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RESEARCH ARTICLE

Falling Platelets, Raising Suspicion and a Surprising Diagnosis: A Rare Presentation of Sjögren's Syndrome with Bicytopenia and Bleeding Gums

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Abstract: Sjögren's syndrome is a chronic autoimmune condition that primarily affects exocrine glands, leading to sicca symptoms. Although systemic manifestations are recognized, severe hematological involvement with bleeding is rare. We describe a 52-year-old woman who presented with bleeding gums for one month and melena for 10 days. She also reported fatigue and leg pain. Clinical evaluation revealed pallor without hepatosplenomegaly or lymphadenopathy. Laboratory investigations showed severe anemia (hemoglobin 4.3 g/dL) and thrombocytopenia (9,000/µL). Peripheral smear revealed microcytic hypochromic anemia, and bone marrow aspiration was a dry tap. ANA testing revealed anti-Ro/SSA positivity, and Schirmer's test confirmed severe dry eyes. Minor salivary gland biopsy demonstrated fibro-fatty infiltration with periductal lymphocytes. A diagnosis of Sjögren's syndrome with bicytopenia was made. She was managed with blood and platelet transfusions, corticosteroids, azathioprine, insulin, thyroxine, atorvastatin, and lubricating eye drops, with significant improvement at follow-up. This case highlights a rare presentation of Sjögren's syndrome with bicytopenia and mucosal bleeding. Autoimmune etiologies should be considered in unexplained cytopenias. Early recognition and prompt immunosuppression can improve outcomes.

Keyword: Sjögren's syndrome,Bicytopenia,Thrombocytopenia,Anemia,Bleeding gums,Autoimmune cytopenia,Bone marrow biopsy,Megakaryocyte hyperplasia,Anti-Ro/SSA antibodies,* Immunosuppressive therapy

Introduction

Sjögren's syndrome is an autoimmune disease in which immune cells target salivary and lacrimal glands, producing dryness of the eyes and mouth.[1] It can occur as primary disease or in association with systemic autoimmune disorders such as systemic lupus erythematosus or rheumatoid arthritis, termed secondary Sjögren's syndrome.[2]

Systemic features are common and may involve the musculoskeletal, renal, pulmonary, vascular, and nervous systems.[3] Hematological abnormalities are also reported, including anemia, leukopenia, and thrombocytopenia.[4] Severe cytopenias with bleeding manifestations are distinctly rare.[5]

We present an unusual case of Sjögren's syndrome with bicytopenia and bleeding gums as the initial manifestation, emphasizing the diagnostic challenge and therapeutic implications.

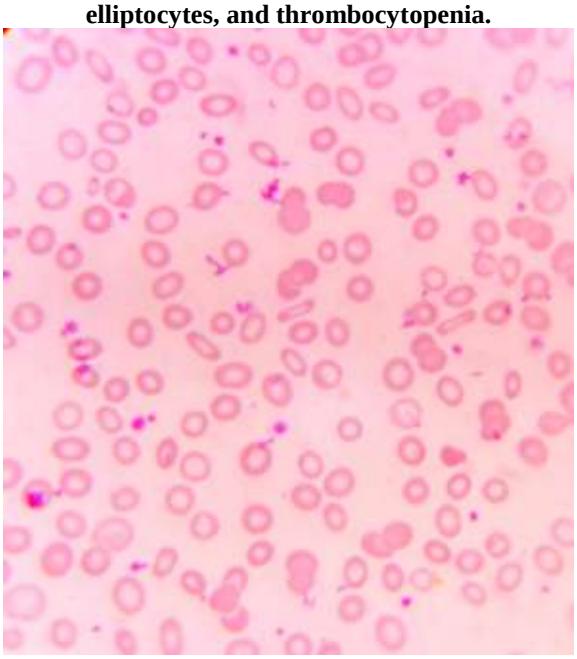
Case Presentation

A 52-year-old woman presented with spontaneous gum bleeding for one month, worsening over the preceding three days with clot formation. She also reported fatigue, bilateral leg pain, and black stools suggestive of melena. She denied fever, chest pain, syncope, or bleeding from other sites.

Her past history included type 2 diabetes mellitus (controlled with metformin, glimepiride, and

teneligliptin), hypothyroidism on thyroxine replacement, and dyslipidemia on atorvastatin. She was postmenopausal, vegetarian, and had no history of smoking or alcohol use. On examination, she appeared pale but was hemodynamically stable (pulse 99/min, BP 110/70 mmHg, RR 18/min, SpO_2 100% on room air). No lymphadenopathy, edema, or organomegaly was detected. Systemic examination was unremarkable except for a paraumbilical swelling with cough impulse. Laboratory tests revealed hemoglobin 4.3 g/dL (ref: 12–16 g/dL), platelets 9,000/µL (ref: 150,000–450,000/µL), total leukocyte count 9,500/µL (ref: 4,000–11,000/µL), PCV 14% (ref: 36–46%), and MCV 79 fL (ref: 80–96 fL). Peripheral smear demonstrated microcytic hypochromic anemia with anisopoikilocytosis, pencil cells, and elliptocytes, along with thrombocytopenia (figure 1). Bone marrow aspiration yielded a dry tap. Bone marrow biopsy done showed megakaryocyte hyperplasia (figure 2) and reticulin staining showed grade 1 fibrosis (figure 3). ANA testing was positive with anti-Ro/SSA antibody positivity. Schirmer's test confirmed severe dry eyes. Ultrasound showed no salivary gland enlargement. Minor salivary gland biopsy revealed fibro-fatty infiltration with periductal lymphocytes. The trend of hemoglobin and platelet recovery is shown in Figure 4. A diagnosis of Sjögren's syndrome with bicytopenia was made, with comorbid diabetes, hypothyroidism, and dyslipidemia. She was treated with red blood cell and platelet transfusions, high-dose oral prednisolone (60 mg daily), azathioprine, insulin, thyroxine, atorvastatin, and lubricating eye drops. She improved clinically and remained stable at two-month follow-up.

Figure1.Peripheral smear showing microcytic hypochromic anemia with anisopoikilocytosis, pencil cells, elliptocytes, and thrombocytopenia.



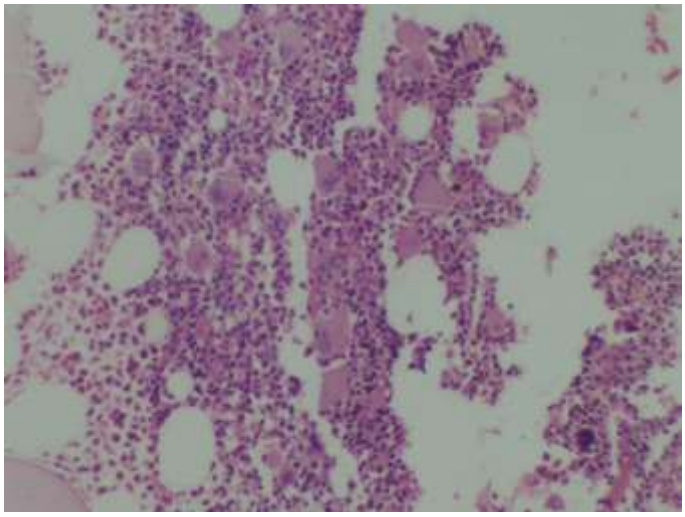


Figure 2: Bone marrow biopsy showing megakaryocyte hyperplasia

Figure 3: Bone marrow reticulin stain showing grade 1 fibrosis

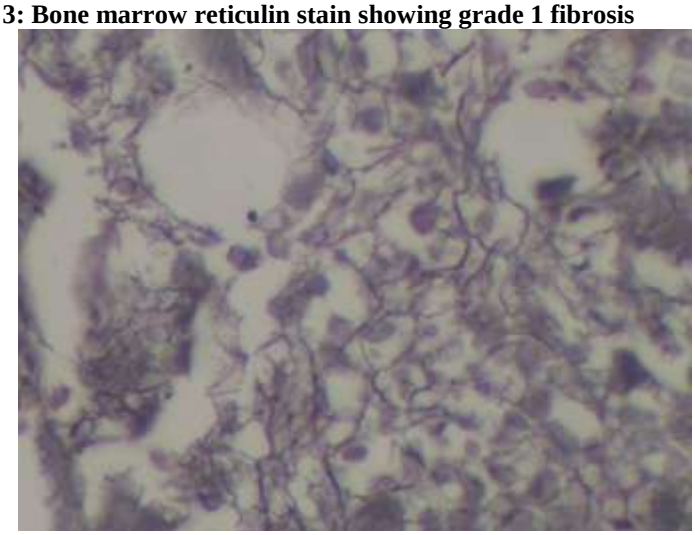
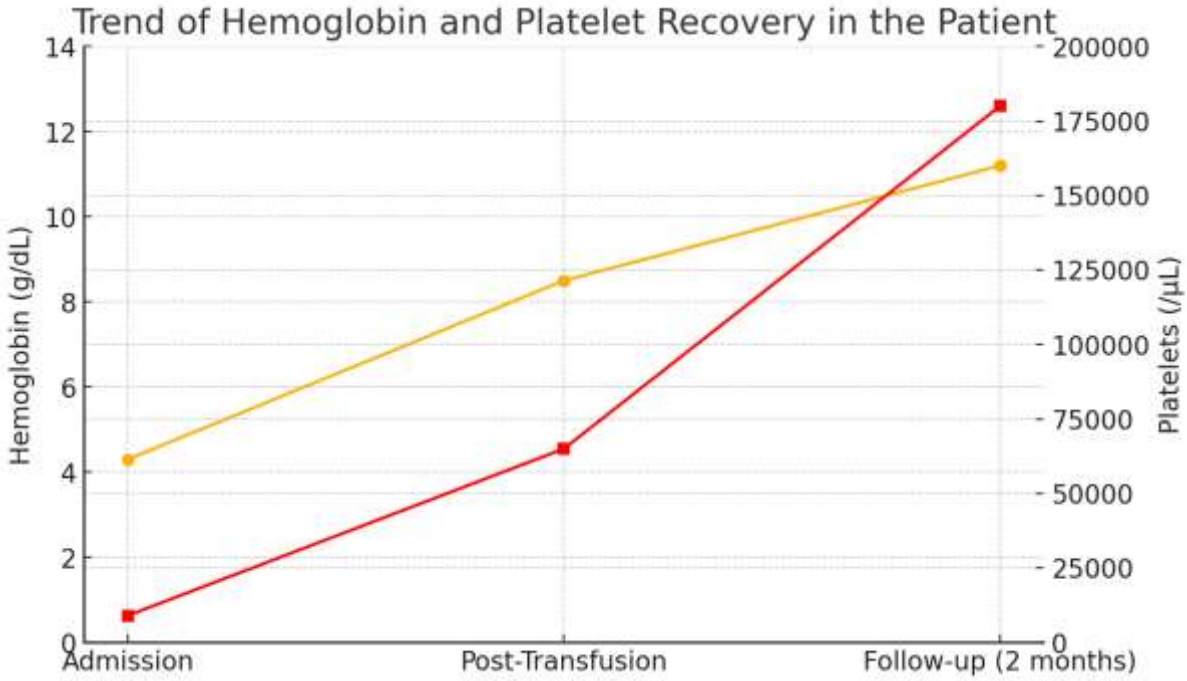


Figure 4. Trend of hemoglobin and platelet recovery in the patient with Sjögren's syndrome and bicytopenia. The patient presented with severe anemia (hemoglobin 4.3 g/dL) and thrombocytopenia (9,000/µL) at admission. Following transfusions and initiation of corticosteroids with azathioprine, hemoglobin and platelet counts improved significantly at post-transfusion assessment and normalized at two-month follow-up.



Discussion

Although dryness of the eyes and mouth is the hallmark of Sjögren's syndrome, the disease often extends beyond exocrine gland involvement, producing systemic manifestations.[1,3] Hematological abnormalities are reported in up to one-third of cases, most often presenting as anemia, leukopenia, or mild thrombocytopenia.[4,6] Severe cytopenias with bleeding are distinctly uncommon and reported mainly in isolated case reports.[8]

The mechanisms of cytopenias are multifactorial, including immune-mediated bone marrow suppression, peripheral destruction of circulating cells, and systemic inflammatory effects.[7] In rare cases, severe thrombocytopenia requires aggressive immunosuppression, as documented in prior case reports.[8]

Treatment depends on severity. Acute cases with life-threatening cytopenias require transfusion support. Corticosteroids remain the first-line therapy, while immunosuppressive agents such as azathioprine,

mycophenolate, or rituximab are used in refractory disease.[9,10]

Our patient responded well to corticosteroids and azathioprine, underscoring the importance of prompt recognition and early intervention.

Conclusion

This case describes an unusual initial presentation of Sjögren's syndrome with bicytopenia and mucosal bleeding. Autoimmune etiologies should be considered when evaluating unexplained cytopenias. Multidisciplinary care and timely immunosuppressive treatment can prevent complications and improve patient outcomes.

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Declarations

Conflicts of Interest: The authors declare no conflicts of interest.

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Patient Consent: Written informed consent was obtained from the patient for publication of this case report.

Ethics Statement: Ethical approval was not required for this single-patient case report as per institutional policy.