Phaeochromocytoma: A Rare Cause of Hypertension in Pregnancy

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Introduction

Hypertension is common (8% to 10% incidence) in pregnancy and has well-defined management guidelines [1]. Phaeochromocytomas are a **rare** (1 in 54,000 pregnancies) cause of **severe** hypertension associated with significant morbidity and mortality if missed [2]. This case report highlights the diagnosis and subsequent clinical management of a case at our tertiary obstetric unit.

Case Discussion

A 34-year-old G4P1, presented at 33 weeks gestation with incidental severe hypertension (195/120 mmHg) at routine antenatal review. Gestational diabetes in her previous pregnancy was controlled with metformin, but required insulin therapy in this pregnancy. She was admitted and commenced on labetalol and then nifedipine as per local guidelines, but remained hypertensive despite maximal therapy. She was otherwise asymptomatic.

Pre-eclampsia tests (protein-creatinine ratio and placental growth factor) were negative, so a renal ultrasound scan was performed. This showed a retroperitoneal lesion, with MRI confirming a 5x5x6cm right adrenal mass. Paired with her clinical features this was suggestive of phaeochromocytoma. Normetadrenaline levels were 16 times greater than normal, confirming the diagnosis.

Blood pressure was controlled with doxazosin, bisoprolol and nifedipine and she was transferred to our centre. There was a significant reduction in insulin requirements, triggering a review for signs of placental failure. Growth scans were normal so it was felt to be an effect of alpha and beta blockade antagonizing the effects of catecholamines on insulin production and resistance.

The MDT decision was to proceed with elective caesarean section at 37 weeks.

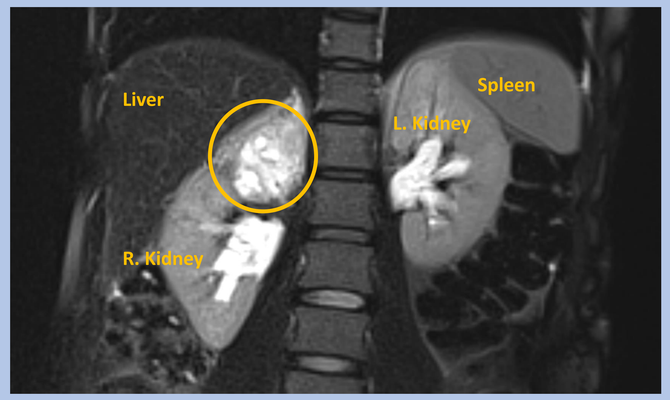


Figure 1: T2 MRI Adrenal Scan showing a markedly hyperintense right adrenal gland (Phaeochromocytoma, Highlighted)

After arterial line insertion, combined spinal epidural anaesthesia was complicated by accidental dural puncture and further attempts by paresthesia on threading the catheter. Single shot spinal anaesthesia produced adequate anaesthesia without major haemodynamic consequences.

Blood pressure was maintained with a metaraminol infusion. Caesarean section was uneventful and forceps were used electively to deliver the head with the aim to avoid tumour compression secondary to fundal pressure.

The patient remained hypertensive post-delivery until switched from doxazosin to phenoxybenzamine. Post dural puncture headache required an epidural blood patch 5 days later. Laparoscopic adrenalectomy was performed nine weeks post-partum and the tumour was confirmed histologically as a phaeochromocytoma.

Conclusions

Phaeochromocytoma is an important differential in any pregnant woman with **atypical severe hypertension** in pregnancy. Misdiagnosis as pre-eclampsia can easily occur with disastrous results, with deaths from undiagnosed cases featuring in MBRRACE reports [2].

Doxazosin has a lower feto-maternal ratio than phenoxybenzamine so may be the preferred option for alpha blockade in pregnancy [3].

Planned caesarean section is preferable to allowing labour with its potential for unplanned emergency interventions.

Anaesthetic considerations for delivery include avoiding hypertensive crises from catecholamine release (can be triggered by direct tumour compression or by drugs such as ephedrine, suxamethonium, morphine and anticholinergic agents) and the potential for uncontrolled hypotension resistant to vasopressors from the combination of regional anaesthesia and alpha and beta blockade.

Key Points

- Phaeochromocytoma may present with atypical severe hypertension and abnormal glucose tolerance.
- Doxazocin has a better fetal side effect profile than phenoxybenzamine.
- Planned delivery via caesarean is preferable to allowing labour, although tumours may be resected in early pregnancy.
- Avoid tumour compression from fundal pressure at delivery and be aware of the drugs that can trigger catecholamine release.
- Anticipate resistance to vasopressors in the alpha blocked patient.

References:

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