

A challenging case of primary aldosteronism presenting in pregnancy

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INTRODUCTION

- Primary aldosteronism (PA) is the most common form of secondary hypertension.
- PA is uncommonly diagnosed during pregnancy¹.
- Uncontrolled blood pressure in pregnancy can result in adverse outcomes for both the mother and fetus¹⁻⁴.
- Here we report a case presenting in pregnancy whose management proved particular challenging, due to variable compliance with therapy, frequent non-attendance at clinic, a subsequent pregnancy and negative imaging.

CASE

- A 24 year old woman presented at 26 weeks gestation with severe pre-eclampsia, hypokalaemia (2.1mmol/l) and a metabolic alkalosis.
- She received intravenous potassium replacement, labetalol and nifedipine
- She had to undergo an emergency caesarean section 3 days after admission.
- Investigation postnatally revealed a significantly raised aldosterone/renin ratio at >12250 (aldosterone 2450 pmol/L, plasma renin activity<0.2 pmol/mL/h).
- High dose oral potassium replacement was commenced alongside ongoing intravenous potassium.
- Plasma aldosterone remained elevated (1580pmol/l) following intravenous saline loading, confirming a diagnosis of PA. Spironolactone was commenced.
- CT and MRI scans showed normal adrenals.
- She remained hypokalaemic and hypertensive despite being prescribed spironolactone and oral potassium supplements; compliance was poor.
- She did not attend for further planned investigations and missed some clinic appointments.
- Twelve months after initial presentation she became pregnant again. Spironolactone was stopped.
- She struggled with her potassium supplements due to vomiting and remained hypokalaemic. She was subsequently treated with amiloride and labetalol during pregnancy and delivered a healthy baby boy at nearly 34 weeks gestation following elective caesarean section.
- Postpartum, poor treatment compliance and missed visits to clinic continued.
- A dexamethasone suppression test excluded glucocorticoid remediable aldosteronism.
- Repeat adrenal imaging (MRI) was normal.
- Adrenal vein sampling (AVS) indicated right adrenal aldosterone excess.
- Right adrenalectomy was performed and a small adenoma identified.
- She has remained normotensive and normokalaemic off treatment postoperatively.

INVESTIGATIONS

INVESTIGATION	RESULT
Aldosterone/renin ratio	>12250
Aldosterone level	2450 pmol/L
Plasma renin activity	<0.2 pmol/ml/h
Plasma aldosterone post saline loading	1580 pmol/L
CT	Normal adrenals
MRI	Normal adrenals
Low dose dexamethasone suppression test	Negative Pre-Dexamethasone Aldosterone- 799 pmol/L Post-Dexamethasone Aldosterone 547 pmol/L
Adrenal vein sampling	Right adrenal aldosterone excess (right adrenal vein [RAV] aldosterone: cortisol ratio 245.4, left adrenal vein [LAV] aldosterone: cortisol ratio 3.6, Lateralisation Index RAV: LAV 68.2)
Right adrenalectomy & histology	Adenoma

DISCUSSION

- There are less than 50 case reports in the literature describing PA in pregnancy¹⁻³.
- Spironolactone is commonly used as a first line treatment for PA in non-pregnant patients but is not recommended during pregnancy, due to potential anti-androgenic effects causing ambiguous genitalia in males¹⁻³.
- The optimal management of PA during pregnancy is unclear. Initial pharmacological management should include antihypertensive drugs approved for pregnancy, along with potassium supplements. Eplerenone or amiloride can be considered as therapeutic options, although more evidence is needed regarding their safety and efficacy in pregnancy¹⁻⁵.
- PA increases the risk of maternal and fetal morbidity and mortality, including preterm delivery, placental abruption, intrauterine growth retardation, end-organ damage and stillbirth¹⁻⁴. In our case, her first child had multiple medical issues.
- The course of PA in pregnancy is variable, possibly in part due to the anti-mineralocorticoid effect of progesterone¹⁻³.
- The aim of management is to control blood pressure and normalise potassium levels.
- For aldosterone-producing adenomas, laparoscopic adrenalectomy may occasionally need to be considered as a therapeutic option in the second trimester of pregnancy. Ideally, if pregnancy is planned, adrenalectomy should be performed pre-conception^{2,3}.
- This case also confirms the importance of AVS which identified unilateral aldosterone production despite normal imaging and enabled cure of her PA.

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