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Epistaxis as The Initial Presentation of Primary Sjögren Syndrome: A Case Report

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Abstract

Background: Primary Sjögren syndrome is chronic systemic autoimmune disorder characterized by lymphocytic infiltration of the exocrine glands, most commonly presenting with dryness of the mouth and eyes. This case is very rare, which the antibody in the Sjogren's syndrome case concerns platelets. **Objective:** This case report emphasizes the occurrence of epistaxis in Sjögren Syndrome. **Case Presentation:** This case describes a 30-year-old male presenting with recurrent epistaxis as the initial clinical manifestation. Further clinical evaluation revealed hypertensive crisis and dryness in the mouth and nasal mucosa. The clinical presentation raised suspicion of an underlying autoimmune condition, prompting further testing, which confirmed the presence of anti - Sjögren's syndrome type B (SSB) antibodies on an antinuclear antibody (ANA) profile. **Result:** The criteria for Sjogren's syndrome are based on the criteria if eye and mouth symptoms, eye and mouth clinical signs, and one of autoantibodies are found Anti-Ro (SSA), Anti-La (SSB), Antinuclear antibodies (ANA), Rheumatoid factor (RF). From this case has found symptoms of mouth and antibodies SSB. **Conclusion:** This case highlights that epistaxis can be caused by autoimmune cases, for epistaxis it is better to do screening for autoimmune examination test, emphasizing the need for further research to elucidate the diverse clinical manifestation, progression and prognosis factor.

Keywords: Autoimmune, Epistaxis, Sjögren Syndrome

Case Report

INTRODUCTION

Epistaxis or nasal bleeding is among the most frequently encountered event in emergency department. It can manifest spontaneously across all age groups, with the highest incidence observed in individuals

under 10 years old and those aged 70 to 79 (Senthilkumar & Jeba, 2022; Yang et al., 2024). Prevalence epistaxis in Sjögren's syndrome it's about 13.9% of SS patients with mild to severe thrombocytopenia experienced epistaxis (Wu et al.,2022). Therefore, a thorough evaluation is crucial to identify any underlying etiology. Although epistaxis often presents without an identifiable cause, it may also be triggered by localized trauma or systemic conditions, including autoimmune disorders such as Sjögren's syndrome. (McDonald et al., 2015; Parajuli, 2015).

Sjögren's syndrome is a systemic autoimmune disorder characterized by lymphocytic infiltration of exocrine glands, commonly presenting with dryness of the eyes and mouth. This condition exhibits a marked predilection for women, with a female-to-male ratio of approximately 9:1, and an incidence rate of 6.92 per 100,000 individuals (Carsons & Patel, 2023a; Zhan et al., 2023). We report a rare case of recurrent epistaxis in a 30-year-old male, where extensive evaluation revealed primary Sjögren's syndrome. This case highlights that epistaxis can be caused by autoimmune cases,for epistaxis it is better to do screening for autoimmune examination test to exclude any autoimmune cases. This cases taken in public hospital type.

This case highlights the rare presentation of epistaxis as an initial sign of Sjögren's syndrome and underscores the importance of comprehensive clinical assessment and appropriate diagnostic approaches.

CASE PRESENTATION

A 30-year-old asian male admitted directly to the emergency department on November 30th, 2023 with recurrent, profuse epistaxis originating from both the right and left nasal cavities, accompanied by fever, nausea, and a persistent bitter taste in the mouth. There is no medical history. Patient never had a blood transfusion before. Patient had no drug allergies before. The patient uses glasses because he often feels dazzled. Family history: There is no family member suffering from the same disease as the patient. There is no family member suffering from cancer or autoimmune disease. The patient has never had a smoking habit. The patient denied any associated signs of lacerations, crepitus, or neoplastic growth.A thorough physical examination revealed a hypertensive crisis (202/126 mmHg) alongside substantial bilateral nasal hemorrhage, hematemesis, and xerostomia. The patient received therapy with adrenaline tampon therapy, Tranexamat acid 500mg IV injection, Esomeprazole 40mg IV injection, Captopril 25mg Sublingual, Carbazchrome sodium sulfonate 10mg injection.

Laboratory examinations were promptly conducted on December 1st, 2023, revealing a normal complete blood count. Moreover, clinical chemistry analysis indicated normal biochemical parameters across multiple systems. The patient's urea level was 7.72 mg/dL (reference range: 5–20 mg/dL), creatinine level was 0.72 mg/dL (0.7–1.2 mg/dL), potassium level was 3.71 mMol/L (3.5–5.0 mEq/L), and sodium level was 136 mMol/L (136–145 mMol/L). Additionally, calcium and magnesium levels were measured at 8.4 mg/dL (8.5–10.5 mg/dL) and 2.08 mg/dL (1.3–2.1 mEq/L), respectively. Liver function test, including SGOT level of 26.8 U/L (normal range: 8–45 U/L) and SGPT level of 50.2 U/L(normal range: 7–56 U/L), were within the normal range. Coagulation parameters, comprising of activated partial thromboplastin time (APTT) at 30.09 seconds (25–35 seconds) and prothrombin time (PT) at 12.22 seconds (11–13.5 seconds), also remained within reference limits. Furthermore, uric acid was recorded at 5.93 mg/dL (4.0–8.5 mg/dL), while thyroid function tests revealed a free T4 level of 1.21 ng/dL (0.8–1.9 ng/dL) and a TSH level of 1.03 µIU/mL (0.5–5.0 µIU/mL). Blood glucose regulation appeared intact, with a blood glucose level of 126 mg/dL (<126 mg/dL), fasting blood sugar at 98 mg/dL (70–100 mg/dL), and HbA1c at 5.60% (4.0–5.6%). Collectively, these findings suggest preserved renal, hepatic, coagulation, and glycemic functions. . However, lipid profile assessment demonstrated a markedly low high-density lipoprotein (HDL) level of 25 mg/dL (normal: 35-65 mg/dL) and a significant elevation in triglycerides, measured at 534 mg/dL (normal: < 150 mg/dL), suggesting a dyslipidemic profile. The international normalized ratio (INR) measured at 0.85, slightly below the standard reference range of 1.0, indicating a mild tendency towards reduced blood clotting ability.

On December 2nd, 2024, the patient still complained of blood leakage on the tampon. Because there was still blood leakage, a platelet aggregation test was performed. A platelet aggregation test (PAT) showed epinephrine at 20 (normal: 15-49), adenosine diphosphate (ADP) at 47 (normal: 50-82), and collagen at 5 (normal: 55-92). These findings indicated hypo-aggregation to both ADP and collagen, a result commonly associated with autoimmune conditions. Therapy for this platelet disorder uses folic acid 1 mg therapy twice daily and mecobalamin 500 micrograms once daily.

Imaging studies were performed on December 2nd, 2024 to assess the potential presence of tumors as the primary cause of epistaxis. A chest X-ray (anteroposterior view in a semi-upright position) and electrocardiography (ECG) demonstrated normal findings. Head CT angiography identified minimal hemorrhagic fluid within the left maxillary sinus and left nasal cavity, but no masses or abnormal lesions were observed in the nasal cavity, paranasal sinuses, nasopharynx, or pharynx. A small, well-defined lesion was observed in the left lateral pterygoid muscle, measuring 2.9 x 2.1 x 1.5 cm, thus confirming a presence of focal sialadenitis.

On December 3th, 2024 Anti-nuclear antibody (ANA) profile was performed, subsequent testing of the anti-nuclear antibody (ANA) profile indicated a positive result for the anti-Sjögren syndrome type B (SSB) antibody. Taken together, the diagnosis of primary Sjögren's syndrome (pSS) was established.

DISCUSSION

Sjögren's syndrome is characterized by autoimmune manifestations of dryness affecting the eyes, mouth, and other mucous membranes due to the infiltration of the lacrimal and salivary glands (Carsons & Patel, 2023b; Niu et al., 2017). Primary Sjögren's syndrome is diagnosed when these manifestations occur independently, without other underlying autoimmune disorders. However, these clinical features can overlap with conditions such as rheumatoid arthritis and systemic lupus erythematosus, complicating definitive diagnosis (Bowman, 2018; Carsons & Patel, 2023b; Hou et al., 2024).

The pathogenesis of primary Sjögren's syndrome remains complex (Mititelu et al., 2024). This condition exhibits diverse otolaryngologic manifestations, including glandular, ocular, oral, otologic, pharyngeal, laryngeal, and sinonasal involvement. Our patient presented significant oral involvement, with pronounced xerostomia. Salivary hypofunction, a hallmark feature in Sjögren's syndrome, is defined as a salivary flow rate reduced to less than 50% of normal (Amulya et al., 2024). Research suggests that this condition may result from chronic lymphocytic infiltration and inflammation of acinar cells, leading to exocrine fibrosis and subsequent glandular damage. Other studies propose that dysregulation of nerve stimuli may also contribute (Ishikawa, 2018; Rihab et al., 2023).

While oral manifestations are common in Sjögren's syndrome, our patient exhibited nasal involvement, specifically epistaxis, which is relatively uncommon for this disorder. Emerging evidence indicates that these manifestations may arise from autoimmune-mediated damage to nasal exocrine glands, disrupted mucosal muscarinic receptor signaling, and extracellular matrix degradation (Amulya et al., 2024; Beckman et al., 2017). Reports suggest nasal symptoms may include rhinorrhea, congestion, crusting, epistaxis, and olfactory dysfunction, attributed to submucosal infiltration by collagen bundles penetrating the disrupted basal lamina (Amulya et al., 2024; Eren et al., 2021; Lee & Kim, 2023).

The broad spectrum of clinical manifestations of primary Sjögren's syndrome presents diagnostic challenges. Unfortunately, no single diagnostic test can definitively confirm the syndrome. Instead, a comprehensive diagnostic approach is required, incorporating Schirmer's test, ocular surface staining, salivary gland function assessment, ultrasonography, sialometry, biopsy, and ANA testing (Freitas et al., 2024; Wedari et al., 2022). Among these, ANA testing is crucial, as the presence of anti-Ro/SSA and/or anti-La/SSB autoantibodies is observed in approximately 70% of primary Sjögren's syndrome cases, as noted in our patient (Parisis et al., 2020). However, these autoantibodies

are not disease-specific, as they can also appear in conditions like systemic lupus erythematosus, scleroderma, and rheumatoid arthritis (Barnado et al., 2024; Nosal et al., 2022; Wedari et al., 2022).

The heterogeneity of the primary Sjögren's syndrome necessitates careful consideration, with individualized treatment should be provided based on the expected course of the disease. Therapeutic strategies should be designed based on the anticipated disease progression and the extent of its impact on the patient's quality of life (Puwen et al., 2020).

Methycobalamin and folic acid are critical for the methylation of proteins, phospholipids, neurotransmitters, RNA, and DNA. Methycobalamin (mecobalamin) and folic acid play an important role in the formation of blood cells and DNA synthesis. In the process of cell development, it usually takes about 5-7 days from the initial stage of differentiation to becoming mature blood cells. However, the effects of vitamin B12 and folic acid supplementation on increasing cell production can begin to be seen within a few days (Sobczyńska-Malefora et al, 2021). After this therapy, the patient's complaints clinically improved but no further platelet aggregation tests were performed.

The strength of this case is that the cause of the platelet dysfunction is an autoimmune disease, but the weakness of this case is that platelet aggregation tests were not done again so that platelet function could not be monitored after therapy.

CONCLUSION

Primary Sjögren's syndrome is a multifaceted and heterogeneous autoimmune disorder with diverse clinical manifestations, typically characterized by dryness and discomfort in the mouth and eyes. While uncommon, Sjögren's syndrome may also involve the nasal cavity, presenting atypically with epistaxis. These observations underscore the importance of comprehensive clinical evaluation, accurate diagnosis, and timely treatment to ensure better outcomes.

CONFLICT OF INTEREST

The authors declare that they have no competing interests. The fund come from authors.

ETHICAL CLEARANCE

This study has received ethical clearance from Universitas Wijaya Kusuma Surabaya with the number 129/SLE/FK/UWKS/2024.

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