


Case Report

Three in One: Systemic Lupus Erythematosus, HELLP Syndrome, and Antiphospholipid Syndrome: A Case Report and Literature Review

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Abstract

Introduction

Systemic lupus erythematosus (SLE) is a multisystem autoimmune disease commonly affecting women of reproductive age. Its overlap with HELLP syndrome (Hemolysis, Elevated Liver Enzymes, and Low Platelet Count) and antiphospholipid syndrome (APS) during pregnancy is rare and presents diagnostic and therapeutic challenges due to shared clinical and laboratory features. This report describes a case of pre-existing SLE complicated by the concurrent occurrence of SLE flare, HELLP syndrome, and APS during pregnancy.

Case presentation

A 26-year-old female with known SLE presented with right upper quadrant abdominal pain and hypertension two days following a mid-trimester abortion at 22 weeks of gestation. Laboratory evaluation revealed thrombocytopenia, elevated liver enzymes, and hemolysis, consistent with HELLP syndrome. The presence of lupus anticoagulant and anticardiolipin antibodies, in conjunction with her obstetric history, supported the diagnosis of APS. The patient was treated with high-dose corticosteroids and anticoagulation, resulting in significant clinical improvement.

Literature review

A review of literature highlights the consistent presentation of nonspecific symptoms such as abdominal pain, nausea, and hypertension in patients with overlapping SLE, HELLP syndrome, and APS. Laboratory findings often reveal thrombocytopenia, elevated liver enzymes, and hemolysis, reflective of underlying microangiopathic processes. Therapeutic strategies typically involve corticosteroids and anticoagulation, with plasmapheresis reserved for severe cases.

Conclusion

The coexistence of SLE flare, HELLP syndrome, and APS during pregnancy is a rare and complex condition that requires careful evaluation. Early recognition and appropriate management are crucial for achieving favorable outcomes.

1. Introduction

Systemic lupus erythematosus (SLE) is a chronic autoimmune disease with multisystem involvement, affecting approximately 20 to 150 per 100,000 individuals worldwide, with a strong female predominance [1,2]. It commonly presents with fatigue, arthralgia, skin manifestations, and renal or hematological complications, but its presentation can vary widely depending on disease activity [3]. Pregnancy in patients with SLE is considered high-risk due to increased risks of preeclampsia, fetal loss, and preterm delivery [4]. Antiphospholipid syndrome (APS) is another autoimmune disorder that frequently overlaps with SLE. It is characterized by the presence of antiphospholipid antibodies and is associated with thrombotic events and pregnancy complications such as recurrent miscarriage and intrauterine growth restriction [5,6]. HELLP (Hemolysis, Elevated Liver Enzymes, and Low Platelet Count) syndrome is a severe form of preeclampsia that can develop in up to 0.9% of pregnancies, usually leading to hepatic dysfunction, hypertension, and coagulopathy [7]. These conditions exhibit overlapping clinical and laboratory features such as thrombocytopenia, elevated liver enzymes, and hemolysis, making diagnosis and management particularly challenging [3-7].

This report presents a known case of SLE with the simultaneous occurrence of SLE flare, HELLP syndrome, and APS during pregnancy. The report was prepared in accordance with the CaReL guidelines, with all references evaluated for reliability and scientific integrity [8,9].

2. Case Presentation

2.1. Patient information

A 26-year-old woman presented with a one-week history of right upper quadrant abdominal pain. She had a second-trimester abortion at 22 weeks of gestation, delivered vaginally two days earlier. Her medical history included SLE, diagnosed eight years earlier following a miscarriage. Because of recurrent abortions, she was placed on aspirin during pregnancy. She also had a history of gestational hypertension.

2.2. Clinical findings

Examination revealed significant tenderness in the epigastric region upon light palpation, with no changes in mental status.

2.3. Diagnostic approach

Laboratory investigations revealed markedly elevated liver enzymes, including alanine aminotransferase (ALT: 2500 U/L; reference: 7–56 U/L) and aspartate aminotransferase (AST: 3000 U/L; reference: 10–40 U/L). She also had thrombocytopenia, with a platelet count of 50,000/mm³ (reference: 150,000–450,000/mm³), and a prolonged prothrombin time (PT) of 22 seconds (reference: 11–15 seconds) with an elevated international normalized ratio (INR: 2.0; reference: 0.8–1.2). D-dimer levels were markedly elevated at 2500 ng/mL (reference: <500 ng/mL), while fibrinogen was decreased to 50 mg/dL (reference: 200–400 mg/dL). Peripheral

blood smear demonstrated schistocytes, indicative of ongoing microangiopathy. Autoimmune testing confirmed active SLE, with a positive antinuclear antibody (ANA) and elevated anti-double-stranded DNA (anti-dsDNA) titers. Imaging studies, including an abdominal computed tomography scan, demonstrated severe hepatic necrosis without evidence of rupture or hematoma. These findings confirmed the coexistence of SLE flare, HELLP syndrome, and APS, a rare and complex presentation.

2.4. Therapeutic intervention and follow-up

The patient was admitted to the intensive care unit for supportive care and initiated methylprednisolone pulse therapy for three consecutive days, resulting in significant improvement. Liver enzymes normalized, platelet counts increased, and symptoms resolved. Anticoagulation with warfarin was started for APS, and hydroxychloroquine was continued for SLE management. At discharge, she was stable and referred to rheumatology for long-term follow-up.

3. Discussion

Pregnancy in patients with SLE carries an increased risk of adverse maternal and fetal outcomes, including preeclampsia, fetal loss, and preterm birth, particularly in cases complicated by active disease [5]. The presence of APS, a thrombotic disorder defined by antiphospholipid antibodies, further increases pregnancy risks by promoting thrombosis, recurrent miscarriages, and placental insufficiency [6].

HELLP syndrome, a severe form of preeclampsia, presents with hepatic dysfunction, endothelial injury, and hematologic abnormalities, leading to substantial maternal and fetal morbidity [7].

The present case developed HELLP syndrome and APS following a mid-trimester abortion at 22 weeks, reflecting the complex interplay of these conditions and the challenges in diagnosis and management. A brief literature review identified 4 reported cases of concurrent (SLE and APS), (SLE and HELLP syndrome), or (APS and HELLP syndrome). All occurred in young women aged 26–39 years, underscoring the well-established female predominance of these conditions (Table 1) [3-7].

Pregnancy is a known trigger for SLE flares, and the additional presence of APS further increases the risk of obstetric complications [2,4,5]. The present case is consistent with these observations, as the patient's history of SLE and pregnancy loss conferred an elevated risk for the development of APS and HELLP syndrome.

The most common symptoms reported in the reviewed cases were right upper quadrant abdominal pain, hypertension, nausea, and dyspnea, which are classic signs of HELLP syndrome [3-7]. Sakhel et al. reported tachypnea and dyspnea,

Table 1. Summary of the reviewed case reports describing the occurrence of at least two of SLE, APS, and HELLP syndrome concurrently.

Author name/year	Age (years)	Gender	Country	Associated conditions	Symptoms	Key laboratory findings	Therapeutic approach	Outcome	Follow-up (months)
Osmanagaoglu et al., 2004[4]	33	F	Turkey	SLE, HELLP	Synovitis, butterfly rash, confusion, convulsions	Elevated liver enzymes, proteinuria, and low platelets	Prednisolone, therapeutic abortion	Died	-
Veres et al., 2007[5]	26	F	Hungary	APS, HELLP	Hypertension, chest discomfort, vision changes	Elevated ALT, AST, LDH, low platelets, hemolysis	Plasmapheresis, anticoagulation	Recovered	24
Yamamoto et al., 2004[6]	26	F	Japan	SLE, HELLP	Severe epigastric pain, hypertension	Elevated liver enzymes, microangiopathic hemolysis	Plasma exchange, steroids	Recovered	12
Sakhel et al., 2006[7]	39	F	Lebanon	SLE, APS	Severe upper abdominal pain, tachypnea, dyspnea	Elevated liver enzymes, thrombocytopenia, hemolytic anemia	Methylprednisolone, heparin	Improved	6

SLE: Systemic Lupus Erythematosus; APS: Antiphospholipid Syndrome; HELLP: Hemolysis, Elevated Liver enzymes, Low Platelets; F: Female; ALT: Alanine Aminotransferase; AST: Aspartate Aminotransferase; LDH: Lactate Dehydrogenase

which can indicate pulmonary involvement or preeclampsia-related respiratory distress [7]. Osmanagaoglu et al. reported another case presenting with synovitis, butterfly rash, and confusion, indicative of either neurological lupus involvement or eclampsia [4]. Some cases presented with vision disturbances and chest discomfort, which may indicate APS-related microvascular thrombotic events [5]. While some patients had neurological involvement, the present case exhibited a severe hypertensive episode, reinforcing the complexity of these overlapping conditions.

Laboratory investigations across cases demonstrated consistent hematologic abnormalities, including thrombocytopenia, elevated liver enzymes, and hemolysis, which are defining features of HELLP syndrome [3]. One case reported proteinuria, which is more suggestive of SLE nephritis or preeclampsia-related renal dysfunction [4]. In another case, hemolysis was confirmed through microangiopathic findings rather than an immune-mediated mechanism [6]. Patients with APS frequently exhibited prolonged PT/INR, elevated D-dimer levels, and thrombocytopenia, reflecting the hypercoagulable state of APS [5].

The present case demonstrated severe thrombocytopenia, elevated ALT/AST, and hemolysis, with positive lupus anticoagulant and anticardiolipin antibodies confirming APS and elevated anti-dsDNA titers indicating active SLE. This profile closely mirrors the reviewed cases of the literature, emphasizing the diagnostic overlap between HELLP syndrome, SLE flares, and APS.

Although imaging findings were not reported in all cases, hepatic involvement was a significant feature in those with

severe HELLP syndrome or APS-associated thrombosis. Osmanagaoglu et al. reported a case of hepatic infarctions and subcapsular hematomas, suggesting vascular damage secondary to HELLP syndrome or APS-related thrombosis [7]. Veres et al. described another case with hepatic microangiopathic changes in the absence of infarction, highlighting the diverse spectrum of hepatic involvement in such patients [5]. The present case exhibited hepatic involvement in imaging, consistent with hepatic dysfunction in severe HELLP syndrome, reinforcing the value of imaging studies in distinguishing these conditions.

Management strategies varied depending on the severity of the disease, but high-dose corticosteroids were universally used to suppress SLE flares and reduce systemic inflammation [6]. APS patients received anticoagulation therapy with heparin or warfarin to prevent thrombotic complications. In severe cases, plasmapheresis and intravenous immunoglobulin were administered to manage catastrophic APS or refractory hemolysis [5]. The case reported by Osmanagaoglu et al. required therapeutic abortion due to the severity of maternal complications, highlighting the need for individualized obstetric decision-making in these high-risk pregnancies [4].

The present case was treated with methylprednisolone for SLE flare and heparin for APS, resulting in significant clinical improvement, which mirrors the treatment success observed in the reviewed cases (Table 1).

Outcomes among reported cases varied based on disease severity and timing of intervention. While most patients responded well to treatment, the case reported by Osmanagaoglu et al. resulted in maternal mortality, particularly due to multi-organ failure and delayed diagnosis [4]. Other cases reported full

recovery following corticosteroid therapy and anticoagulation [5,6]. The case reported by Appenzeller et al. demonstrated partial resolution of symptoms but continued to require long-term monitoring due to APS-related complications [3].

Early intervention played a crucial role in improving prognosis, as seen in cases that received timely corticosteroid and anticoagulation therapy. The present case, diagnosed early and treated promptly, achieved clinical stabilization, underscoring the critical role of timely diagnosis and multidisciplinary management in improving maternal outcomes.

4. Conclusion

The coexistence of SLE flare, HELLP syndrome, and APS during pregnancy is a rare and complex condition that requires careful evaluation. Early recognition and appropriate management are crucial for achieving favorable outcomes.

Declarations

Conflicts of interest: The authors declare no conflicts of interest.

Ethical approval: Not applicable.

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Use of AI: ChatGPT-4 was used to assist in language editing and improving the clarity of the manuscript. All content was reviewed and verified by the authors. Authors are fully responsible for the entire content of their manuscript.

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