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Case Report

Thrombotic Thrombocytopenic Purpura and Systemic Lupus Erythematosus – A Rare Association

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ABSTRACT

Thrombotic thrombocytopenic purpura (TTP) is a life-threatening disorder characterized by a pentad of microangiopathic hemolytic anemia (MAHA), thrombocytopenia, renal dysfunction, neurological symptoms, and fever. Systemic lupus erythematosus (SLE) is a chronic autoimmune disease primarily affecting young females, involving multiple organ systems due to immune complex deposition and autoantibody-mediated tissue damage. The coexistence of SLE and TTP is rare but presents a significant diagnostic and therapeutic challenge. We report a case of an 18-year-old female presenting with fever, menorrhagia, and neurological deficits. Despite platelet transfusions, her cytopenia persisted. Laboratory investigations revealed hemolysis, ANA positivity, and MRI brain abnormalities. Due to financial constraints, plasmapheresis could not be performed, and the patient was successfully treated with corticosteroids and cyclophosphamide. This case highlights the importance of early recognition and alternative treatment strategies in resource-limited settings.

Keywords: Thrombotic thrombocytopenic purpura, Systemic lupus erythematosus, Autoimmune hemolytic anemia, Cyclophosphamide, Microangiopathy

INTRODUCTION

Thrombotic thrombocytopenic purpura (TTP) is a rare but life-threatening hematological disorder caused by severe deficiency of ADAMTS13, a von Willebrand factor-cleaving protease. This leads to excessive platelet aggregation and widespread microvascular thrombosis. Classic TTP presents with a pentad of findings: Microangiopathic hemolytic anemia (MAHA), Thrombocytopenia, Renal failure, Neurological abnormalities and Fever. [1,2]

Systemic lupus erythematosus (SLE) is a systemic autoimmune disease characterized by immune complex-mediated damage to various organs. It predominantly affects young women, with multisystem involvement, including renal, hematological, and neurological manifestations. The co-occurrence of SLE and TTP is uncommon but represents a diagnostic and therapeutic challenge due to overlapping clinical features. Prompt identification and treatment are essential to prevent fatal outcomes.[1,2,3]

This report presents a rare case of an 18-year-old

female with TTP associated with SLE, successfully managed with corticosteroids and cyclophosphamide in the absence of plasmapheresis.

Case Presentation

An 18-year-old female presented with complaints of Fever (on and off) for 10 days, Menorrhagia for 10 days and Joint pain and headache. She had been admitted to another hospital, where she received platelet and packed red blood cell (PRBC) transfusions. However, her cytopenia persisted, leading to referral to our hospital. On Day 4 of admission, she had a generalized tonic-clonic seizure (GTCS) and was in a confused state for a day.

On Day 12, she developed Left upper limb weakness, Right lower motor neuron (LMN) facial palsy and Persistent altered sensorium since Day 5.

Examination Findings

Neurological Examination showed Altered consciousness from Day 5, Left upper limb weakness and Right LMN facial palsy. Hematological Examination showed Petechiae noted on the lower limbs, Cardiorespiratory & Abdominal Examination and Unremarkable.

Investigations

Table 1: Hemogram

Parameter	Result	Reference Range
Complete Blood Count (CBC)		
Hemoglobin	7.2 g/dL	12-16 g/dL
Total Leukocyte Count	6,800/mm³	4,000-11,000/mm³
Platelet Count	18,000/mm³	150,000-450,000/mm³
Reticulocyte Count	5.2%	<2.5%

Peripheral Smear showed Anisopoikilocytosis, schistocytes, macrocytes, tear-drop cells. Direct Coombs Test (DCT) was Negative and Indirect Coombs Test (ICT) was also Negative.

Table 2: Renal function tests

Parameter	Result	Reference Range
Serum Creatinine	1.8 mg/dL	0.5-1.2 mg/dL
Blood Urea	28 mg/dL	7-20 mg/dL
24-hour Urinary Protein	891 mg/24 hours	<150 mg/24 hours

- Autoimmune Workup showed ANA Positive (10.80)- Negative
- ANA Profile: Sm Ab (+), Sm/RNP Ab (+)- Negative
- MRI Brain: Multiple tiny foci of GRE hypo intensities within bilateral cerebellum and bilateral perisylvian cortex Normal
- Plasmic Score 6 (suggestive of TTP)
- Due to resource limitations, ADAMTS13 activity could not be tested. The diagnosis of TTP was made based on clinical criteria and the Plasmic Score.

Management & Treatment

From Day 1-5 she was given with 3 units of PRBC transfusion and 8 units of platelet transfusion → No improvement in cytopenia. From Day 5-9 she was given Pulse methylprednisolone therapy (1 g

IV for 3 days) and Planned plasmapheresis but not affordable. On Day 12, she has been given Cyclophosphamide, initiated due to persistent cytopenias and neurological symptoms. Significant improvement in platelet count, hemoglobin, and renal function. Neurological symptoms improved

DISCUSSION

The coexistence of SLE and TTP is rare but potentially life-threatening. The pathophysiology of TTP in SLE is believed to involve Endothelial Injury & Autoantibodies and immune complexes damage endothelial cells, leading to platelet aggregation. ADAMTS13 Deficiency leads to Reduced enzyme activity results in accumulation of ultralarge von Willebrand factor (vWF) multimers, causing microvascular thrombosis.[3,4,5]
Differential Diagnosis Considerations [6,7]:

- SLE flare vs. TTP: Overlapping features make differentiation challenging.
- Antiphospholipid Syndrome (APS): Ruled out due to the absence of thrombosis and negative lupus anticoagulant.
- Hemolytic Uremic Syndrome (HUS): Distinguished by severe renal dysfunction, which was not predominant in our case.

Standard Treatment Approach: First-line therapy for TTP is Plasmapheresis (PLEX). Corticosteroids Reduce inflammation and autoantibody production and Rituximab or Caplacizumab can be Used in refractory cases.[5,8,9]
Alternative Treatment Strategy in Resource-Limited Settings: Due to financial constraints, cyclophosphamide was used as an adjunct therapy, leading to significant improvement. Although cyclophosphamide is not a standard therapy for TTP, it has shown effectiveness in severe autoimmune cases.[5,6,11]

CONCLUSION

TTP and SLE can coexist, posing a diagnostic and therapeutic challenge. Early recognition and intervention are crucial for preventing life-threatening complications. Plasmapheresis remains the gold standard for TTP, but in resource-limited settings, cyclophosphamide and corticosteroids can be considered as alternative therapy. This case highlights the need for tailored management strategies based on patient affordability and available resources.

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