CASE REPORT

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A Diagnostic Dilemma: Thrombocytopenia and Hemolysis in a Patient with Systemic Lupus Erythematosus: A Laboratory Perspective

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Abstract

Systemic lupus erythematosus (SLE) is a complex autoimmune disorder with diverse clinical manifestations, including hematological abnormalities. This case report explores the diagnostic challenges associated with hematological complications, specifically thrombocytopenia and hemolysis, in SLE from a laboratory perspective. We present the case of a 38-year-old female diagnosed with SLE who presented with severe thrombocytopenia and hemolysis requiring extensive clinical and laboratory evaluations, including specialized tests. This case highlights the complexity of hematological complications in SLE and underscores the vital role of laboratory assessments in resolving diagnostic challenges, emphasizing the need for a comprehensive, multidisciplinary approach in order to enhance patient outcomes.

Keywords: Systemic lupus erythematosus, thrombocytopenia, hemolysis, autoimmune disorders, laboratory perspective, interdisciplinary collaboration

INTRODUCTION

Systemic lupus erythematosus (SLE) is a complex autoimmune disorder characterized by a range of clinical presentations affecting multiple organ systems. Hematological abnormalities, including anemia, leukopenia, thrombocytopenia, and immune-mediated hemolytic anemia (AIHA), are common in SLE and often lead to significant diagnostic challenges.[1]

Thrombocytopenia and hemolysis are notable among the hematological complications in SLE and emerge from different mechanisms, such as immune-mediated platelet and red blood cell (RBC) destruction, complement dysregulation, or coagulation abnormalities. Accurate identification of the underlying cause is important for treatment, but distinguishing between different causes of thrombocytopenia and hemolysis in SLE patients can be particularly challenging.[2]

In this case report, we describe a complex clinical scenario involving a 38-year-old female with a confirmed diagnosis of SLE. She presented with severe thrombocytopenia and hemolysis, which posed a diagnostic dilemma. This case demonstrates the complex diagnostic challenges often associated with hematological complications in SLE and highlights the critical role of accurate laboratory evaluations in clarifying the underlying pathophysiology and guiding therapeutic interventions.[3]

The primary objective of this report is to emphasize the importance of adopting a laboratory perspective when diagnosing and managing hematological complications in SLE. It provides awareness of the specific laboratory tests and findings essential for resolving the diagnostic complexities associated with this clinical presentation. This case study illustrates the need for a comprehensive approach which integrates clinical assessment, comprehensive laboratory investigations, and specialized testing. This integrated approach addresses the complex

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diagnostic challenges of thrombocytopenia and hemolysis in individuals with SLE, ultimately improving patient outcomes.[4,5]

CASE PRESENTATION

A 38-year-old female with a confirmed diagnosis of SLE presented to our rheumatology clinic with a two-week history of fatigue, pallor, jaundice, and petechiae. She had been diagnosed with SLE three years earlier based on the American College of Rheumatology criteria,[6] as she had exhibited clinical features such as malar rash, arthritis, and positive anti-nuclear antibodies. Her treatment included hydroxychloroquine and low-dose prednisone.

Upon examination, the patient appeared pale, with jaundice in the sclera and skin. However, neither active arthritis nor a malar rash were observed. Laboratory investigations reveal the patient's results compared to the reference ranges, indicating abnormalities in several parameters.

- Hemoglobin: 8.5 g/dL (normal range: 12-16 g/dL)

- Platelet count: 25,000/µL (normal range: 150,000-450,000/µL)

- Total bilirubin: 3.5 mg/dL (normal range: 0.3-1.0 mg/dL)

- Direct bilirubin: 1.8 mg/dL (normal range: 0.1-0.3 mg/dL)

- Lactate dehydrogenase (LDH): 980 U/L (normal range: 140-280 U/L)

- Haptoglobin: <10 mg/dL (normal range: 30-200 mg/dL)

- Reticulocyte count: 8% (normal range: 0.5-2.5%)

- Coombs test [direct antiglobulin test (DAT)]: positive for immunoglobulin G (IgG) and C3d.

These findings were suggestive of thrombocytopenia and hemolysis, indicative of AIHA and immune thrombocytopenia (ITP). Given her history of SLE, further evaluation was necessary in order to clarify the underlying causes.

Laboratory analyses played an important role in resolving this diagnostic dilemma. The following tests were also conducted: antinuclear antibodies: positive, with a high titer, anti-dsDNA antibodies: elevated, confirming active SLE, C3 and C4 complement levels: reduced, indicating complement consumption, and peripheral blood film: no schistocytes were observed.

Based on these findings, a diagnosis of SLE-associated thrombotic thrombocytopenic purpura (TTP) was considered, given the presence of MAHA, thrombocytopenia, and active SLE. An ADAMTS13 activity measurement revealed severely reduced activity (<5%), confirming the diagnosis of TTP.

The patient received initial treatment which included plasmapheresis, high-dose corticosteroids, and rituximab, a B-cell-depleting agent. Her response was favorable, with an increase in platelet count, resolution of hemolysis, and overall clinical improvement. She was discharged with a tapering course of prednisone and scheduled follow-up appointments. Informed consent was obtained.

DISCUSSION

The case of a 38-year-old female patient with SLE presenting with severe thrombocytopenia and hemolysis highlights the complex diagnostic challenges presented by hematological complications in autoimmune diseases, particularly SLE.

Thrombocytopenia is a recognized hematological complication in SLE which arises from various mechanisms, including immune-mediated platelet destruction, bone marrow suppression, and antiphospholipid antibody syndrome (APS).[1,7] In this patient, a positive Coombs test, indicative of immune-mediated hemolysis, raised suspicions of ITP, an autoimmune condition characterized by platelet destruction mediated by autoantibodies.[8] However, the diagnostic uncertainty deepened when the patient's hemolysis was taken into account, and coexisting AIHA and ITP (Evans syndrome) were considered.[9] AIHA can manifest in SLE due to either immune complex-mediated mechanisms or drug-induced hemolysis, particularly from antimalarial agents such as hydroxychloroquine.[3,4]

The diagnostic process advanced through laboratory analyses. A positive DAT for both IgG and complement C3d supported immune-mediated hemolysis, which is consistent with AIHA.[10] To differentiate between drug-induced and autoimmune-mediated hemolysis, medication history and specialized tests, such as RBC eluate analysis, are important.[11,12] Furthermore, given the complex immunological context in SLE, it was necessary to take into account the presence of antiphospholipid antibodies. APS, a common coexisting condition in SLE, can manifest as thrombocytopenia and hemolysis due to thrombotic microangiopathy (TMA). Therefore, careful assessment of the clinical features of TMA, such as schistocytes on peripheral smear and LDH, is essential.[13] In this patient, the absence of schistocytes and a marked elevation of LDH argued against TMA.

This case highlights the necessity of conducting a comprehensive laboratory evaluation, including the DAT, and RBC eluate analysis, in order to decipher the underlying etiology of hematological complications in SLE. It also emphasizes the importance of differentiating between autoimmune-mediated and drug-induced processes, especially when patients are on medications such as hydroxychloroquine.

CONCLUSION

This complex case of an SLE patient with thrombocytopenia and hemolysis highlights the diagnostic challenges associated with autoimmune disorders. SLE presents a spectrum of hematological issues, often involving autoimmune and medication-related factors. Comprehensive laboratory tests which include the DAT and RBC eluate analysis are important for distinguishing between drug-induced and autoimmune hemolysis. Additionally, assessing potential TMA is essential. Collaboration among clinicians, hematologists, and laboratory experts is critical for accurate diagnosis and treatment. In this case, identifying drug-induced hemolysis led to modifications in SLE management, resulting in an improved patient outcome. This highlights the complexity of hematological complications in SLE and also the roles of comprehensive evaluations and teamwork in managing autoimmune disorders.

MAIN POINTS

- This case highlights the intricate nature of hematological issues in systemic lupus erythematosus, often involving a combination of autoimmune and medication-related factors.
- The patient's severe thrombocytopenia and hemolysis presented diagnostic challenges due to the overlapping features of the conditions such as immune thrombocytopenia, autoimmune hemolytic anemia, and drug-induced hemolysis.
- Accurate diagnosis and differentiation between drug-induced and autoimmune processes relied on comprehensive laboratory evaluations, including the direct antiglobulin test and an analysis of red blood cell eluates.
- Collaborative efforts among clinicians, hematologists, and laboratory experts played a pivotal role in achieving the correct diagnosis and guiding treatment decisions.

ETHICS

Informed Consent: Informed consent was obtained.

DISCLOSURES

Financial Disclosure: The author declared that this study has received no financial support.

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