
Funding: None; *Competing interests*: None stated.

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