May 2014

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CASE REPORT

Surgical management of pheochromocytoma in a 13-week pregnant woman

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Accepted 20 April 2014

SUMMARY

A 34-year-old 13-week pregnant woman presented with hypertension refractory to medical therapy and on workup was found to have a right adrenal mass. Due to her persistent increased blood pressure she was advised urinary vanillylmandelic acid (VMA) and its level was raised. MRI of the abdomen showed a well-circumcised lesion in the right adrenal of 3.0×2.5 cm suggestive of pheochromocytoma. The patient was started on antihypertensives including α-blockers and β-blockers and planned for right open adrenalectomy. Intraoperatively, blood pressure was raised up to 180/110 mm Hg on slight manipulation of adrenal gland which was controlled with glyceryl-trinitrate and volatile agents. Postoperatively urinary VMA decreased to normal range and all antihypertensives were gradually stopped. She had uneventful pregnancy and delivered vaginally. This case report highlights the importance of surgical management of pheochromocytoma in second trimester of pregnancy to avoid catastrophic complications later in pregnancy.

BACKGROUND

Pheochromocytoma (PCC) is a neuroendocrine tumour of the adrenal medulla (originating in the chromaffin cells). PCC is a very rare condition in pregnancy and it carries significant risks to the mother as well as to the fetus. Therefore, PCC should be considered as a possible cause of refractory hypertension in pregnancy and appropriate investigations should be carried out. The diagnosis can be established by measuring catecholamines and metanephrines levels in the plasma (blood) or through a 24-hour urine collection for VMA. The tumour is generally localised by MRI during pregnancy. Definitive treatment of PCC is surgical but initially it should be treated medically to control blood pressure with α-adrenergic blockers, followed by β-blockers. In this case report PCC was detected in the second trimester, and managed successfully.

CASE PRESENTATION

A 34-year pregnant woman was referred to our clinic because of hypertension refractory to medical therapy and right adrenal mass. She had two normal deliveries before this pregnancy and had no complications. She was diagnosed to have essential hypertension when non-pregnant and was prescribed ACE inhibitor (enalapril 10 mg twice daily). Calcium channel blocker and β-blocker (amlodipine 10 mg four times a day and atenolol 50 mg twice daily) were later added by the family physician due to refractory hypertension. Meanwhile, the patient became pregnant.

INVESTIGATIONS

Owing to persistent increased blood pressure (BP) she was advised urinary vanillylmandelic acid (VMA) which was raised 30 mg/24 h (normal level 2–7 mg/24 h). Later the family physician advised ultrasound which revealed an adrenal mass. She had an MRI subsequently which confirmed a well-circumscibed lesion in the right adrenal mass of 3.0×2.5 cm suggestive of PCC (figures 1 and 2).

DIFFERENTIAL DIAGNOSIS

Hypertension during pregnancy is usually attributable to pre-eclampsia (pregnancy-induced hypertension) or chronic hypertension but secondary causes of hypertension should be considered in patients refractory to medical therapy. These include renal vascular hypertension, coarctation of the aorta and adrenal causes of hypertension (Cushing syndrome, Conn's syndrome, PCC).

TREATMENT

Soon after these diagnostic tests, the patient developed symptoms including sweating, flushing and chest pain with BP 200/120 mm Hg. Cardiac evaluation was performed including ECG and echocardiogram which revealed no abnormality. The treatment was revised and phenoxybenzamine 10 mg thrice daily was initiated resulting in adequate control of hypertension as well as symptoms. Obstetric examination and ultrasound of the fetus showed a single alive fetus with normal fetal growth parameters. Considering the risk of hypertension during pregnancy and the possibility of aggravation during later stages of pregnancy we decided to excise the mass. Surgical risk and risk to fetus was discussed with the patient. Obstetrician also agreed with the plan and counselled the patient about the possible chances of miscarriage. The patient was planned for adrenalectomy during 13th week of pregnancy.

The patient was admitted for right open adrenalectomy and started on progesterone pessaries 400 mg twice daily. Intraoperatively, BP raised to 180/110 mm Hg on slight manipulation of adrenal gland which was controlled by the anaesthetist with glyceryl-trinitrate and volatile agents included isoflurane and nitrous to control the BP during surgery. Operative time was 160 min with a blood loss of only 200 mL and without any need of transfusion. Postoperatively in the recovery room the

To cite: Memon MA, Aziz W, Abbas F. *BMJ Case Rep* Published online: [*please include* Day Month Year] doi:10.1136/bcr-2013-202838

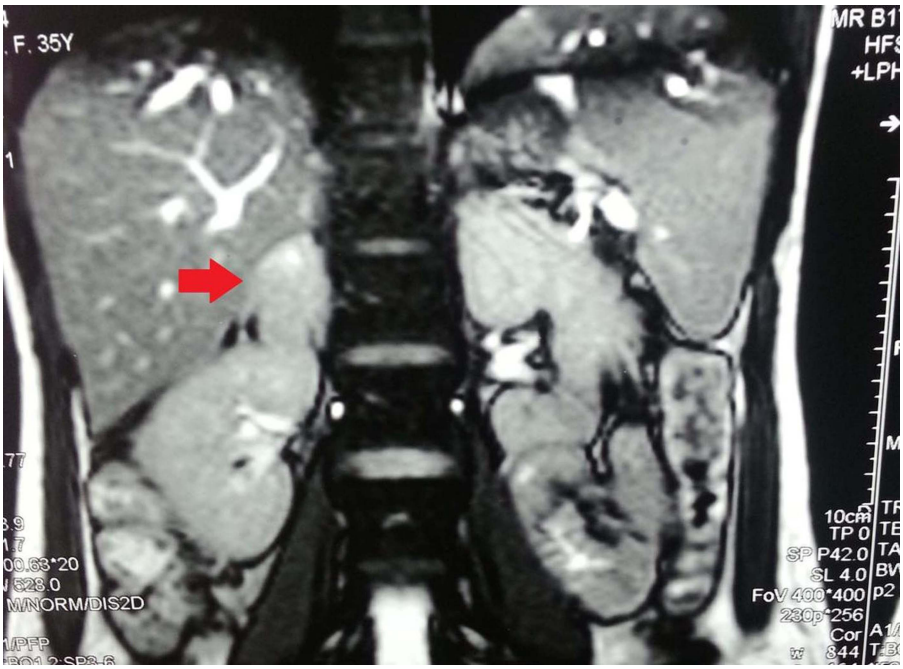

Figure 1 Abdominal MRI (coronal view) showing the right adrenal mass (arrow).

obstetrician performed ultrasound and the fetal heart was checked, which was found normal.

For pain management the anaesthetist started pethidine via patient-controlled intravenous analgesia (PCIA). She was prescribed intravenous paracetamol and morphine after discontinuation of PCIA. The patient was kept in the intensive care unit for labile BP for 2 days which was managed by fluid resuscitation and adjustment of dosage of antihypertensives. The endocrinologist gradually tapered off the antihypertensive medication over 3 days postoperatively. On second postoperative day, slight per vaginal spotting was noticed which settled with observation. The patient was discharged on seventh POD with propranolol 10 mg twice daily and progesterone pessaries 400 mg twice daily.

OUTCOME AND FOLLOW-UP

The patient was followed up in the clinic after 2 weeks, she was fine with normal fetal growth parameters on ultrasound. Urinary VMA decreased to normal range and all antihypertensives were stopped. She was followed up in the obstetric clinic, had safe pregnancy and delivered vaginally at term.

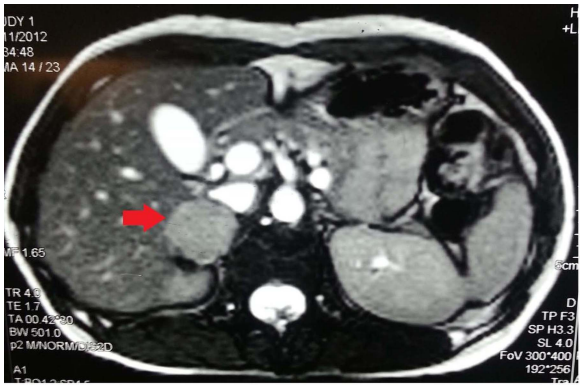

Figure 2 Abdominal MRI (axial view) showing the right adrenal mass (arrow).

DISCUSSION

PCC accounts for 0.1–1% of all cases of hypertension. Precise incidence in pregnancy is difficult to determine but more than 200 cases have been reported in the published literature. PCC is a very rare neuroendocrine tumour which originates from the adrenal medulla (chromaffin cells). This tumour is notorious for its devastating consequences.[1 2] PCC is occasionally referred to as '10% tumors' because 10% are bilateral, 10% are extra-adrenal and 10% are malignant.[2 3] In pregnancy, the presence of PCC may be difficult to detect owing to the more prevalent diagnosis of pregnancy-induced hypertension. It is usually suspected when patient is not responding well to antihypertensives. Untreated PCC carries a risk of mortality for the mother and the fetus, as high as 58%.[4 5] Early diagnosis is vital and symptoms and signs vary which includes: hypertension (98% of cases), orthostatic hypotension, palpitations, tachycardia, headache, sweating, seizure disorders and anxiety attacks. Other symptoms are chest pain, nausea and vomiting, pallor and flushing.[2 6] Pregnancy does not alter urinary catecholamines hence diagnosis is confirmed by 24 h urine VMA, metanephrines or catecholamines. Metanephrines and catecholamines can be measured in the blood as well.[7 8] In adults, approximately 80% of PCC are unilateral and solitary. For localisation ultrasound of the abdomen should be performed as it is easily accessible, cheap and a safe modality in pregnancy and has 89–97% sensitivity. MR has the advantages of greater accuracy, high-quality images and lack of ionising radiation.[9–12] As the complication rate increases with progression of pregnancy, late first trimester and second trimester are the ideal times for surgical treatment after organogenesis is completed. Surgery should be avoided in early first trimester because of high chances of miscarriage and in late second trimester and third trimester because of abdominal exploration and access is difficult.[12 13]

PCC should always be treated first medically to stabilise the BP and symptoms. α-Adrenoceptor blockade that is either phenoxybenzamine or prazosin and β-blockade is used to control tachycardia and dysrhythmia.[8 11 12 14] The aim of this pretreatment is twofold: first, before undergoing surgery, blood

pressure, heart rate and volume depletion should be restored as far as possible. Second, the patient should be protected from the toxic cardiovascular effects of preoperative surges of catecholamines.[1 15] The definitive treatment of PCC is surgical excision either open, laparoscopic or robotic.[4 13] A brief literature review showed that patients with PCC during pregnancy were managed with one of the two approaches. First, medically up to the end of pregnancy and then tumour excision along with C section.[2 16 17] This approach appears to be more suitable for patients who present late in pregnancy and/or adequately manageable with antihypertensives. The second approach is resection of PCC preferably during the second trimester. Robotic adrenalectomy during pregnancy has been reported and the patient had no perioperative complication.[13] Individualised management is appropriate as no single protocol is suitable for all patients given the rarity and the complexity of the problem.

Learning points

- Although pheochromocytoma (PCC) has been treated by controlling blood pressure in the pregnancy, surgical treatment in the second trimester is preferable.
- Patients presenting early in the second trimester are best candidates owing to low risk for maternal–fetal death during this period compared with the first or third trimester.
- Management of PCC should be multidisciplinary in which the endocrinologist, obstetrician, urologist and the anaesthesiologist should be included to minimise the probabilities of complications.
- Strict blood pressure control perioperatively is the key to avoid complications.
- PCC must be suspected as a cause of refractory hypertension in pregnancy.

Acknowledgements The authors thank Ms Munira Amin for her help with literature search.

Contributors MAM and WA drafted the manuscript and performed literature search. FA is the consultant surgeon responsible for patient management.

Competing interests None.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

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