

Haemolytic Anaemia in Pregnancy: A Clinical Conundrum

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Objective

To highlight Evans Syndrome as a rare cause of haemolytic anaemia in pregnancy, and its management.

Differential Diagnoses of Haemolytic Anaemia

CONDITION	CLINICAL FEATURES	BIOCHEMICAL FEATURES
Evans Syndrome	Jaundice, haematuria, epistaxis, melena, bruising, may be asymptomatic.	Raised reticulocytes, raised bilirubin, positive DAT, thrombocytopenia. Features on blood film: polychromasia, macrocytosis, spherocytes
Pre-eclampsia/HELLP	Haemolysis, abdominal pain, headache, proteinuria, visual disturbance, upper abdominal pain, increased blood pressure	Raised transaminases, raised LDH, raised blood pressure, proteinuria, IUGR, raised creatinine, thrombocytopenia. Normal bilirubin.
Haemolytic Uraemic Syndrome	Haematuria, gastroenteritis-like symptoms, bloody diarrhoea, family history. ADAMTS13 activity normal/moderately reduced.	Anaemia, raised bilirubin, raised LDH, marked renal impairment
Thrombotic Thrombocytopenic Purpura	Headaches, migraines, depression, history of IUD or IUGR, reduced ADAMTS13 activity	Anaemia, raised bilirubin, raised LDH, renal impairment
Disseminated Intravascular Coagulation	Haematuria, severe obstetric complications	Thrombocytopenia, abnormal clotting screen

Case Report

A multiparous woman (P1 – NVD) of Albanian descent presented at 34+2 weeks gestation with flu-like symptoms, frank haematuria, abdominal pain, lower limb cramps and reduced fetal movements. She appeared clinically jaundiced, with a normal blood pressure, and no proteinuria. The cardiotocography identified a fetal bradycardia, and therefore a Grade 1 Caesarean Section was performed under General Anaesthesia (EBL 250mL).

Pre-Operative Blood Results	
Hb	83 (116 at 28/40)
WCC	5.02
Plt	157
LDH	865
Bili	78
Cr	64
ALT	62
Blood Film	Left shift of leucocytes

Opinions were sought from our medical, haematology, and rheumatology colleagues. Further investigations agreed by the MDT including Hepatitis, CMV, EBV and HIV serology were all unremarkable. COVID antibodies were detected, with negative COVID-19 PCR. The Coomb’s test was positive, suggestive of auto-immune haemolytic anaemia. The auto-antibody screen was negative. An abdominal ultrasound identified no abnormalities of the liver or urinary tract. A diagnosis of Evans Syndrome was made.

Discussion

Evans syndrome in pregnancy is rare, with only a small number of cases published¹. It is described as the coexistence autoimmune haemolytic anaemia and immune thrombocytopenia¹; neutropenia is not common in pregnancy¹. Women often present with epistaxis, haematuria, bruising, and fever¹. Laboratory findings include raised reticulocyte count and unconjugated bilirubin, with a positive indirect Coombs test¹. The management during pregnancy aims to ameliorate the haemoglobin and platelet counts, and may include steroids and IV Gammaglobulin, with Azathioprine or splenectomy being considered in persistent cases¹. The fetus may be affected due to the transplacental passage of antibodies, resulting in fetal anaemia and thrombocytopenia; antenatal ultrasound monitoring for this is advised. Good maternal clinical outcome are often seen, with the condition resolving following delivery¹.

Other differential diagnoses included partial HELLP Syndrome, Haemolytic Uraemic Syndrome and Thrombotic Thrombocytopenic Purpura.

Conclusion

Evans syndrome is a rare cause of haemolytic anaemia in pregnancy, characterised by the co-existence of immune haemolytic anaemia and thrombocytopenia. It may be diagnosed with a full blood count, blood film and Coomb’s test.

References

Lefkou E, Nelson-Piercy C, Hunt BJ (2009) Evans Syndrome in pregnancy: a systematic literature review and two new cases. European Journal of Obstetrics & Gynaecology and Reproductive Biology. 149 (1): 10-17.