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Concomitant Presentation Of Adult Sjögren's Syndrome With Lupus Anticoagulant-Hypoprothrombinemia Syndrome: A Case Report

BinHan Liu, Yi Liu*

Department of Rheumatology and Immunology, West China Hospital, Sichuan University, Chengdu 610041, Sichuan, China.

***Correspondence to:**
Yi Liu,
Department of Rheumatology and Immunology, West China Hospital, Sichuan University, Chengdu 610041, Sichuan, China.
Tel.: +0086-15328075110
E-mail: yiliu_2020@scu.edu.cn

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1. Abstract

In the context of LAHS, a rare hemorrhagic disorder linked with systemic lupus erythematosus (SLE) and viral infections, we report an adult patient diagnosed with primary Sjögren's syndrome who later developed LAHS. The patient responded positively after undergoing steroid therapy and receiving the immunosuppressive agentcyclophosphamide. Remarkably, a 2-month follow-up revealed significant recovery. This case underscores the unique coexistence of Sjögren's syndrome and LAHS and the efficacy of the combined treatment approach.

2. Keywords:
Sjögren's syndrome, LAHS, factor II deficiency, lupus anticoagulant, steroids, cyclophosphamide.

3. Introduction

Lupus Anticoagulant-Hypoprothrombinemia Syndrome (LAHS) is a rare and intricate hemorrhagic disorder1. It is characterized by the unique simultaneous occurrence of acquired factor II deficiency and lupus anticoagulant (LA) presence. Historically, the incidence of LAHS has been predominantly observed among children and women. Moreover, it often presents with systemic lupus erythematosus (SLE) and is intricately linked with viral infections, adding complexity to its diagnosis and management2. Sjögren's, on the other hand, is an autoimmune disease in which the immune system mistakenly attacks the glands that produce tears and saliva, leading to dry eyes and mouth. Its primary form, where the syndrome exists independently, is distinguishable from its secondary form, which coexists with other autoimmune diseases3. The coexistence of Sjögren's syndrome with other autoimmune disorders is not uncommon. However, its concurrence with LAHS has been sparsely documented, making it a topic of immense medical interest4.

The epidemiology of both conditions provides valuable insights. While Sjögren's syndrome affects an estimated 1-4 million people in the US alone, its prevalence is much higher in adults, especially women, and LAHS remains a rare phenomenon5. Previous research has individually delved into the etiopathogenesis, clinical features, and management strategies for both conditions. However, limited literature is available on the simultaneous presentation of both disorders, emphasizing the value of this case report in bridging that knowledge gap6. The interplay between autoimmune diseases is intricate and not entirely understood. Their combined presentation can pose diagnostic challenges and demand a multidimensional approach to treatment. In this context, the current report aims to shed light on a unique case where an adult patient with primary Sjögren's syndrome developed LAHS7. By exploring this intersection, this study endeavors to provide a comprehensive understanding of the potential overlaps, pathophysiological mechanisms, and therapeutic interventions pertinent to these conditions8. Understanding such cases is pivotal in light of the limited existing literature on the confluence of Sjögren's syndrome and LAHS9. It enriches the clinical and academic discourse and offers new avenues for therapeutic strategies. Furthermore, by elucidating the intricacies of this combined presentation, the medical community can be better equipped to manage similar cases in the future, ensuring optimized patient care10.

4. Case report

4.1. Chief Complaint: A 57-year-old female presented with a 13-year history of joint pain and complaints of gum bleeding and skin bruising for the past 3 months.

4.2. History of Present Illness: The patient first experienced joint pain 13 years ago, which has been persistent. For the last three months, she noticed gum bleeding and skin bruises without any evident cause. The patient had a previous medical history of cerebral infarction.

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4.3. Physical Examination: On admission, ecchymosis was observed on both lower extremities and dental caries were present in the oral cavity. No other significant abnormalities were detected.

Laboratory Findings:

- Positive anti-nuclear antibodies with a titer of 1:320 show a granular and homogeneous pattern.
- The ENA profile revealed elevated anti-SS-A and anti-Ro-52 antibody levels, while anti-SS-B was lower.
- RF was 20.60 IU/mL, and the anti-CCP antibody was 15.40 U/mL.
- Positive Coombs test.
- Decreased complement levels of C3 and C4.
- ESR was elevated at 63.0 mm/h, and CRP was 7.06 mg/L.
- Positive phospholipid syndrome-related antibodies: ACAIgA 66.20 APLU/mL, ACA IgG >120.00 GPLU/mL, ACAIgM 64.20 MPLU/mL.
- Elevated levels of anti-β2GP1 IgA, IgG, and IgM.

4.4. Additional Examinations: Salivary gland SPECT scan indicated impairment in both parotid and submandibular glands. Peripheral venous color Doppler ultrasound showed no thrombus formation. Diagnosis and Treatment: The patient did not meet the diagnostic criteria for rheumatoid arthritis (RA) due to the absence of significant joint symptoms. The revised diagnosis was “Sjogren’s syndrome; lupus anticoagulant-hypocoagulable syndrome”. Upon admission, the patient received 40mg of intravenous methylprednisolone daily, which led to a noticeable improvement in factor II activity. Subsequently, the patient was prescribed 50mg of oral cyclophosphamide daily for immune suppression and was successfully discharged.

Follow-up: A two-month follow-up at the outpatient clinic revealed significant improvement in factor II activity. The platelet count was 41*10^9/L, and all phospholipid syndrome-related antibodies tested positive. The patient will continue to be monitored closely, with plans to taper off the steroid dosage, evaluate the efficacy of cyclophosphamide, and explore new and effective treatment strategies.

5. Discussion

LAHS represents a less common coagulation disorder and an autoimmune disease characterized by lupus anticoagulants and diminished prothrombin levels11. Lupus anticoagulants are a subset of autoantibodies that interrupt the routine clotting process, increasing predisposition to excessive blood clot formation12. Conversely, hypoprothrombinemia is typified by the reduction in the levels of a crucial protein, prothrombin, within the coagulation system, which facilitates clot formation13. Clotting gets compromised when there’s a drop in prothrombin levels13. The precise etiology of LAHS remains enigmatic, but research points towards aberrant immune system functionality as a pivotal component in its pathogenesis14. In its aberrancy, the immune system produces antibodies that target the body’s clotting factors, disrupting the routine clotting cascade14. Clinically, this syndrome manifests as recurrent bouts of both thrombosis and bleeding15. Patients may manifest with conditions like thrombotic venous inflammation, deep vein thrombosis, and pulmonary embolism16. Furthermore, owing to the diminished activity of clotting factors, patients may also manifest bleeding tendencies, such as skin ecchymosis, epistaxis, and menstrual irregularities15. In the tapestry of prior case reports or diminutive case series studies, LAHS appears to chiefly afflict children and women . It’s observed that the younger cohort is more likely to have associations with infections rather than autoimmune disorders17. However, among adults, LAHS frequently correlates with autoimmune diseases18. The primary manifestation of LAHS is bleeding, but a subset around 10% of patients, experience thrombotic events. These thrombotic events occur more in patients with LAHS and autoimmune disorders than in those with infections alone19. For LAHS spurred by viral infections, it’s conventionally believed to resolve autonomously within a set timeframe20. Conversely, immune-mediated LAHS typically mandates immunosuppressive therapy21. The patient under our lens manifests a novel case of LAHS triggered by Sjögren’s syndrome, an erstwhile undocumented eventuality but still categorized under autoimmune disorders22.

At this juncture, no standardized therapeutic protocol or guideline is specifically tailored for LAHS23. The consensus pivots around supportive care and immunosuppressive therapy24. Corticosteroids emerge as the front-line therapeutic recourse, and the deployment of immunosuppressive agents becomes imperative for tapering down steroids25. Additionally, there’ve been isolated case reports that spotlight the efficacy of other therapeutic modalities like cyclophosphamide, azathioprine, intravenous immunoglobulin, and rituximab26, 27, 28, 29. It’s worth noting, however, that a subset of cases doesn’t register the anticipated therapeutic outcomes30. In this vignette, the therapeutic regimen encompassing methylprednisolone and cyclophosphamide manifested a commendable therapeutic response. Post a 2-month follow-up, the patient’s coagulation profile and factor II activity rebounded robustly, even breaching the upper echelons of the normal range31. Prospectively, deliberation will ensue to ascertain the potential incorporation of aspirin as a prophylactic measure against thrombosis32.

6. Conclusion

In summation, LAHS emerges as a rare disorder, presenting with a confluence of lupus anticoagulant and diminished prothrombin levels. This distinctive disorder is earmarked by recurrent episodes of both thrombosis and bleeding. This case study unfurls the maiden documented instance of adult-onset Lupus anticoagulant-hypoprothrombinemia syndrome (LAHS) juxtaposed with Sjögren’s syndrome. This revelation acts as a crucial beacon, signaling the medical fraternity to delve deeper into whether the pathogenesis, clinical features and prognosis of LAHPS might oscillate depending on the underlying autoimmune disorder. Additionally, the timely diagnosis and initiation of the apt therapeutic regimen are paramount in amplifying the prognosis for such affected individuals.

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