Rare disease

Cardiac pheochromocytoma presenting during pregnancy

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Summary

A 25-year-old woman presented at 12 weeks gestation, with symptoms and laboratory investigations consistent with pheochromocytoma. Imaging modalities available during pregnancy were limited and MRI scan of the abdomen and the neck failed to localise the tumour. Postpartum imaging, including 131-metaiodobenzylguanidine and octreotide scans, cardiac CT, cardiac MRI and cardiac catheterisation, allowed accurate localisation of the tumour and helped plan for successful surgical removal.

BACKGROUND

Cardiac pheochromocytoma is a rare condition, and to our knowledge this is the first time it has been reported in pregnancy. Although imaging modalities available during pregnancy are limited, making the diagnosis difficult, a possible

cardiac location needs to be considered. In contrast, the imaging modalities available post-partum provide a wealth of information allowing accurate tumour localisation and surgical planning.

Figure 1 Short-axis MRI image with delayed enhancement technique: the tumour is located anterior inferiorly behind the sternum just above the liver (arrow); there is delayed contrast enhancement at the core of the tumour.

Figure 2 CT scan images without contrast (A) and with contrast (B–D). (A) At tumour level: the tumour can not be distinguished from the normal heart; (B) above tumour level: multiple arteries (accolade) are present adjacent to the origin of the right coronary artery (RCA) (arrow). (C) At tumour level: tangle of vessels inside tumour (arrowhead), with the RCA located in the lateral wall of the tumour (arrow). (D) Below tumour level: a enlarged cardiac vein (arrow head) is seen adjacent to the distal RCA (arrow).

CASE PRESENTATION

A 25-year-old woman, G2P1, presented at 12 weeks gestation after experiencing episodic bouts of diaphoresis, palpitations, headaches and light headedness. Her systolic blood pressure was 190/145 mm Hg, and her heart rate was 226 bpm. Her past medical history and family history were unremarkable. A 24-h urine collection, collected at 14 weeks gestation, demonstrated a dramatically elevated excretion of norepinephrine at 14 969 nmol/24 h (normal non-pregnant range <500) and normetanephrine at 18.4 μ mol/24 h (normal non-pregnant range <3.3). A presumptive diagnosis of pheochromocytoma was made and medical therapy with α -blockade (phenoxybenzamine) and β -blockade (metoprolol) was initiated.

INVESTIGATIONS

An MRI scan of the abdomen and neck failed to localise a tumour. A caesarean section was performed at 34 weeks gestation without complications, and a healthy daughter, weighing 2166 g, was born.

Postpartum, further imaging studies were able to be performed to localise the source of the excessive catecholamine secretion. An octreotide and a ¹³¹-metaiodo-

benzylguanidine (MIBG) scan both showed uptake in the right lower retrosternal area. A dedicated cardiac MRI (figure 1) and cardiac CT (figure 2) showed a highly vascular tumour located in the right atrioventricular (AV) groove in the RCA (right coronary artery) territory, closely attached to the myocardium; this was confirmed on cardiac catheterisation (figure 3).

TREATMENT

The patient was taken to the operating room and intraoperatively; each time the tumour was touched (even delicately) the heart went into supraventricular tachycardia. These episodes responded to intravenous esmolol boluses. The tumour was found to be in the layer between the epicardium and the myocardium, without myocardial invasion. It extended from the right AV groove at the tricuspid valve to one-third of the way down the anterior wall of the right ventricle and did not involve the tricuspid valve. The entire tumour was resected successfully, with reconstruction of the right atrial and ventricular walls with bovine pericardium as well as tricuspid valve repair and CABG of the RCA (native RCA was sacrificed during tumour removal). Pathological examination confirmed a paraganglioma.

Figure 3 Coronary angiography of the right coronary artery (RCA) taken from the left anterior oblique view with cranial angulation. The tumour encases the mid-segment of the RCA and derives its entire dense vascular supply from the RCA.

OUTCOME AND FOLLOW-UP

Postsurgery, despite discontinuation of α - and β -blockade, the patient was bradycardic and hypotensive requiring epinephrine, vasopressin and norepinephrine infusions. The epinephrine and vasopressin infusions were weaned by postoperative day (POD) #1, and the patient was off all vasopressors by POD#2. She could not be extubated until POD#2, due to excessive lethargy and weakness. Currently, 18 months postsurgery, she is well; her spells have completely resolved with normalisation of heart rate, blood pressure and urinary excretion of norepinephrine and normetanephrine.

DISCUSSION

Catecholamine-secreting tumours arise from chromaffin cells of the adrenal medulla (pheochromocytoma) or the autonomic nervous system (extra-adrenal pheochromocytomas, also called extra-adrenal catecholamine-secreting paragangliomas) and are rare with an annual incidence of 2–8 cases per million.¹ Of these, only 2% are mediastinal paragangliomas.² Brown *et al*³ described a case series of 12 patients with mediastinal paragangliomas; in all patients the tumour was adjacent to the heart or great vessels. Medical pretreatment, often with α - and β -blockade, is recommended to prevent a hypertensive crisis. The majority of cardiac pheochromocytomas are successfully treated with surgical resection. However, in cases where these vascular tumours are significantly invading the myocardium, there can be significant mortality and inability to completely excise the tumour; therefore, it has been suggested that cardiac transplantation could be an alternative option.⁴

To our knowledge, cardiac pheochromocytoma presenting during pregnancy has not been previously described.

Learning points

- ▶ In the non-pregnant patient, a wide range of imaging modalities is available for tumour localisation after a biochemical diagnosis of pheochromocytoma is made.
- ▶ During pregnancy, the imaging options are limited due to safety considerations, and, as in our case, the source of excess catecholamine production may not be found until after the child is born.
- ▶ The anatomic extent of the cardiac pheochromocytoma and the extent of involvement of the coronary arteries needs to be assessed, allowing for successful surgical planning and preparation.

Competing interests None.

Patient consent Obtained.

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