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Acquired Immune-Mediated Thrombotic Thrombocytopenic Purpura as the Initial Presentation of Systemic Lupus Erythematosus in a Pediatric Patient: A Case Report

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1. Abstract

Thrombotic thrombocytopenic purpura (TTP) is a rare and potentially fatal thrombotic microangiopathy characterized by microangiopathic hemolytic anemia, severe thrombocytopenia, and organ ischemia resulting from severe ADAMTS13 deficiency. Pediatric acquired immune-mediated TTP is uncommon, and its coexistence with systemic lupus erythematosus (SLE) is exceptionally rare. We report the case of a 13-year-old previously healthy female who presented with non-blanchable purpuric rash, severe anemia, thrombocytopenia, recurrent vomiting, and neurological manifestations in the form of nocturnal hallucinations. Laboratory evaluation demonstrated microangiopathic hemolytic anemia with elevated lactate dehydrogenase, reticulocytosis, low haptoglobin, Negative Direct Antiglobulin Test, normal coagulation and 9% schistocytes on peripheral blood smear. ADAMTS13 testing confirmed acquired immune-mediated TTP with activity <0.01% and markedly elevated ADAMTS13 antibodies. Autoimmune workup revealed positive antinuclear antibody (22 U/ml), positive anti-SS-A antibodies and low complement C3 level (79 mg/dl). The patient required aggressive management with therapeutic plasma exchange, pulse corticosteroids, rituximab, and hydroxychloroquine. Her clinical course was complicated by refractory thrombocytopenia requiring escalation to twice-daily plasmapheresis, and increasing the steroid dose. Gradual hematological and clinical improvement was achieved following combined immunosuppressive therapy and plasmapheresis. This case highlights the diagnostic complexity and therapeutic challenges of pediatric SLE-associated acquired TTP and emphasizes the importance of early recognition, prompt ADAMTS13 testing, and aggressive multidisciplinary management to improve outcomes.

2. Introduction

Thrombotic thrombocytopenic purpura (TTP) is a rare, life-threatening thrombotic microangiopathy characterized by microangiopathic hemolytic anemia, severe thrombocytopenia, and ischemic organ injury caused by disseminated platelet-rich microvascular thrombi. It is associated with severe deficiency of ADAMTS13 activity, either congenital or, more commonly, acquired through inhibitory autoantibodies [1]. Pediatric acquired immune-mediated TTP is uncommon. Published pediatric cohorts and reviews demonstrate that autoimmune disorders, particularly systemic lupus erythematosus (SLE), are important associated conditions in older children and adolescents [2-4].

Systemic lupus erythematosus (SLE) is a chronic multisystem autoimmune disease with heterogeneous hematologic, neurologic, renal, and mucocutaneous manifestations. The coexistence of immune-mediated TTP and SLE is uncommon, particularly in pediatric patients, and creates a diagnostic challenge because active SLE may itself cause thrombocytopenia, hemolytic anemia, neurologic manifestations, renal abnormalities, and other forms

of thrombotic microangiopathy [5].

Early recognition of TTP is critical because treatment delay can be fatal. International Society on Thrombosis and Haemostasis guidance supports urgent therapeutic plasma exchange and corticosteroids for acute immune-mediated TTP and conditionally recommends adding rituximab and caplacizumab [6,7]. We report a rare case of acquired immune-mediated TTP associated with newly diagnosed SLE in a 13-year-old female who presented with severe microangiopathic hemolytic anemia, thrombocytopenia, and neurological manifestations requiring urgent intensive care management and aggressive immunosuppressive therapy.

3. Case Presentation

A 13-year-old previously healthy female patient was admitted to the Pediatric Intensive Care Unit on 13/01/2026 as a case of suspected thrombotic thrombocytopenic purpura (TTP) requiring urgent plasmapheresis. She had been in her usual state of health until five days prior to admission, when she developed a non-blanchable purpuric rash involving both lower extremities. Initial medical evaluation at an outpatient clinic revealed anemia and thrombocytopenia with hemoglobin of 8 g/dL and platelet count of 17 $\times 10^9$/L. The patient was discharged home with advice to repeat laboratory investigations after two days. Subsequently, her condition deteriorated with development of recurrent vomiting, cough, and episodes of nocturnal hallucinations, prompting presentation to the Hospital. Further laboratory investigations demonstrated severe microangiopathic hemolytic anemia and thrombocytopenia, with hemoglobin of 7 g/dL, platelet count of 10 $\times 10^9$/L, markedly elevated lactate dehydrogenase (LDH), increased reticulocyte count, low haptoglobin level, and schistocytes on peripheral blood smear. These findings were highly suggestive of thrombotic thrombocytopenic purpura, and the patient was urgently referred for plasmapheresis. There was no history of fever, urinary symptoms, abnormal movements, or preceding infectious illness.

Past medical history was unremarkable. However, the patient had a notable history of recurrent oral ulcers, for which upper and lower gastrointestinal endoscopy had been performed in April 2025 to rule out Crohn's disease, and both studies were unremarkable. Family history was significant for systemic lupus erythematosus (SLE) in her mother. Further history also revealed intermittent malar rash episodes.

On admission, laboratory evaluation confirmed severe microangiopathic hemolytic anemia and thrombocytopenia. Peripheral blood film demonstrated anemia with 9% schistocytes and no blasts. Hemolytic workup showed elevated indirect bilirubin, markedly elevated LDH, reticulocytosis, and decreased haptoglobin level. Direct and indirect Coombs tests were negative. Coagulation profile and renal function tests were preserved. Complement studies revealed low C3 level (79 mg/dL) with normal C4 level (12 mg/dL). Autoimmune workup demonstrated strongly positive antinuclear antibody (ANA) with a titer of 1:325 and positive anti-SS-A antibodies (+2). Antiphospholipid antibodies were negative. ADAMTS13 testing confirmed the diagnosis of acquired immune-mediated TTP, revealing ADAMTS13 activity <0.01%, ADAMTS13 antigen level of 0.01, and markedly elevated ADAMTS13 antibodies >80. Based on the clinical presentation, laboratory findings, autoimmune profile, recurrent oral ulcers, malar rash history, low complement level, and positive family history, the patient was diagnosed with acquired TTP associated with systemic lupus erythematosus.

Radiological evaluation by oncology computed tomography scan performed on 13/01/2026 demonstrated few bilateral prominent axillary lymph nodes, bilateral cervical lymphadenopathy measuring up to 9 mm, and para-aortic lymph nodes up to 1 cm in short axis. Mild hepatomegaly was noted with liver span measuring 14.5 cm, along with splenomegaly measuring 13 cm. Both kidneys were normal in size and morphology with mildly decreased attenuation and no hydronephrosis.

The patient was managed aggressively with prednisone 1mg /kg and daily therapeutic plasma exchange starting on 13/01/2026, as well as Rituximab 375mg/m2 mg weekly for four doses. Due to delayed platelet response, plasmapheresis intensity was escalated to twice daily sessions on 21/01/2026 for three consecutive days, then de-escalated back to once daily sessions. At the same time switched prednisone to methyl prednisone 500mg daily for five days then brought back again to oral prednisone 1mg/kg daily.

Immunosuppressive therapy included initially prednisone 1mg /kg but due to delayed response escalated to pulse methylprednisolone 500 mg daily for five days then de-escalated back to oral prednisone 1 mg/kg daily. Following rheumatology consultation, hydroxychloroquine (Plaquenil) 200 mg daily was initiated on 20/01/2026 for management of underlying SLE. Aspirin 100 mg daily was started but temporarily discontinued on 30/01/2026 because of severe thrombocytopenia, with plans to resume therapy once platelet count improved.

Supportive management included vitamin D supplementation with cholecalciferol 50,000 IU weekly for three months, calcium supplementation (Calcivid), proton pump inhibitor prophylaxis with Nexium 40 mg daily, and trimethoprim-sulfamethoxazole prophylaxis (Sulprim Forte every other day) during prolonged immunosuppressive therapy. The patient showed gradual clinical and hematological improvement with combined plasmapheresis and immunosuppressive therapy and was discharged on Prednitab, Plaquenil, aspirin, calcium/vitamin D supplementation, Nexium, and prophylactic Sulprim Forte, with close hematology and rheumatology follow-up arranged.

4. Discussion

Thrombotic thrombocytopenic purpura (TTP) is a rare but life-threatening thrombotic microangiopathy characterized by severe thrombocytopenia, microangiopathic hemolytic anemia (MAHA), and ischemic organ injury due to severe ADAMTS13 deficiency [1]. Pediatric acquired immune-mediated TTP is uncommon, and its association with SLE is particularly rare, posing substantial diagnostic and therapeutic challenges [2-4].

The presented case illustrates severe acquired immune-mediated TTP in a pediatric patient whose subsequent evaluation supported SLE as an associated autoimmune disorder. She initially presented with purpura, severe thrombocytopenia, and hemolytic anemia, followed by nocturnal hallucinations. Neurologic involvement in TTP may range from headache and confusion to seizures and altered mental status because of microvascular ischemic injury [1]. Early recognition is essential because TTP was associated with mortality exceeding 90% before effective plasma-exchange therapy became available [8].

The diagnosis of TTP in this patient was supported by severe thrombocytopenia, direct-antiglobulin-test-negative MAHA, elevated lactate dehydrogenase, reticulocytosis, reduced haptoglobin, and marked schistocytosis on peripheral blood smear. [1] Preserved coagulation parameters and renal function also favored TTP over disseminated intravascular coagulation and severe hemolytic uremic syndrome. Definitive testing showed ADAMTS13 activity <0.01% with markedly elevated anti-ADAMTS13 antibodies, confirming immune-mediated TTP [6].

An important aspect of this case was the coexistence of SLE. The patient had recurrent oral ulcers, a history of malar rash, positive ANA and anti-SS-A antibodies, low complement C3, and a maternal history of SLE. SLE is a recognized autoimmune disorder associated with immune-mediated TTP [5]. Complement activation and endothelial injury may also contribute to thrombotic microangiopathy in SLE [9]. Distinguishing SLE-associated immune-mediated TTP from other hematologic manifestations of active SLE is challenging because both can present with thrombocytopenia, hemolytic anemia, neurologic manifestations, and renal abnormalities; demonstration of severe ADAMTS13 deficiency with an inhibitor is therefore central to confirming immune-mediated TTP [5,6].

Therapeutic plasma exchange has historically been the cornerstone of acute immune-mediated TTP treatment because it removes circulating anti-ADAMTS13 autoantibodies and replenishes functional ADAMTS13. Current guidance recommends plasma exchange with corticosteroids and conditionally recommends the addition of rituximab and caplacizumab [7]. In this case, the patient received pulse methylprednisolone followed by oral corticosteroids and weekly rituximab. Rituximab depletes autoreactive CD20-positive B lymphocytes and is used to reduce anti-ADAMTS13 autoantibody production [7]. The hematologic response after combined plasma exchange, corticosteroids, and rituximab supports the value of early immunosuppression in immune-mediated TTP.

Hydroxychloroquine was initiated after rheumatology evaluation for the associated SLE. Beyond its established role in lupus control, hydroxychloroquine has immunomodulatory and potential antithrombotic effects [10]. In a retrospective cohort, Yue and colleagues reported different clinical features and outcomes in SLE-associated immune-mediated TTP compared with primary immune-mediated TTP; patients with SLE-associated disease and ADAMTS13 inhibitor positivity had favorable survival, possibly reflecting earlier recognition and intensive treatment of both conditions [5].

Our patient's presentation as an adolescent female is consistent with the reported epidemiology of pediatric acquired TTP. Pediatric cohorts describe a predominance of girls and an association with autoimmune disease, including SLE, particularly in older children and adolescents [2-4]. These observations support interpreting her TTP within the autoimmune spectrum of newly recognized SLE.

This case highlights the importance of maintaining a high index of suspicion for TTP in children presenting with MAHA and thrombocytopenia, particularly when neurologic manifestations are present [1,2]. It also emphasizes evaluation for associated autoimmune disease when clinical and immunologic features suggest SLE. Rapid initiation of TTP-directed therapy while ADAMTS13 testing is pending is essential to reduce morbidity and mortality [6,7].

In conclusion, acquired immune-mediated TTP associated with SLE in pediatric patients is a rare but potentially catastrophic condition requiring rapid multidisciplinary management. Our case demonstrates the diagnostic complexity, aggressive disease course, and therapeutic challenges associated with this overlap syndrome. Early recognition of MAHA, urgent ADAMTS13 testing, and aggressive treatment with plasma exchange and immunosuppressive therapy were critical in achieving clinical remission and favorable outcome in this patient.

5. Consent

Written informed consent was obtained from the patient for publication of this case report and any accompanying images.

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