

## CASE REPORT

## Left atrial phaeochromocytoma

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Phaeochromocytomas of the heart are very rare. This report describes the case of a 69 year old woman presenting with persistent hypertension and a left atrial phaeochromocytoma diagnosed by  $^{131}\text{I}$ -metaiodobenzylguanidine scintigraphy scanning. She was successfully treated by surgical excision with the aid of cardiopulmonary bypass and perioperative  $\alpha$  and  $\beta$  adrenergic blockade.

Phaeochromocytomas are relatively rare tumours, the majority of which are located in the abdominal cavity, in association with the adrenal glands. Only 2% of these tumours are found in the thoracic cavity, with cardiac localisation being very rare.<sup>1</sup> We present the case of a left atrial phaeochromocytoma successfully treated with surgical management.

## CASE REPORT

A 69 year old woman presented with poorly controlled hypertension, despite being treated with phenoxybenzamine 10 mg twice daily, nifedipine 60 mg once daily, and ramipril 2.5 mg twice daily. The patient did not experience any headaches, dizziness, or palpitations. There was no family history of phaeochromocytoma or other related disorders. Physical examination showed a raised blood pressure of 180/130 mm Hg. There was no evidence of abdominal masses or bruits or any radiofemoral delay. Fundoscopic examination was unremarkable. A chest radiograph showed the presence of a mediastinal mass visible just below the left pulmonary artery. Initial biochemical investigations showed greatly raised urinary noradrenaline (norepinephrine) concentrations (3375 nmol/24 hours; normal < 450 nmol/24 hours) but normal adrenaline (epinephrine) (62 nmol/24 hours; normal < 100 nmol/24 hours). A clonidine suppression test was positive, suggesting a diagnosis of phaeochromocytoma.

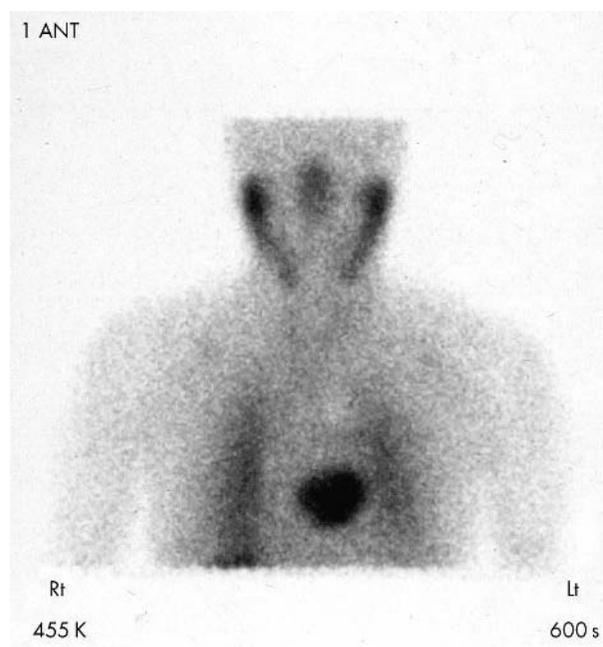
In view of these findings, an iodine-131 labelled metaiodobenzylguanidine (MIBG) scintillation scan was arranged, which showed an area of intense uptake in the middle mediastinum (fig 1). Although an initial computed tomogram (CT) failed to visualise any tumour, subsequent magnetic resonance imaging showed a 7.2 × 5.6 × 4.6 cm mass attached to the left atrium, lying posterior to the pulmonary artery and aortic root (fig 2) and corresponding to the area of increased uptake on the  $^{131}\text{I}$ -MIBG scan. Echocardiography confirmed an otherwise normally functioning heart with only mild mitral regurgitation.

After meticulous anaesthetic preparation with  $\alpha$  and  $\beta$  adrenergic blockade, a median sternotomy showed a 6 cm reddish tumour mass attached to the left atrium. After instituting cardiopulmonary bypass through the right atrium and ascending aorta, systemic cooling to 28°C and cardioplegia to arrest the heart, the tumour mass was excised from the left atrium and posterior aspect of the aortic root (fig 3). The postoperative course was uncomplicated and the patient was discharged on the seventh day after her operation.

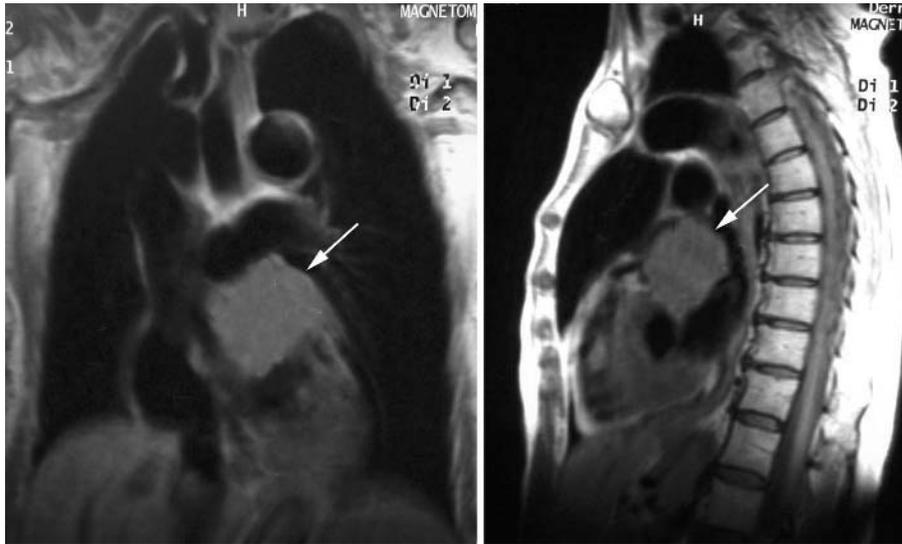
Immunohistochemical analysis of the tumour mass confirmed phaeochromocytoma. At subsequent follow up two years later, her blood pressure was well controlled with ramipril 10 mg once daily.

## DISCUSSION

Phaeochromocytomas are functionally active catecholamine secreting tumours arising from chromaffin cells of the sympathetic nervous system. In adults, the majority of chromaffin cells are located in the adrenal medulla, but aberrant collections are also scattered throughout the body. These have been classified into four groups based on anatomical location and innervation: sympathetic chain; nodose and jugular ganglia of the vagus nerve; visceral paraganglia of heart, liver, bladder, and gut; and branchial arch derived structures including jugular, tympanic, and laryngeal paraganglia, and carotid and subclavian bodies.<sup>2</sup> Subsequently, the majority of phaeochromocytomas are found in the abdomen, with fewer than 2% in the chest, usually in the posterior mediastinum along the paravertebral sympathetic ganglia.<sup>1</sup> Phaeochromocytomas of the middle mediastinum are rare and arise either from coronary or aortopulmonary paraganglia (branchial arch derived structures) or visceral autonomic paraganglia of the heart. Cardiac phaeochromocytomas are most often found on the roof of the left atrium but can also be found on the intra-atrial septum and anterior surface of the heart.<sup>3</sup>



**Figure 1** Iodine-131 labelled metaiodobenzylguanidine (MIBG) scintillation scan illustrating an area of intense uptake in the middle mediastinum. ANT, anterior; Lt, left; Rt, right.



**Figure 2** Coronal and sagittal magnetic resonance imaging showing the left atrial phaeochromocytoma lying close to the pulmonary artery (arrows).

The first phaeochromocytoma involving the heart was described in 1974, being surgically resected from the left atrial wall.<sup>4</sup> Since then only 46 cases of cardiac phaeochromocytoma have been described in the literature. These patients usually present with arterial hypertension, as in our patient, or with the classic symptoms of headache, palpitations, and sweating. Cardiac arrhythmias are also common in patients with mediastinal phaeochromocytomas.<sup>5</sup> These symptoms are caused by release of large amounts of catecholamines, mainly noradrenaline and occasionally adrenaline.<sup>6</sup> These secreting tumours are usually diagnosed by urine catecholamine studies. As the majority of functioning tumours are intra-adrenal in origin, contrast enhanced CT of the abdomen with thin slices of the adrenal glands is carried out to locate the tumour. As with our patient, however, CT may not always detect extra-adrenal phaeochromocytomas and total body <sup>131</sup>I-MIBG scintigraphy scanning is required when the adrenals are normal. The radioisotope <sup>131</sup>I-MIBG accumulates preferentially in these tumours in proportion with catecholamine synthesis. As these lesions may be malignant, whole body screening with <sup>131</sup>I-MIBG may provide preoperative information on the extent of tumour spread and the presence of metastases. As

opposed to CT, magnetic resonance imaging has been found to be more sensitive in localising extra-adrenal phaeochromocytomas, with its greater resolution and ability to differentiate soft tissues.<sup>7</sup>

Once localised, cardiac phaeochromocytomas are treated by surgical excision to provide relief of symptoms and effective control of blood pressure, and also because a proportion of these tumours are malignant. Treatment strategies include simple excision, as was feasible with our patient, excision with pericardial patch reconstruction, autotransplantation after excision of the tumour, and orthotopic cardiac transplantation, depending on the extent of the disease.<sup>3</sup> These tumours can be approached by median sternotomy or by left or right thoracotomy, depending on where the tumour is located in the heart. As manipulation of these tumours intraoperatively can lead to severe hypertension and arrhythmias, perioperative  $\alpha$  and  $\beta$  adrenergic blockade is required to suppress the catecholamine activity. Furthermore, cardiopulmonary bypass can be used to maintain cardiac stability by isolating the heart from the circulation; it also allows safe dissection of the tumour from the coronary arteries if required.

In conclusion, this report describes the successful treatment of a rare left atrial phaeochromocytoma with surgical excision under cardiopulmonary bypass and meticulous perioperative  $\alpha$  and  $\beta$  adrenergic blockade.

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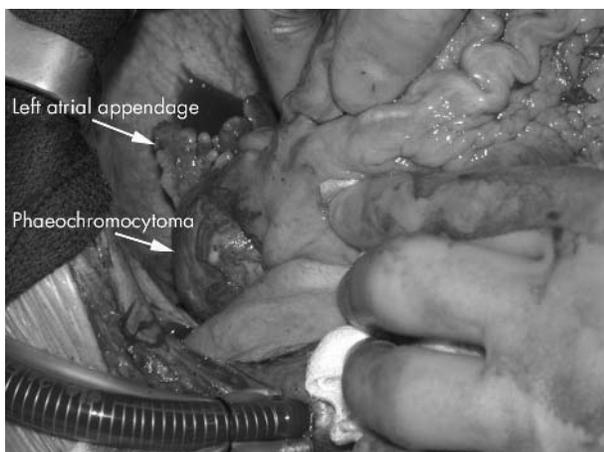
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**Figure 3** Intraoperative image showing the phaeochromocytoma attached to the superior aspect of the left atrium.

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