



Primary Adrenal Insufficiency in Undiagnosed Antiphospholipid Syndrome: A Postpartum Diagnostic Dilemma

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Objective

Primary adrenal insufficiency (PAI) secondary to bilateral ischaemic adrenal infarction in the context of undiagnosed antiphospholipid syndrome (APS) in the postpartum period is uncommon, but important. This case adds to existing literature by highlighting potential aberrancies in the clinical course.

Case Introduction

A South-Indian 40-year-old female presented 10-days postpartum with epigastric pain and hypertensive urgency. Antenatal issues necessitating caesarean section at 33+4 weeks included preeclampsia, intrauterine growth restriction and gestational diabetes. The male infant, weighing 1467grams (6th centile) with APGARs 8 (1minutes) and 9 (5minutes), had a prolonged admission for necrotising enterocolitis. Acute abdominal/aortic pathology was excluded on CT aortogram and ultrasound. Cholelithiasis was noted.

Abdominal pain improved with pantoprazole and new antihypertensive treatment including hydralazine, labetalol and nifedipine. She was discharged after 3 days but re-presented that night with migrating epigastric, back and thoracic pain with nausea and vomiting.

Case Progress

She was hypertensive, dry and lethargic. Hyponatraemia, cholestatic liver derangement and hypoalbuminaemia were identified. Subsequent reduction in blood pressure, cessation of antihypertensive medications and hyperpigmentation prompted cortisol, thyroid and autoimmune screening, identifying severe hypocortisolaemia (12nmol/L). Sheehan syndrome was excluded on neuroimaging. Short synacthen test had no response, confirming PAI. Hydrocortisone was commenced. CTPA excluded pulmonary embolism for new interscapular pain but revealed interval enlargement of the left adrenal gland. Abdominal CT confirmed bilateral adrenal gland enlargement with ischaemic changes.

Recurring abdominal pain, new haemoglobin decrease and rising inflammatory markers developed. Haemolytic screen was negative. Magnetic resonance cholangiopancreatography excluded hepatobiliary pathology but demonstrated bilateral adrenal infarction and a non-occlusive right renal vein thrombosis. She was biochemically coagulopathic. Investigations showed a markedly positive anti-B2-glycoprotein, anti-cardiolipin and lupus coagulant antibody. Subsequently, placenta histopathology showed infarcts with thrombi.

Worsening hypoalbuminaemia and proteinuria (6.15g/day) continued until anticoagulation was commenced. Peripheral serology for other causes of glomerulonephritis were negative. Anti-coagulation was commenced with coumadin, aiming for an international normalised ratio between 2.5-3.5 with ongoing oral hydrocortisone supplementation.

Discussion

PAI secondary to thrombotic APS is a rare, potentially life-threatening entity that requires a high index of suspicion. Multiple aspects complicated diagnosis. Firstly, PAI typically results in hyponatraemia and hyperkalaemia but our patient was hypokalaemic. Secondly, initial hypertension was unexpected but may be due to overlap of preeclampsia in the puerperium. The combination of hypotension after preeclampsia, non-improving proteinuria and abdominal pain impacted prompt diagnosis. These symptoms improved once anticoagulation was commenced.

Conclusion

This case highlights the importance of re-evaluation in an evolving diagnostic dilemma.

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