

## An intrapericardial phaeochromocytoma

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*A case history is presented of a patient with symptoms of a phaeochromocytoma that could not, however, be located in the abdomen. She also had physical and radiological signs of mitral valve disease. After a laparotomy, cardiac investigations suggested the site of the tumour as retrocardiac. Operation confirmed this hitherto unreported site of a phaeochromocytoma, deforming a normal mitral valve. Surgery and the postoperative course were uneventful.*

The localization of a suspected phaeochromocytoma usually presents a diagnostic challenge that can be overcome. In this case report an elusive tumour was eventually traced to an unconventional site and was removed.

### Case report

A 36-year-old woman was first seen in April 1967 with a history of attacks of palpitation which had started during her first pregnancy in 1954. At this time her blood pressure was stated to be normal. During her second pregnancy she had had further similar episodes, and after delivery a clear attack of pulmonary oedema, at which time her blood pressure was recorded as 180/120 mmHg. In a further pregnancy she had had another attack of severe breathlessness with some haemoptysis just before delivery. Otherwise, apart from these episodes, she had remarkably few symptoms other than fatigue and slight shortness of breath on exertion. There was no significant family history.

On examination, though very thin, her general condition was good. Her blood pressure varied between 140/90 and 170/100 mmHg in all positions. There was some left ventricular overactivity and a late systolic murmur was heard at the apex. No abnormality was found in any other systems. Urinary vanillylmandelic acid (VMA) estimations varied from 10 mg to 19.2 mg/24 hr. Free noradrenaline varied between 1 to 2 mg/24 hr. The homovanillic acid was conspicuously raised. Retroperitoneal air insufflation showed both kidneys and the suprarenal areas were well outlined with no tumour visible. An intravenous pyelogram was normal and abdominal aortography was also normal, showing no evidence of large vessels in the suprarenal areas or any mass in the para-aortic or pelvic regions. X-ray of the chest showed some prominence of the upper left heart border but the heart itself was not enlarged. It was considered that a phaeochromocytoma was almost certainly present and exploration of the suprarenals via loin incisions was carried out in May 1967 by Mr. Kenneth Owen. Both glands were normal. No tumour could be found in the para-aortic region despite an extensive search, and cystoscopy was also normal.

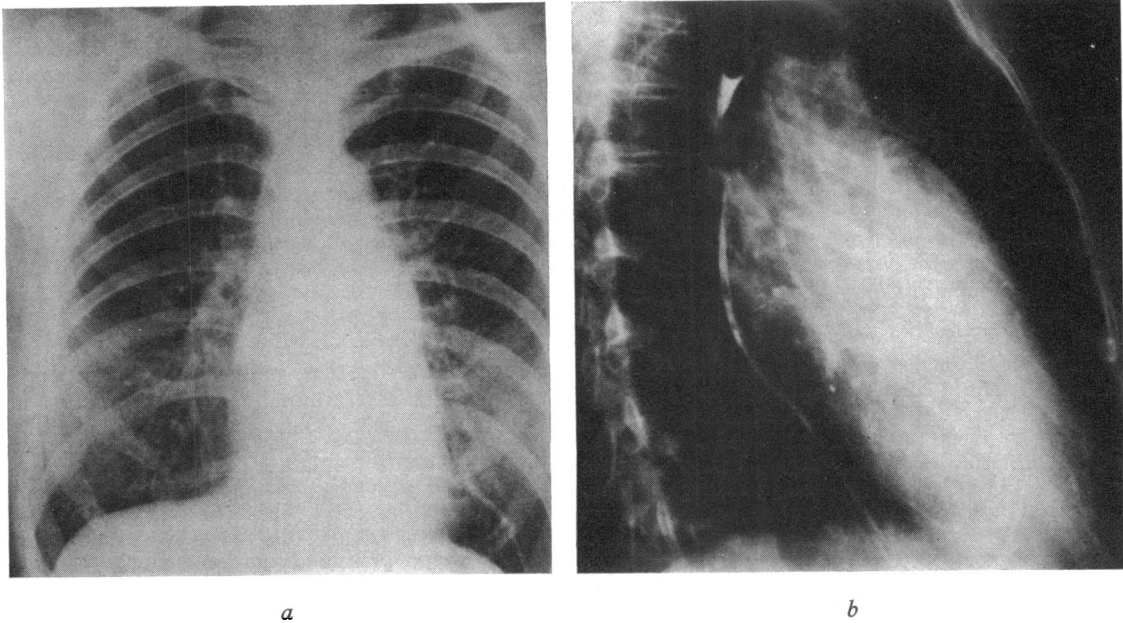
At the end of the procedure she developed pulmonary oedema but responded rapidly to treatment and then made a normal postoperative recovery.

Before returning home she had venous samples taken from the inferior vena cava at various levels with stimulating doses of tyramine being given intravenously during this procedure. No excess of pressor amine was discovered at any level.

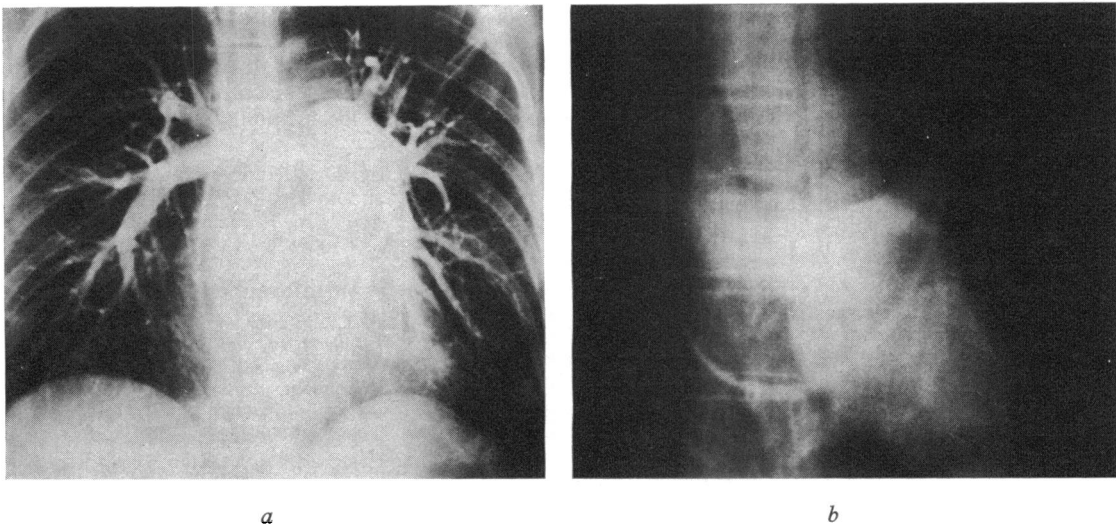
She was readmitted in January 1970 for review. Her symptoms were much as before: mild shortness of breath on exertion, fatigue, and occasional attacks of palpitation. The apical systolic murmur was still present. The chest x-ray again showed a bulge on the left border thought to be due to slight enlargement of the left atrium and appendage compatible with a diagnosis of mitral disease (Fig. 1a). Barium swallow also suggested this (Fig. 1b). She had had more blood staining of her sputum but bronchoscopy showed no abnormality. Right heart catheterization was carried out and revealed normal pulmonary artery and wedge pressures both at rest and on effort. Right-sided angiocardiology showed a high left pulmonary artery (Fig. 2a) which was displaced laterally as well as upwards and was simulating an enlarged left atrial appendage. The left atrium itself was in fact small and, when opacified, appeared to be displaced downwards (Fig. 2b). This chamber bore no relation to the indentation of the barium-filled oesophagus (Fig. 1b).

Comparison of the straight and oblique x-rays with the angiocardiology suggested that a mass was displacing the pulmonary artery upwards and to the left, and was also displacing a small left atrium in a downward direction; it was also indenting the oesophagus. Subsequent left ventricular angiocardiology confirmed the presence of mitral regurgitation into a small left atrium and root aortograms showed the mass received a large arterial supply from many small vessels. Thus an apparently enlarged left atrium was in fact a vascular tumour that was distorting the mitral valve and causing functional mitral regurgitation in late systole. 24-hour urinary VMA estimations continued to be raised.

Pressor assay of blood samples taken during angiocardiology showed high concentrations of catecholamines in the left and right atrium as well as the aorta as compared with the peripheral venous sample (Fig. 3). This, therefore, localized the tumour in the atrial region



**FIG. 1** a) Posteroanterior chest x-ray showing bulge on the left heart border. b) Barium swallow showing indentation of the oesophagus higher than usually seen when due to left atrial enlargement.



**FIG. 2** a) Pulmonary arteriogram showing upward and lateral displacement of the left pulmonary artery. b) Later angiogram showing downward displacement of the left atrium.

with drainage into both atria since the pressor levels were roughly equivalent.

It was, therefore, concluded that the mass in relation to the left atrium was almost certainly a phaeochromocytoma and removal was advised. This was carried out on 13 May 1970 by midline sternotomy and on cardio-

pulmonary bypass. A large mass was found arising from the posterior pericardium and superior aspect of the left atrium above the pulmonary veins. The tumour was enucleated through a very vascular plane, multiple small arteries supplying the tumour. The tumour was firmly connected to the postero-superior wall of the left atrium,

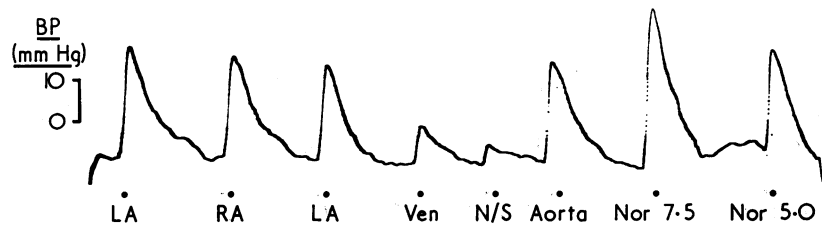


FIG. 3 Pressor assay in the anaesthetized ganglion blocked rat of plasma taken from various intravascular sites by catheter. LA, left atrium; RA, right atrium; Ven, peripheral venous; N/S, normal saline; Nor, noradrenaline: dose, 7.5 and 5 ng.

and final freeing entailed removal of a circular portion of extremely thinned atrial wall approximately 2 cm in diameter. Haemostasis was difficult to achieve, and it was necessary to divide the aorta and pulmonary artery to obtain satisfactory exposure. It was then possible to suture the capsule of the tumour and to close the defect in the wall of the left atrium. The aorta and pulmonary arteries were then repaired and, the cardiovascular state being satisfactory, perfusion was stopped and the chest closed.

An adrenaline drip was maintained for 12 hours, but after this the systemic blood pressure was satisfactorily maintained. The patient was slow to return to full consciousness, but 24 hours after operation she was awake and mentally alert.

The tumour weighed 80 g, measured 8 × 5 cm, and contained the equivalent of 1 mg noradrenaline per gram. There was a little adrenaline present. Histologically it was more vascular than a typical phaeochromocytoma but there was no evidence of malignancy.

Her subsequent postoperative progress was uneventful and at the time of discharge from hospital, 4 weeks after operation, urinary VMA estimation was less than 7 mg/24 hr. Two years after operation her condition was satisfactory, with a normal blood pressure and no mitral systolic murmur. Urinary VMA estimation was again less than 7 mg/24 hr.

### Discussion

A phaeochromocytoma is a rare benign tumour and can arise wherever there are chromaffin cells. Thus, the vast majority of these tumours arise where chromaffin tissue is most plentiful, in one or other adrenal gland. However, about 6 per cent arise in other sites, especially retroperitoneally in the para-aortic region close to the origin of the inferior mesenteric artery, in the bladder wall, or within the thorax (Symington and Goodall, 1953). McNeill, Groden, and Neville (1970) reviewed 22 reported examples of intrathoracic phaeochromocytomata and added

a further case. In these patients the tumours presented in the paravertebral gutter in relation to the sympathetic chain. As far as is known, an intrapericardial tumour has not been previously described, yet chromaffin cells can be shown in close relation to the heart and great vessels alongside peripheral fine sympathetic nerve fibres (Coupland, 1965; 1971, personal communication). Chemoreceptor carotid body type tissue is also plentiful within the pericardium in small aggregates, and chromaffin type cells may be seen within such tissue (Boyd, 1961). Whether these cells are catecholamine secreting or have tumour potential is speculative. Nevertheless, in this patient, though the exact site of origin and tissue of origin of the phaeochromocytoma could not be determined with certainty, its intrapericardial origin seemed undisputed. This site should be borne in mind when there are difficulties in localization.

We are indebted to Dr. J. Finberg for carrying out the pressor assays, and to Dr. D. Sutton for radiological assistance.

### References

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