Pheochromocytoma in pregnancy: a case report
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ABSTRACT
A case of large 11cm pheochromocytoma at 35 weeks with preceding diagnoses of pre-eclampsia and gestational diabetes (GDM), which confounded initial management.

Pheochromocytoma in pregnancy is rare, with an incidence of 0.007%, but carries high mortality of 40–50% if untreated. Our case highlights the diagnostic challenges and complexity of management.

A 24-year-old primigravida presented at 33+2 weeks with symptoms of anxiety, nausea and vomiting. Her blood pressure was 206/90mmHg. She had persistent sinus tachycardia but an otherwise normal examination. The bloods were unremarkable, but proteinuria was evident (protein creatinine ratio 134.9mg/g). A large for gestation fetus was identified (>95th percentile, estimated fetal weight 3082g). OGTT at 34 weeks confirmed gestational diabetes (GDM).

Pre-eclampsia was diagnosed and oral labetalol commenced. Blood pressure on the ward was labile and her sinus tachycardia persisted. Once her symptoms and hypertension settled, she was discharged on oral labetalol.

At 35 weeks she re-presented with intermittent headaches, ongoing anxiety, a systolic pressure of 210mmHg (the diastolic was not recorded) and ongoing maternal tachycardia, which prompted further investigation. IV hydralazine and magnesium sulphate were commenced for acute hypertensive control and eclampsia prevention.

She was found to have paroxysmal hypertension and no excessive weight gain despite diagnosis of GDM and pre-eclampsia on assessment by the endocrinologist.

On investigation, plasma normetanephrines were raised (58,153pg/mL) and she had newly deranged liver function tests but no other evidence of end organ damage. Ultrasound revealed a 9x8x11cm right suprarenal mass.

Upon suspecting pheochromocytoma, phentolamine and magnesium infusion was commenced and the patient was admitted to ICU. Initially, the plan was made for phentolamine for seven days prior to surgery. However, the next day, following a multi-disciplinary team (MDT) discussion including anaesthetics, obstetrics, endocrinology and obstetric medicine, a lower segment caesarean was performed under general anaesthesia at 36+6 weeks with ongoing IV phentolamine infusion and magnesium. The indication was suboptimal control of blood pressure in pregnancy and risk of severe morbidity and mortality to both mother and fetus. Efforts were made to minimise pressure exerted on the tumour at delivery. There were no episodes of labile blood pressure intraoperatively, and a well male infant of 3,395gm was delivered by forceps via hysterotomy without fundal pressure.

Post-partum MRI revealed imaging consistent with a pheochromocytoma. The patient underwent successful transperitoneal laparoscopic excision of adrenal tumour two months later and T2Nx pheochromocytoma was confirmed. Family history and genetic testing for common susceptibility genes were negative and she is receiving ongoing endocrinology follow-up.

Diagnosis of pheochromocytoma can be difficult in pregnancy. More common causes of hypertension, such as pre-existing hypertension and pregnancy-related hypertension, are difficult to differentiate from rare causes such as pheochromocytoma. Labile, paroxysmal, difficult-to-control blood pressure associated with additional features, including palpitation/tachycardia, flushing, sweating and light-headedness, should
prompt further investigation. Fifty percent experience sustained hypertension, 35–45% paroxysmal hypertension and up to 50% are found to have orthostatic hypotension. Presence of proteinuria does not exclude the diagnosis and is not uncommon, as an altered renal function can be attributed to catecholamine-mediated renovascular changes found to reverse after tumour removal. GDM is also common and reported in a third of cases. This may be explained by altered glucose metabolism by inhibition of insulin secretion or induced insulin resistance.

Twenty-four-hour catecholamine urine collection is recommended, as pregnancy does not elevate urinary catecholamine levels within the diagnostic range for phaeochromocytoma. Plasma metanephrines can be considered, but specificity is low (85–89%). False positives are reported with concurrent use of antihypertensives or incorrect collection method. MRI is advised over USS as the gravid uterus can make visualisation difficult.

The primary goal is to prevent complication from hypertensive crisis. Although catecholamines do not cross the placenta into the fetal circulation, the paroxysmal hypertension can lead to placental abruption as rebound hypotension causes severe hypoxia and fetal demise. Medical treatment with alpha-blockade should be started as soon as diagnosis is made. Caution should be given before prescribing beta-blockade; if administered alone, it may cause precipitous blood pressure rise due to unopposed alpha-adrenergic effects, as observed in our case. Other commonly used medications, such as oxytocin, methyldopa and anaesthetic agents, should also be avoided. Surgery is the definitive treatment. Successful adrenalectomy has been reported in the second trimester. In cases of later gestation diagnosis, the general consensus is for adrenalectomy after an elective caesarean section (CS). Vaginal delivery is associated with a higher mortality (31%) compared with CS (19%). Vaginal delivery is considered in multiparous mothers who have had rapid, safe deliveries. The evidence for timing of delivery, adrenalectomy and mode of delivery is based on case reports and remains an area of ongoing research.

Our case demonstrates the importance of considering the diagnosis of phaeochromocytoma in the context of resistant hypertension or pre-eclampsia in the third trimester. A careful, individualised approach with MDT collaboration is required.
Competing interests:
Nil.

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REFERENCES