remedied with improved product design.

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Cardiac Pheochromocytoma Originating in the Interaltral Septum

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A 36-year-old woman with classic clinical and biochemical features of pheochromocytoma was found to have a cardiac pheochromocytoma originating in the interaltral septum and a significant obstructive lesion in the left anterior descending coronary artery. Complete resection of the pheochromocytoma and an aortosaphenous vein graft were performed. This is the first reported case of successful resection of an interaltral pheochromocytoma. (Chest 1990; 97:760-62)

MRI = magnetic resonance imaging, 131I-MIBG = 131I-meta iodobenzylguanidine; VMA = vanillylmandelic acid

Cardiac pheochromocytoma is extremely rare. Only 14 cases have been reported. The patient reported here was found to have a cardiac pheochromocytoma involving the interaltral septum. She underwent surgical resection with a successful outcome.

CASE REPORT

A 31-year-old married black woman was seen at the Harbor City Kaiser medical clinic for the first time in May of 1982 for treatment of chronic hypertension. She had a history of hypertension since 1973. She described frequent palpitations, excessive diaphoresis and light-headedness. Blood pressure with the patient in the supine position was 220/140 and it dropped to 90/70 mm Hg when the patient stood up.

The patient was admitted to the hospital with a strong suspicion of pheochromocytoma. Total 24-h urinary VMA was elevated at 26 mg (normal, 0-7 mg). An adrenal angiogram and selective venous samplings were suggestive of a possible tumor in the left adrenal gland. She underwent an exploratory laparotomy. The left adrenal gland was resected, but no pheochromocytoma was found. She was discharged on a regimen of phenoxymethylamine and atenolol with controlled blood pressure. Several months later she developed overt diabetes mellitus and was treated with an oral hypoglycemic agent. In March of 1985, a routine chest x-ray film revealed an unusually prominent right atrial border but the patient declined further...
investigations and surgery.
In December of 1987, the patient developed intermittent chest pain and progressive exertional dyspnea. After an episode of epistaxis lasting two weeks the patient agreed to undergo surgery. Physical examination revealed an obese woman who was 167 cm tall and weighed 96 kg. Blood pressure with the patient in the supine position was 180/120 mm Hg and upon standing blood pressure dropped to 120/80 mm Hg. Pulse was 110 beats per minute and regular. Pertinent cardiac findings were moderate parasternal lift and prominent left ventricular impulse. The ECG showed left ventricular hypertrophy. The two-dimensional echocardiogram revealed hyperdynamic and hypertrophic left ventricle. The apical four-chamber view demonstrated a large round mass located in the upper interatrial septum (Fig 1). Gated MRI showed a large mass (6 x 6 x 5 cm) in the interatrial septum with some encroachment upon the right atrial lumen. The \textsuperscript{123}I-MIBG scan was positive.

Cardiac catheterization revealed normal cardiac pressures, but arterial pressure peaked up to 250/190 mm Hg. Selective blood samples revealed the highest norepinephrine level, 16,522 pg/ml (normal, 138 to 480 pg/ml) in the coronary sinus, indicating intracardiac location of a pheochromocytoma. Selective coronary angiogram revealed dense vasculization to the tumor from the left circumflex coronary artery and a critical stenosis in the middle of the left anterior descending coronary artery (Fig 2).

On February 5, 1988, a median sternotomy was done and a 7 x 6 x 5-cm ovoid, firm, solid tumor was resected along with most of the interatrial septum and adjacent right and left atrial walls using cardiopulmonary bypass (Fig 3). The tumor was dissected from the aorta and superior vena cava which was totally detached from the right atrium to facilitate removal of the tumor. Reconstruction of the interatrial septum, right and left atrial walls was done with pericardial patches and the superior vena cava was reattached to the right atrium. An aortosaphenous vein bypass was performed with endarterectomy of the left anterior descending coronary artery. Microscopic examination revealed the typical appearance of a benign pheochromocytoma. She was discharged on a regimen of a small dose of furosemide for pedal edema.

During the subsequent one year follow-up, the patient has been completely asymptomatic. Blood pressure has ranged from 120/80 to 160/90 with normal 24-h urinary VMA. Diabetes mellitus disappeared completely.

**Discussion**

Pheochromocytoma is a functionally active catecholamine-producing tumor arising from chromaffin-positive cells of the sympathetic nervous system. This tumor creates a hypersympathetic state of varying degree by producing large amounts of catecholamines, primarily norepinephrine and less frequently epinephrine. The incidence of pheochromocytoma is 0.1 percent among hypertensive patients; 90 percent of these tumors are benign and potentially curable.

![FIGURE 1. Two-dimensional echocardiogram demonstrating sound mass in the interatrial septum. Left: apical four chamber view. Right: diagram.](image)

![FIGURE 2. Left coronary angiogram (30 degree RAO view) demonstrating tumor neovascularization from the left circumflex coronary artery and critical lesion in the middle of the left anterior descending artery.](image)
Cardiac pheochromocytoma is very rare and so far 14 cases have been reported since the first case was described by Besterman et al. The most common location of this tumor was in the left posterior atrial wall, which occurred in eight previously reported patients. Two patients had interatrial involvement. In one patient the tumor was found during autopsy. The other patient died during attempted operative resection. The patient reported here had a large interatrial septal pheochromocytoma involving both adjacent atrial walls. The tumor was resected en bloc and reconstructed with pericardial patches. This is the first reported successful resection of cardiac pheochromocytoma originating in the interatrial septum.

Localization of the cardiac pheochromocytoma became easier with the advent of 123I-MIBG scintigraphy introduced by the University of Michigan in 1982. This radiopharmaceutical's molecular structure closely resembles that of norepinephrine and it concentrates in catecholamine storage vesicles such as within the adenalin medulla and in pheochromocytoma. At the present time the gated MRI is the best technique for the demonstration of the detailed anatomy of cardiac pheochromocytoma and its relationship to other structures.

Selective blood samplings of catecholamines during cardiac catheterization can confirm the site of a functioning pheochromocytoma in the heart by demonstrating the highest concentrations of catecholamines in the coronary sinus, as shown in this case.

Echocardiography is generally not helpful because these tumors frequently arise from the posterior left atrial wall area which is difficult to differentiate from the surrounding structures. In this case, however, the echocardiogram revealed the tumor clearly because of its location between the right and left atrial chambers. This is the first reported two-dimensional echocardiogram demonstrating an interatrial pheochromocytoma.

Another unique feature of this case was the presence of a significant atherosclerotic lesion in the left anterior descending coronary artery which was bypassed with an aortosaphenous vein graft. Two previous patients with cardiac pheochromocytomas had received aortosaphenous vein bypass grafts because of location of tumors over coronary arteries. Although this patient was only 36-years-old, the presence of premature atherosclerotic coronary artery disease was not surprising in view of several serious coronary risk factors including severe hypertension, overt diabetes mellitus and hyperlipidemia, all of which were exacerbated by the catecholamine-producing pheochromocytoma. In light of this experience any patient with pheochromocytoma should be evaluated carefully for the presence of premature atherosclerotic coronary disease.

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