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CASE REPORT

Thrombotic Thrombocytopenic Purpura as the Initial Manifestation of Systemic Lupus Erythematosus: A Case Report

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ABSTRACT

Background: Thrombotic thrombocytopenic purpura (TTP) rarely heralds systemic lupus erythematosus (SLE). We report such a presentation.

Methods: A 29-year-old woman presented with microangiopathic hemolytic anemia, thrombocytopenia and neurologic symptoms.

Results: ADAMTS13 activity was severely reduced. Autoimmune work-up revealed positive antinuclear and anti-dsDNA antibodies with low complement, consistent with SLE. She responded to plasma exchange and immunosuppression.

Conclusions: TTP may be the first manifestation of SLE. Prompt recognition and plasma exchange are life-saving; autoimmune screening should follow.

KEYWORDS

thrombotic thrombocytopenic purpura; systemic lupus erythematosus; ADAMTS13; plasma exchange; microangiopathy

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INTRODUCTION

TTP is a thrombotic microangiopathy caused by severe deficiency of ADAMTS13. Association with autoimmune disease is recognized, but TTP as the presenting feature of SLE is uncommon. Severe deficiency of ADAMTS13 leads to accumulation of ultralarge von Willebrand factor multimers and platelet-rich microthrombi within the microcirculation. When TTP arises in the context of autoimmunity, distinguishing it from other thrombotic microangiopathies is critical because delayed plasma exchange is associated with high mortality.

METHODS

We describe clinical presentation, laboratory findings, treatment and outcome.

RESULTS

The pentad was incompletely expressed. Blood film showed schistocytes; LDH was markedly elevated. ADAMTS13 activity was <10%. Daily plasma exchange, corticosteroids and rituximab produced remission over two weeks.

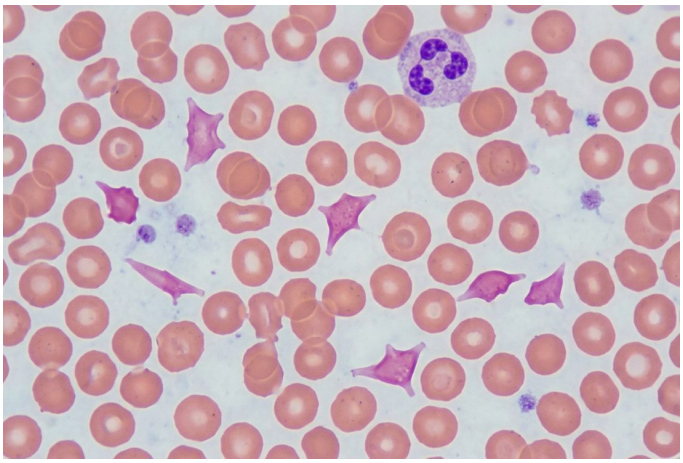
DISCUSSION

The overlap between TTP and SLE reflects shared autoimmune mechanisms. Distinguishing TTP from lupus nephritis-associated microangiopathy is essential because treatment differs. Early plasma exchange remains the cornerstone. The coexistence of TTP and SLE may reflect a shared loss of immune tolerance, with autoantibodies directed against ADAMTS13. Rituximab is increasingly used to reduce relapse in immune-mediated TTP. Long-term rheumatologic follow-up is warranted because the hematologic presentation may precede other manifestations of lupus.

CONCLUSION

Clinicians should screen for SLE in patients presenting with acquired TTP, as concurrent immunosuppression may improve outcomes.

Figure 1. Peripheral blood smear



High-power photomicrograph (Wright-Giemsa) showing numerous fragmented erythrocytes (schistocytes) and thrombocytopenia, characteristic of microangiopathic hemolytic anemia.

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