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PREGNANCY COMPLICATED BY SYSTEMIC LUPUS ERYTHEMATOSUS, AUTOIMMUNE HEPATITIS AND PORTAL HYPERTENSION

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ABSTRACT

Introduction: Pregnancy in women with concurrent systemic lupus erythematosus (SLE), autoimmune hepatitis and portal hypertension is exceptionally rare, with fewer than 10 reported cases of any two conditions together and none describing the full triad with detailed haemostatic assessment.

Case description: A 26-year-old woman at 9–10 weeks of gestation presented with abnormal liver enzymes, thrombocytopenia, and later developed subacute cutaneous SLE. Autoimmune evaluation revealed positive anti-double-stranded deoxyribonucleic acid (45 IU/ml), low complement C3 (0.68 g/l), and elevated serum IgG (18.2 g/l) supporting a diagnosis of SLE and probable autoimmune hepatitis. Doppler ultrasound confirmed compensated portal hypertension with splenomegaly. Multidisciplinary management included methylprednisolone, hydroxychloroquine, and low-dose aspirin. The platelet count remained stable (85–105 × 10^9/l) throughout pregnancy. Second-trimester endoscopy showed grade I varices without red signs. Planned caesarean section was performed at 34–35 weeks after foetal lung maturation, delivering a female neonate weighing 2,270 g with Apgar scores 8/9. Maternal and neonatal outcomes were favourable.

Conclusion: Favourable outcomes are achievable despite the concurrent burden of SLE, autoimmune hepatitis (AIH) and portal hypertension in pregnancy. Key points include interpreting thrombocytopenia as a multifactorial sign, balancing bleeding and thrombosis risks, and delivering individualised multidisciplinary care.

KEYWORDS

Systemic lupus erythematosus, autoimmune hepatitis, portal hypertension, pregnancy, case report

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LEARNING POINTS

- Pregnancy in women with concurrent systemic lupus erythematosus (SLE), autoimmune hepatitis and portal hypertension is rare but can achieve favourable maternal and neonatal outcomes with individualized multidisciplinary care.
- Thrombocytopenia in this setting is often multifactorial (hypersplenism, immune-mediated, pregnancy-related). A stable platelet count above 50 $\times 10^9$/l without bleeding may allow expectant management rather than intervention.
- The bleeding–thrombosis paradox requires balancing prothrombotic tendencies (pregnancy, SLE) against bleeding risks (portal hypertension, varices, and thrombocytopenia). Low-dose aspirin may be used cautiously; low molecular weight heparin is not mandatory in antiphospholipid antibody-negative patients with significant bleeding risk.

INTRODUCTION

Pregnancy in women with systemic lupus erythematosus (SLE), autoimmune hepatitis (AIH) and portal hypertension is rare and carries high maternal–foetal risk[1,2]. Single-disease guidelines do not address the combined scenario[3,4]. The convergence of immune activation, hepatic inflammation, portal haemodynamic stress and haemostatic instability creates a diagnostic and therapeutic challenge[5]. This case illustrates the bleeding–thrombosis paradox and demonstrates that favourable outcomes are achievable with multidisciplinary individualised care[5-7].

CASE DESCRIPTION

A 26-year-old woman presented to our tertiary centre at 9–10 weeks of gestation with incidentally detected abnormal liver enzymes during routine antenatal screening. She had no prior diagnosis of liver disease, autoimmune disorder or portal hypertension. Her medical history was unremarkable. There was no history of thrombosis, bleeding, prior pregnancy loss or adverse obstetric outcomes. Family history was negative. She reported no alcohol use, hepatotoxic drugs or herbal supplements.

At presentation, she was asymptomatic from a hepatic perspective, with no jaundice, pruritus, abdominal pain or bleeding manifestations. Physical examination was unremarkable except for mild fatigue and later-developing cutaneous erythematous lesions on the face and upper trunk, consistent with subacute cutaneous lupus erythematosus.

Initial laboratory evaluation revealed elevated alanine aminotransferase (122 U/l; reference range 10–40 U/l), aspartate aminotransferase (98 U/l; reference range 10–35 U/l), and gamma-glutamyl transferase (85 U/l; reference range 5–35 U/l). Total bilirubin, albumin and international normalized ratio were within normal limits. Complete blood count showed a platelet count of 98 $\times 10^9$/l (reference range 150–400 $\times 10^9$/l), with normal hemoglobin and white blood cell count.

Viral hepatitis serology and Epstein–Barr virus, cytomegalovirus was negative. Autoimmune screen revealed positive antinuclear antibodies (titre 1:320, speckled pattern), positive anti-double-stranded deoxyribonucleic acid (dsDNA) antibodies (45 IU/ml; reference range <25 IU/ml), low complement C3 (0.68 g/l; reference range 0.9–1.8 g/l) and normal C4. Antiphospholipid antibodies were negative.

Serial autoimmune serology confirmed persistent disease activity: anti-dsDNA antibodies ranged from 44.0 to 58.49 IU/ml (reference <25 IU/ml) across multiple time points, and antinuclear antibodies remained positive (0.6–0.72,

Parameter	24.10.2023	27.11.2023	21.12.2023	27.02.2024	Reference range
ANA (index)	0.72	not tested	0.6	0.7	<0.9 (negative)
Anti-ds-DNA (IU/ml)	55	58.11	44	58.49	<25
Anti-SS-A (U/ml)	14.4	not tested	10	not tested	<7
Anti-cardiolipin (IgG/IgM)	3.8/5.9	not tested	not tested	4.74	<10
Anti-β2-GP (IgG/IgM)	4.02/5.6	not tested	not tested	4.2	<7

Table 1. *Serial autoimmune serology during pregnancy and follow-up.*

Abbreviations: ANA, antinuclear antibodies; anti-ds-DNA, anti-double-stranded deoxyribonucleic acid; anti-SS-A, anti-Sjögren's-syndrome-related antigen A; anti-β2-GP, anti-beta-2-glycoprotein.

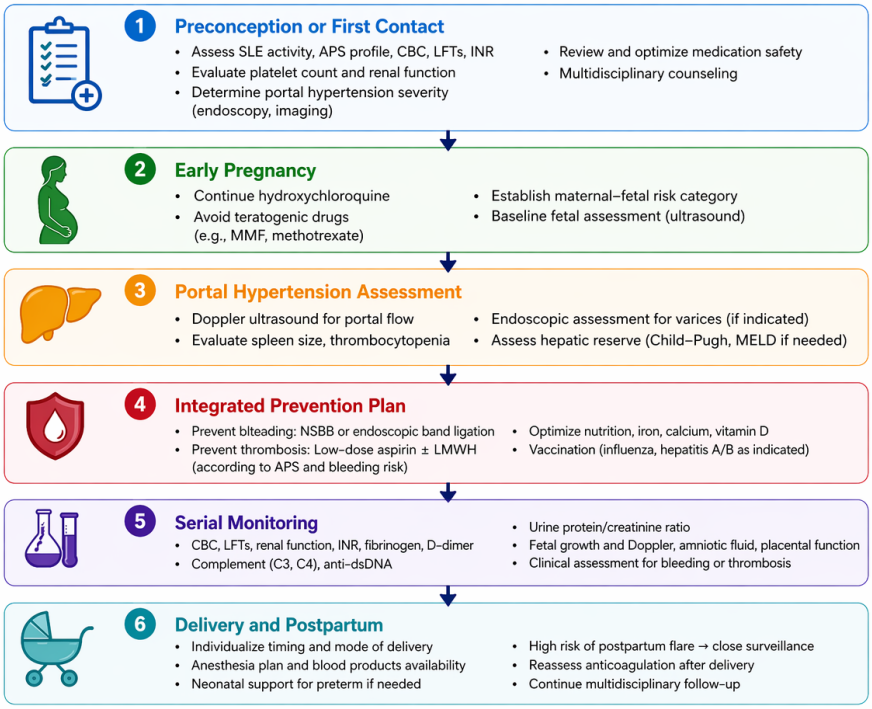


Figure 1. *Multidisciplinary management algorithm for pregnancy complicated by systemic lupus erythematosus, autoimmune hepatitis and portal hypertension. The algorithm outlines six sequential phases: preconception or first contact assessment, early pregnancy evaluation, portal hypertension risk stratification, integrated prevention plan (bleeding and thrombosis), serial maternal–foetal monitoring, and delivery with postpartum follow-up.*

Abbreviations: SLE, systemic lupus erythematosus; AIH, autoimmune hepatitis; LMWH, low molecular weight heparin; ANA, antinuclear antibodies; anti-dsDNA, anti-double-stranded deoxyribonucleic acid; aPL, ant phospholipid antibodies.

index value). Anti-SS-A antibodies were positive at 14.4 U/ml. Antiphospholipid antibodies, including anti-cardiolipin IgG/IgM and anti-β2-glycoprotein IgG/IgM, were repeatedly negative, which informed the decision to withhold low molecular weight heparin despite the prothrombotic state of pregnancy and SLE *(Table 1)*.

Serum immunoglobulin G (IgG) level was elevated at 18.2 g/l (reference range 7–16 g/l), supporting an autoimmune-mediated hepatic process in the absence of liver biopsy.

Abdominal ultrasound showed hepatomegaly with increased echogenicity, splenomegaly (span 14 cm), and a patent portal vein with preserved flow. Doppler ultrasound confirmed compensated portal hypertension (portal vein velocity 12 cm/s; normal >15 cm/s) with no ascites or collateral vessels *(Fig. 1)*.

The patient satisfied the 2019 European Alliance of Associations for Rheumatology (EULAR)/ American College of Rheumatology (ACR) classification criteria for SLE with a cumulative score of 14 points: positive antinuclear antibodies (1:320, speckled) as entry criterion, anti-dsDNA antibodies 45 IU/ml (6 points), low complement C3 0.68 g/l (4 points), and subacute cutaneous lupus (4 points). AIH was diagnosed as probable based on the simplified AIH diagnostic score in the absence of liver biopsy (deferred during pregnancy). Supportive findings included elevated serum IgG (18.2 g/l), positive antinuclear antibodies (ANA), transaminase elevation (alanine aminotransferase 122 U/l, aspartate amintransferase 98 U/l), and exclusion of viral hepatitis, drugs, and alcohol.

The presence of splenomegaly, thrombocytopenia, and Doppler findings (portal vein velocity 12 cm/s) confirmed compensated portal hypertension, most likely on a background of non-cirrhotic portal hypertension related to autoimmune and vascular mechanisms.

Methylprednisolone was initiated at 32 mg/day orally, tapered gradually to 8 mg/day over eight weeks. Hydroxychloroquine 200 mg twice daily was started and continued throughout pregnancy. Low-dose aspirin 100 mg once daily was prescribed for preeclampsia prevention, with careful monitoring. Iron supplementation was given for mild anaemia. No low molecular weight heparin was used because antiphospholipid antibodies (anti-cardiolipin and anti-β2-glycoprotein) were repeatedly negative on serial testing, and there was no prior history of thrombosis. Thrombocytopenia and portal hypertension further increased bleeding risk, making anticoagulation unwarranted in this specific context.

Upper gastrointestinal endoscopy performed in the second trimester (at 22 weeks) showed small non-bleeding oesophageal varices (grade I) with no red signs and no gastric varices. Endoscopic band ligation was not indicated.

Liver enzymes normalised by 16 weeks of gestation. Platelet count remained stable between 85 and 105 $\times 10^9$/l throughout pregnancy. Cutaneous lesions improved. Serial foetal ultrasound showed appropriate growth with no evidence of intrauterine growth restriction.

Planned caesarean delivery was performed at 34–35 weeks after foetal lung maturation. Estimated blood loss was 500 ml. A female neonate weighed 2,270 g (appropriate for gestational age) with Apgar scores 8 at 1 minute and 9 at 5 minutes. No neonatal complications occurred. Postpartum, the patient remained stable. Hydroxychloroquine was continued; methylprednisolone was tapered to 4 mg/day by 6 weeks postpartum. One year after delivery, the patient remained in remission with normal liver enzymes, platelet count 112 $\times 10^9$/l, and no thrombotic or bleeding events.

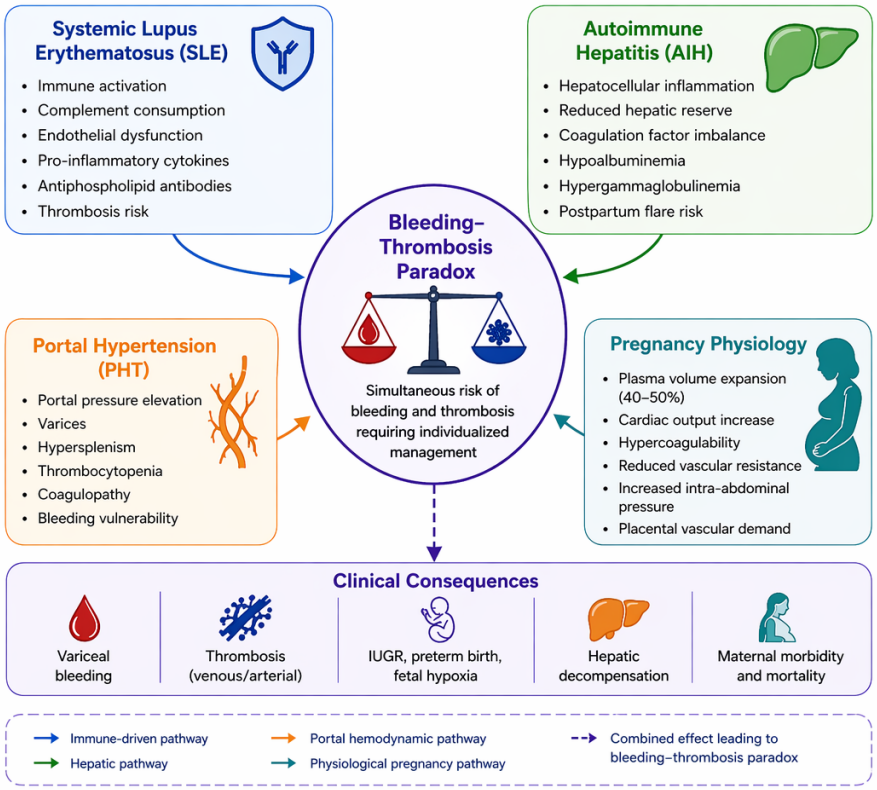


Figure 2. *Immune–hepatic–haemostatic axis and the bleeding–thrombosis paradox in pregnancy with systemic lupus erythematosus, autoimmune hepatitis and portal hypertension. The diagram integrates four contributing pathways: SLE (immune activation, complement consumption, endothelial dysfunction — blue arrow), AIH (hepatocellular inflammation, reduced hepatic reserve — green arrow), portal hypertension (portal pressure elevation, varices, hypersplenism, thrombocytopenia — orange arrow), and physiological pregnancy adaptations (plasma volume expansion, hypercoagulability — purple arrow). The convergence produces a paradoxical clinical state with simultaneous risk of variceal bleeding and thrombosis.*

DISCUSSION

The coexistence of SLE, AIH and portal hypertension in pregnancy is exceptionally rare. To our knowledge, fewer than 10 published cases have described any two of these conditions together, and none have fully characterised the triple combination with detailed haemostatic and haemodynamic assessment during gestation. This case adds clinically relevant information on three interconnected aspects: the diagnostic challenge of overlapping autoimmune and hepatic pathology, the dynamic balance between bleeding and thrombosis risks, and the feasibility of favourable maternal–neonatal outcome with multidisciplinary care.

Although liver biopsy remains the gold standard for AIH diagnosis, it was deferred during pregnancy due to potential procedural risks. In this context, the combination of elevated serum IgG (18.2 g/l), positive ANA, characteristic transaminase elevation, and exclusion of other aetiologies provided sufficient indirect evidence to support a probable AIH diagnosis, consistent with the simplified AIH scoring system.

Pregnancy induces physiological hypervolaemia (plasma volume expansion of approximately 40–50%), increased cardiac output and hypercoagulability[7]. In the presence of portal hypertension, increased splanchnic and portal venous inflow raises variceal wall tension, predisposing to rupture[5,6]. The second and third trimesters, labour and the early postpartum period are highest-risk windows for variceal haemorrhage, as reported in American Association for the Study of Liver Disease (AASLD) guidance[5]. In this case, compensated portal hypertension with grade I varices, no red signs and preserved synthetic function allowed expectant management with planned delivery rather than prophylactic endoscopic band ligation.

Concurrently, SLE contributed endothelial dysfunction, systemic inflammation and antiphospholipid antibody-negative thrombotic risk[1,2]. Low-dose aspirin was used for preeclampsia prevention, guided by the absence of high-risk varices and stable platelet count. Low molecular weight heparin was not given because antiphospholipid antibodies were negative and thrombocytopenia increased bleeding vulnerability. This decision reflects the core dilemma: preventing thrombosis without provoking haemorrhage in a patient with simultaneous prothrombotic (pregnancy, SLE) and prohaemorrhagic (portal hypertension, thrombocytopenia) tendencies[7].

Thrombocytopenia was multifactorial: hypersplenism from portal hypertension contributed to platelet sequestration, while SLE-related immune mechanisms could not be excluded[5,6]. The platelet count remained stable in the 85–105 $\times 10^9$/l range, avoiding the need for platelet transfusion or splenic artery embolization. This pattern suggests that close surveillance without intervention is feasible when counts do not fall below 50 $\times 10^9$/l and there are no bleeding manifestations.

Figure 2 shows the contributing pathways: the immune–hepatic–haemostatic axis and the bleeding–thrombosis paradox in pregnancy with SLE, AIH and portal hypertension.

EULAR and ACR reproductive health recommendations emphasise pregnancy-compatible immunosuppression, continuation of hydroxychloroquine, and disease stability before conception[1,2]. Although conception occurred without prior optimisation, this case demonstrates

that initiating therapy promptly after diagnosis can still achieve remission. European Association for the Study of the Liver guidance on autoimmune hepatitis recommends maintaining immunosuppression during pregnancy to prevent flare[3,4,8], which was followed with methylprednisolone. AASLD guidance on portal hypertension supports risk stratification and individualised bleeding prevention[5]; this case adhered to those principles by performing second-trimester endoscopy and planning delivery in a controlled setting with blood product availability.

This is a single case without histological confirmation of AIH (liver biopsy deferred during pregnancy). Long-term follow-up beyond one year is not available. Generalisability to patients with cirrhosis, active antiphospholipid syndrome or severe thrombocytopenia requires further study.

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