Phaeochromocytoma in Pregnancy
A Dilemma for the Endocrinologist, Endocrine Surgeon, Obstetrician and Anaesthetist

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Introduction

- Phaeochromocytomas are adrenomedullary catecholamine secreting tumours.
- Here we present a challenging case of a phaeochromocytoma diagnosed during pregnancy.
- Given the rarity of this clinical scenario, the management and ethical issues surrounding it are complex and controversial.

History

- A 22-year-old lady presented at 20 weeks gestation with a 4-week history of headaches, palpitations and anxiety.
- From her personal and family history there was nothing else of note.

Clinical Examination

- She was found to be hypertensive (165/107 mmHg), however, her obstetric booking blood pressure had been 100/60 mmHg.
- On suspicion of a phaeochromocytoma, a 24-hour urinary collection showed markedly raised normetadrenaline [52.99 micromols/24hrs (<3)], normal metadrenaline and raised 3-methoxytyramine [6.25 micromols/24hrs (0-2.75)].
- Her corrected serum calcium was 2.58 mmol/L (2.12-2.62) and parathyroid hormone level was <0.3 pmol/L (1.3-7.6). Her calcitonin was <14 ng/L (<15).
- From her personal and family history there was nothing else of note.

Investigations

- Her urea/electrolytes, uric acid levels, autoimmune screen, electrocardiogram and echocardiograph were all normal.
- On suspicion of a phaeochromocytoma, a 24-hour urinary collection showed markedly raised normetadrenaline [6.66 micromols/24hrs (<3.685)], with normal metadrenaline and 3-methoxytyramine.
- Her corrected serum calcium was 2.58 mmol/L (2.12-2.62) and parathyroid hormone level was <0.3 pmol/L (1.3-7.6). Her calcitonin was <14 ng/L (<15).
- Radiological Studies

- MRI of her adrenals showed a 3.2cm high signal mass in the right gland in keeping with a phaeochromocytoma and an intraterine pregnancy (Figs 1 to 3).
- An MRI of her abdomen showed a slightly hypointense mass in the left adrenal (21mm x 14mm in dimension) typical of a left adrenal phaeochromocytoma (Fig 6).

Pre-operative Management

- The lady was adequately alpha-blocked with Phenoxycyzamine and this was followed by beta-blockade with Propranolol, resulting in resolution of her symptoms.
- Following a multidisciplinary discussion, the decision was made to wait till 36-weeks of gestation and to combine delivery via a lower segment caesarean section with removal of the phaeochromocytoma laparoscopically, in the same anaesthetic setting.
- In the following weeks, she was well-controlled with alpha and beta-blockade (Phenoxybenzamine 10mg TDS and Propranolol 60mg TDS), with normal foetal growth.

Surgery

- The combined elective procedures went uneventfully, with a stable patient and a healthy neonate.

Post-operative Investigations

- The histology of the 50x50x25 mm adrenal nodule confirmed a phaeochromocytoma, with MIB-1 proliferation rate of <1% suggesting a benign tumour.
- Genetic testing confirmed von Hippel-Lindau disease (VHL).

Follow-up

- On a six-month follow-up the patient did not complain of any symptoms of adrenergic excess and the baby was healthy.
- Her blood pressure was 100/70 mmHg.
- A24-hour urinary collection showed raised normetadrenaline [6.66 micromols/24hrs (<3.685)], with normal metadrenaline and 3-methoxytyramine.
- The normetadrenaline was raised on two further occasions.

Discussion

- This case posed a dilemma to the multi-disciplinary team, including the Endocrinologists, Endocrine Surgeons, Obstetricians and Anaesthetists.
- The presentation of phaeochromocytoma in pregnancy has been rarely reported and therefore remains a diagnostic and therapeutic challenge.
- Adequate management encompasses early recognition, adrenergic blockade and multidisciplinary management.
- Catecholamine hypersecretion causes hyperensive responses of a variable degree, which are often confused with more common causes of gravid hypertension. Therefore, diagnosis may be overlooked.
- Sudden catecholamine discharge in the mother may be so severe that it may result in pulmonary oedema, myocardial infarction, cerebral haemorrhage, and respiratory distress syndrome.
- The phaeochromocytoma crisis can be caused by anaesthesia, uterine contractions, vigorous foetal movements, mechanical compression by the gravid uterus or medications commonly used during pregnancy.
- The catecholamine excess may cause vasosonstruction of the uteroplacental circulation leading to intraterine foetal retardation, foetal hypoxia, spontaneous abortion or intraterine foetal death.
- The appropriate time of surgical excision is a very difficult decision that has to be balanced between the need to attain foetal maturity and the efficacy of medical treatment in restraining the adrenergic excess.

Reference


Fig 1 Coronal T2 MRI of the abdomen shows an intrauterine pregnancy (arrow).

Fig 2 Coronal T2 MRI of the abdomen shows a large high T2 signal lesion in the right adrenal gland (arrow). Note is made of bilateral hydronephrosis.

Fig 3 Axial T1 MRI of the upper abdomen shows a large low signal T1 lesion in the right adrenal gland (arrow).

Fig 4. Coronal view of an MIBG scan shows a focus of increased MIBG uptake corresponding to the left adrenal (arrow).

Fig 5. Transverse view of an MIBG scan shows a focus of increased MIBG uptake corresponding to the left adrenal (arrow).

Fig 6. Coronal MRI of the abdomen shows a T1 signal lesion in the left adrenal gland (arrow).

MRI head and spine were normal.

The patient is to be seen the outpatients to discuss the post-operative scans.