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Severe Thrombocytopenia, TTP, and Renal TMA in Systemic Lupus Erythematosus: Pathophysiologic Links and Therapeutic Implications

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Abstract

Introduction:

Systemic lupus erythematosus (SLE) is a chronic autoimmune disease with multisystem involvement. Thrombotic microangiopathy (TMA) complicates lupus nephritis in a subset of patients and is associated with poor renal outcomes. Key etiologies of TMA in SLE include thrombotic thrombocytopenic purpura (TTP), antiphospholipid syndrome (APS), and complement-mediated TMA. Concurrent presentation of these processes represents a severe and diagnostically challenging phenotype. This case highlights the convergence of ADAMTS13-deficient TTP, APS-associated TMA, and immune-mediated thrombocytopenia in a single patient with SLE.

Case Presentation:

A 29-year-old female with SLE (anti-Smith, anti–dsDNA, and anti-RNP positive) presented with petechiae and headache. Laboratory evaluation revealed severe thrombocytopenia (10,000/µL), proteinuria, hematuria, and elevated creatinine, consistent with an active lupus flare. CT brain was unremarkable. She was treated with high-dose intravenous methylprednisolone followed by oral prednisone, intravenous immunoglobulin (IVIG), and platelet transfusion, with platelet recovery to 132,000/µL by day 7. Kidney biopsy demonstrated class IV lupus nephritis with superimposed TMA (NIH activity index 11/24, chronicity index 0/12). ADAMTS13 activity was <10%, confirming TTP. Triple-positive antiphospholipid antibodies were also identified.

She was started on immunosuppressive therapy with prednisone, mycophenolate mofetil, and tacrolimus. Warfarin anticoagulation (target INR 2–3) was initiated for APS. Given the coexistence of APS and ADAMTS13 deficiency, anticoagulation was prioritized over complement inhibition.

Discussion:

This case illustrates the overlapping mechanisms of TMA in SLE, including ADAMTS13-deficient TTP, APS-related thrombosis, and immune thrombocytopenia. Severe ADAMTS13 deficiency promotes formation of platelet-rich microthrombi, while antiphospholipid antibodies further amplify thrombotic risk through endothelial and platelet activation. The patient's thrombocytopenia was likely multifactorial, reflecting platelet consumption, immune-mediated destruction, and antibody-driven activation.

Key Clinical Insight:

In patients with SLE and TMA, early differentiation among TTP, APS, and complement-mediated TMA is essential, as treatment strategies differ significantly, ranging from immunosuppression to anticoagulation or complement inhibition, and delays in appropriate therapy can lead to irreversible organ damage. This case underscores the importance of a comprehensive diagnostic approach, including ADAMTS13 testing and antiphospholipid antibody evaluation, to guide targeted, mechanism-driven treatment and improve clinical outcomes.