A functioning intrapericardial paraganglioma (pheochromocytoma)

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SUMMARY A patient with a functioning intrapericardial paraganglioma (pheochromocytoma) that was located at the root of the aorta overlying the right coronary artery and adherent to the right ventricular wall is reported. The tumour was successfully removed under total cardiopulmonary bypass without inducing cardiac arrest.

Since 1974 when Besterman *et al* reported an intrapericardial pheochromocytoma,¹ other cases have been reported.²⁻¹⁰ In most reported cases¹⁻⁶ the tumour seemed to be related to the left atrial wall. We report a case in which the pheochromocytoma was located at the root of the aorta.

Case report

About 10 years before presentation this 49 year old woman had first noted palpitation, headache, and weakness in her feet. Appendectomy (at age 19) and two pregnancies and deliveries (at 27 and 31) had been uneventful. She had an intraoperative hypertensive crisis and a postoperative hypotensive crisis at total hysterectomy and bilateral adnexectomy at 43.

On 16 February 1980, she was admitted to the Keio University Hospital. She was of normal size and shape, and had two abdominal scars. No mass was palpable in the abdominal area. Blood pressure was 150/80 mm Hg and rose to 210/110 mm Hg within a few minutes of the intravenous injection of 10 mg of metoclopramide.¹¹ Urinary excretion of noradrenaline was 4782 nmol/24 h (809 µg/24 h) (normal 59-532 nmol/24 h) (10 to 90 µg/24 h) and the concentration of plasma noradrenaline was 12.05 nmol/l (2.04 ng/ml) when the patient was recumbent (normal < 2.4 nmol/l (0.40 ng/ml)). We suspected that she had a pheochromocytoma.

Because the site of the tumour was not known she was treated with labetalol (400 mg per day). On 14

October 1985 she was readmitted to our hospital because of an attack of paroxysmal hypertension and palpitation. Urinary excretion of noradrenaline was 25.7 µmol/24 h (4.352 µg/24 h) and the concentration of plasma noradrenaline while the patient was recumbent was 36 nmol/l (6.10 ng/ml). On 20 November 1985 whole body ¹³¹I-metaiodobenzylguanidine scintigraphy revealed a lesion in the thorax (fig 1a). Computed tomography of the chest on 4 December 1985 showed an abnormal mass in the mediastinum (fig 1b). On 13 December 1985 an arteriogram obtained after injection of contrast into the left internal mammary artery showed hypervascularity of the upper part of the tumour (fig 2).

At operation on 22 January 1986 a dark red and smooth tumour was found when the pericardium was opened. The tumour arose from the root of the aorta; it overlay the right coronary artery and adhered to the right ventricular wall. It was removed under total cardiopulmonary bypass and mild hypothermia without inducing cardiac arrest. The tumour was supplied with many arteries derived from the right coronary artery. The tumour was removed together with a part of the right ventricular myocardium.

The patient's blood pressure became normal on the first postoperative day. After operation urinary excretion of noradrenaline fell to 893 nmol/24 h (151 µg/24 h) and plasma noradrenaline fell to 2 nmol/l (0.34 ng/ml). Her blood pressure did not increase after intravenous metoclopramide.¹¹ She was discharged on the twenty-fifth postoperative day off medication.

The tumour measured 5.0 × 4.5 × 3.5 cm. The cut area showed a homogeneous, brown mass of

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Fig 1 (a) ^{131}I -metaiodobenzylguanidine scintiscan (anterior image of chest), three hours after a dose of 1.0 mCi, showing lesion in the thorax. (b) Computed tomographic scan of the chest showing an abnormal mass (arrow).

nodular tissue. The tumour was surrounded by a collagenous capsule and was firmly attached to the myocardium, but there was no infiltration of the myocardium. There were many blood vessels, some of which were dilated and some which were compressed. The tumour cells were arranged in clusters of various sizes which were separated by fibrous septa. The tumour cells had egg shaped hyperchromatic nuclei of slightly varying sizes, but active mitotic figures were not seen. Many had cytoplasmic vacuoles. The tumour cells contained abundant cytoplasmic granules (fig 3). Strong staining of the granules by the Grimelius silver reaction suggested that these were secretory granules (fig 3).

Electronmicroscopy showed that the chief cells

contained cytoplasmic granules with an electron-dense core (fig 3). The diameter of these secretion granules ranged from 150 nm to 200 nm. The tumour contained 3.34 mg of noradrenaline per gram but not adrenaline or dopamine. The tumour was identified as a benign functioning paraganglioma (pheochromocytoma).

Discussion

Intrapericardial paragangliomas are rare tumours. There are two types. The first are called chemodectomas; they are chromaffin negative and they originate from the parasympathetic chain and are seen in chemoreceptor tissue. According to Gopalakrishnan *et al* these tumours are non-functioning in the intrapericardial site.¹² The second group comprises the pheochromocytomas which are chromaffin positive and arise from the sympathetic nervous system. These tumours are catecholamine secreting and functioning paragangliomas. Intrapericardial pheochromocytomas usually involve the left atrial wall. Shapiro and colleagues reported six pheochromocytomas originating from the left atrium and another detected in the aortopulmonary window.⁵ Besterman *et al*, Wilson *et al*, Voci *et al*, Saad *et al*, and Fisher *et al* have all reported cases in which a pheochromocytoma was attached to the left atrium.^{1-4,6} Others have described pheochromocytomas arising from the interatrial septum.^{7,8} David *et al* reported a case in which a pheochromocytoma involved the posterior wall of the left ventricle and coronary sinus.⁹ A right atrial pheochromocytoma has also been reported.¹⁰

To our knowledge, a pheochromocytoma arising from the root of the aorta, overlying the right coronary artery, and adherent to the right ventricular wall has not been described before. A tumour at this site is likely to have arisen from chromaffin cells of the paraganglia in the ventral connective tissues between the descending aorta and the pulmonary trunk. This paraganglion is called the coronary paraganglion and receives its blood supply from the left coronary artery in adults.¹³ Boyd has pointed out that in the fetus the coronary paraganglion receives an additional blood supply from the right coronary artery.¹⁴ The fact that the tumour in our patient was mainly supplied from the right coronary artery suggests that this artery may supply the coronary paraganglia in adults as well. Angiography showed that the upper part of the tumour was supplied from the left internal mammary artery. Because there may be anastomoses between the coronary arteries and the internal mammary artery,¹⁵ it is not impossible that a branch of the internal mammary artery supplied the region in which the tumour was found. We

Fig 2 Arteriograms obtained after injection of contrast into the left internal mammary artery showing hypervascularity along the upper part of the tumour (arrows).

therefore propose that there is another blood supply to the coronary paraganglion in adults.

^{131}I -metaiodobenzylguanidine scintigraphy was most helpful in determining the site of the pheochromocytoma in this case. Computed tomography of the chest was useful in determining the anatomical connections with the intrathoracic organs. ^{131}I -metaiodobenzylguanidine is an appropriate agent for the search for extra-adrenal pheochromocytomas.

Microscopy showed that the tumour was firmly attached to the myocardium though no infiltration was noted. Removal of tumours from this site requires resection of a part of the right ventricular wall. The tumour was completely removed under cardiopulmonary bypass without any functional damage to the right coronary artery. Induction of cardiac arrest was not needed because there was no arrhythmia; thus the right ventricular wall could be reconstructed without any difficulty.

We thank Professor Tadashi Inoue and Takao Sar-

uta for their helpful advice and criticism and Hiroaki Suzuki, Toshio Imafuku, and Hiroaki Nomori for their preoperative management of our patient. We also thank Chikao Torikata for the photomicrographs and photoelectronmicrographs.

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Fig 3 (a) Photomicrograph (haematoxylin and eosin) of the tumour showing the arrangement of cells into alveoli. Tumour cells contain egg-shaped hyperchromatic nuclei and abundant cytoplasmic granules. There are vacuoles in the cytoplasm in many cells. (b) Photomicrograph (Grimelius stain) showing strong staining of cytoplasmic granules which indicates that they are secretory granules. (c) Photoelectronmicrograph of a tumour cell containing many secretion granules. (d) High power photoelectronmicrograph showing that the diameter of a granule is about 150–200 nm.

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