

- pulmonary valve stenosis. *N Engl J Med* 1982;307:540.
6. Cribier A, Saoudi N, Berland J, Savin T, Rocha P, Letae B. Percutaneous transluminal valvuloplasty of acquired aortic stenosis in elderly patients: An alternative to valve replacement? *Lancet* 1986;1:63.
  7. Carver JM, Goldstein J, Jones EL, Knapp WA, Hatcher CR. Clinical hemodynamic and operative descriptors affecting the outcome of aortic valve replacement in elderly versus young patients. *Ann Surg* 1984;199:733.

## Cardiac pheochromocytoma involving the left main coronary artery presenting with exertional angina

Stephen A. Stowers, M.D., Paul Gilmore, M.D., Mack Stirling, M.D., James M. Morantz, M.D., Alan B. Miller, M.D., Laura J. Meyer, C.N.M.T., Gary Glazer, M.D., and Douglas Behrendt, M.D.  
*Jacksonville, Fla., and Ann Arbor, Mich.*

Intrathoracic pheochromocytomas are rare. Presenting symptoms are usually hypertension, sweating, headaches, palpitations, and restlessness. Although paragangliomas involving the coronary arteries have been reported, to our knowledge this is the first case of a functioning paraganglioma (pheochromocytoma) involving the left main coronary artery causing exertional angina.<sup>1-6</sup> Successful excision of these tumors depends on precise assessment of their relationship to surrounding structures. Magnetic resonance imaging was first used in this case to define the extent of cardiac invasion by a primary cardiac pheochromocytoma. Imaging with <sup>131</sup>I-metaiodobenzylguanidine was also employed for tumor localization and detection of metastases. This case also illustrates the failure of bypass surgery in the treatment of this condition and emphasizes that total removal of the tumor and reconstruction of involved cardiac structures are the only means of achieving a cure.

A 28-year-old black woman went to a local community hospital with exertional chest pain and dyspnea. Her pain was substernal, accompanied by shortness of breath, left arm numbness, vomiting, and diaphoresis. The pain had increased in severity and frequency during the 2 months before her visit to the hospital. It was exacerbated by exertion and relieved by rest. Her initial physical examination showed a blood pressure of 140/76 mm Hg, a pulse of 80 bpm, normal temperature, and respirations of 12. The chest was clear to auscultation. S<sub>1</sub> and S<sub>2</sub> were normal. A II/VI systolic ejection murmur was present at the aortic

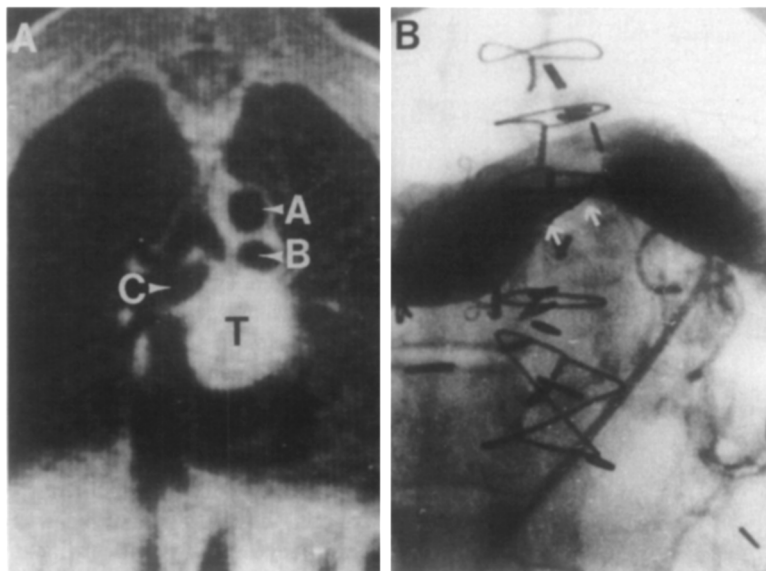


**Fig. 1.** Right anterior oblique cineangiogram demonstrates subtotal occlusion of left main coronary artery (arrow).

area without radiation. No clubbing or cyanosis was observed. The remainder of the physical examination was unrevealing. The ECG at rest revealed 1.5 to 2 mm ST segment depression in precordial leads V<sub>2</sub> to V<sub>6</sub>, 1 mm ST depression in leads II and III, and arteriovenous fistula. The impression was that she was in New York Heart Association class IV because of angina, and she underwent cardiac catheterization. Coronary arteriography displayed a large vascular mass with external compression of the left main coronary artery resulting in 95% obstruction (Fig. 1). The blood supply to this highly vascular mass arose directly from the left main coronary artery. The remainder of the coronary vessels were normal, although retrograde filling of the left anterior descending artery was noted during the right coronary injection. The mass was thought to be unresectable because of involvement of the left atrium. Four-vessel coronary artery bypass grafting was performed to the left anterior descending, ramus intermedius, first obtuse marginal, and second obtuse marginal arteries, and the mass was biopsied. In an attempt to interrupt the blood supply of the tumor, the ostium of the left main coronary artery was oversewn through an incision in the aorta and the bypassed vessels were ligated proximal to their grafts. Biopsy specimen of the mass was consistent with paraganglioma. Subsequent urine testing revealed total catecholamine excretion of 10

From the Division of Cardiology, University of Florida, University Hospital of Jacksonville, and the Departments of Surgery, Radiology, and Internal Medicine (Division of Nuclear Medicine), University of Michigan.

Reprint requests: Stephen A. Stowers, M.D., University Hospital of Jacksonville, 655 W. Eighth St., Jacksonville, FL 32209.



**Fig. 2.** A, Left lateral cineangiogram demonstrates tumor blush with catheter in left coronary ostium (arrow). B, Sagittal magnetic resonance image demonstrates tumor (T) anterior to descending aorta (AO) and occupying left atrium. (LV = left ventricle.)

**Table I.** Case review of intracardiac pheochromocytomas

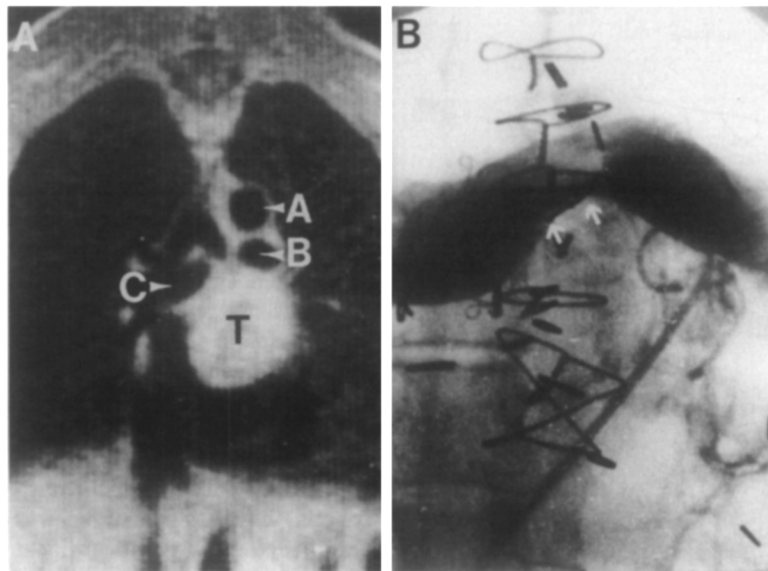
Authors	Patient (age (yr)/sex)	Presentation	Location	Outcome
Awoke and Perlstein <sup>1</sup> (1985)	41/WF	Headache, dizziness, palpitations, diaphoresis	5 × 5 cm tumor from interatrial septum to right superior pulmonary vein	Died of intraoperative hemorrhage (using CPB)
Cooley et al. <sup>2</sup> (1985)	42/M	Arrhythmias, systolic murmur; echocardiogram revealed mass	Left atrium, 5 × 6 cm	Died of intraoperative hemorrhage (using CPB)
Orringer et al. <sup>3</sup> (1985)	24/WF	hypertension	Left atrium, 5 × 3 × 3 cm	Resection, A/W 2 yr follow-up (without CPB)
	18/BF	Hypertension, peripartum hypertensive crisis	Anterior surface of heart encasing LMCA and adhering to LAD, 5 × 5 × 3 cm	Resection A/W (using CPB)
	30/WF	Hypertension	Left atrium, 5 × 6 cm	Resection and reconstruction of left atrial wall with pericardium (using CPB)
Wilson et al. <sup>4</sup> (1974)	36/WF	Hypertension	Left atrium, 6 × 12 cm	Died of intraoperative hemorrhage (using CPB)
	32/M	Hypertension, headaches	Intra-atrial groove (posterior), 4 × 4 cm	Resection (without CPB)
Besterman et al. <sup>5</sup> (1984)	36/M	Palpitations, fatigue	Left atrium	Resection (using CPB)
Geisler et al. <sup>6</sup> (1985)	26/M	Hypertension	Bifurcation of LMCA	Died postoperative Staph sepsis (using CPB)

CPB = cardiopulmonary bypass; A/W = alive and well; LMCA = left main coronary artery; LAD = left anterior descending; Staph = staphylococcus.

times normal values (predominantly norepinephrine), and she was treated with phenoxybenzamine and propranolol. An echocardiogram confirmed left atrial involvement of the tumor. After surgery the patient again began to

have angina associated with an elevated blood pressure (160/110 mm Hg) and, after 4 months, was referred to University Hospital for additional therapy.

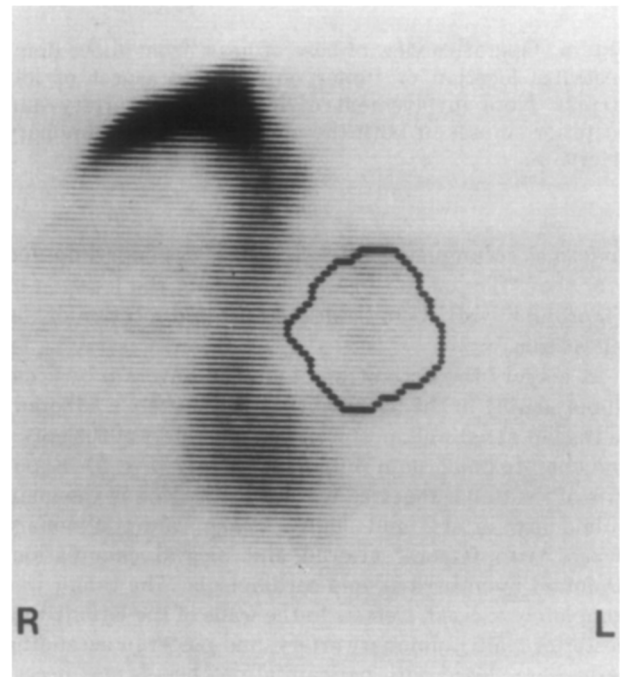
Arteriography again revealed a large vascular tumor



**Fig. 3.** A, Frontal plane magnetic resonance image demonstrates relationship of tumor (T) with right (C) and left (B) pulmonary artery. (A = aorta.) B, Anteroposterior pulmonary angiogram with pigtail catheter in right ventricular outflow tract. Arrows demonstrate extrinsic compression pulmonary artery.

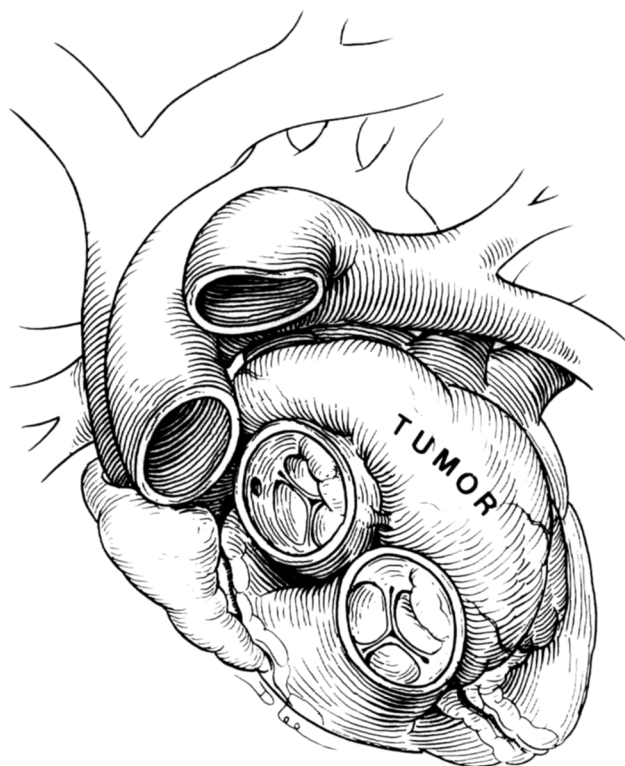
with retrograde flow from the left anterior coronary artery graft (Fig. 2, A). The grafts and coronary arteries were otherwise normal. The main pulmonary artery was distorted and the right pulmonary artery elevated by extrinsic mass effect (Fig. 3), although there was no pressure gradient across the compression. Dynamic computerized transaxial scanning was performed after the bolus injection of contrast material: the scans were only moderately useful in mapping out the relationship of the tumor to the cardiac chambers and great vessels. Magnetic resonance imaging clearly delineated the extent of the tumor and its intimate relation to the left atrium, aorta, and pulmonary arteries (Figs. 2 and 3). Repeat analysis of the urine showed total catecholamines of 5350  $\mu\text{g}/24$  hr (normal range 0 to 230  $\mu\text{g}/24$  hr). The patient's medications were changed from propranolol and phenoxybenzamine to labetalol with better control of hypertension and heart rate. The patient was transferred to the University of Michigan for further studies and surgical resection of the pheochromocytoma.

The patient was prepared for surgery by addition of phenoxybenzamine to her labetalol and by vigorous hydration. The dosage of phenoxybenzamine was progressively increased until the patient developed orthostatic hypotension. Preoperative urine catecholamine levels were elevated: epinephrine, 149  $\mu\text{g}/12$  hr (0 to 20  $\mu\text{g}/12$  hr); norepinephrine, 672  $\mu\text{g}/12$  hr (0 to 100  $\mu\text{g}/12$  hr); metanephrine, 23  $\mu\text{g}/12$  hr (0 to 65  $\mu\text{g}/12$  hr); and normetanephrine, 535  $\mu\text{g}/12$  hr (0 to 163  $\mu\text{g}/12$  hr). Scintigraphic localization of the pheochromocytoma was performed with  $^{131}\text{I}$ -metaiodobenzylguanidine (MIBG) according to standard imaging technique. Faint tracer uptake in the region of the left atrium on the third day after injection suggested the location of the pheochromocytoma to be entirely in the mediastinum with no evidence of metastases. This



**Fig. 4.** First-pass  $^{99\text{m}}\text{Tc}$  red blood cell study with superimposition of the region of interest of abnormal accumulation of  $^{131}\text{I}$  MIBG. Activity is seen in the right subclavian vein, superior vein, superior vena cava, right atrium, and right ventricle. The region of interest corresponds to the expected location of the left atrium.

location was confirmed by performing anterior chest images during a first-pass acquisition with  $^{99\text{m}}\text{Tc}$ -labeled red blood cells and anterior images after the injection of thallium 201. The region of interest drawn around the



**Fig. 5.** Operative view of base of heart from above demonstrates location of tumor on superior aspect of left atrium. Note involvement of left coronary artery and posterior sinuses of both the aorta and main pulmonary artery.

abnormal accumulation of  $^{131}\text{I}$ -MIBG was superimposed over the  $^{99\text{m}}\text{Tc}$  image (Fig. 4), confirming the location of abnormal  $^{131}\text{I}$ -MIBG to be in the expected location of the left atrium.

At surgery the patient was found to have a 6 by 7 cm tumor arising in the left atrioventricular groove adherent to the left atrial wall, posterior proximal part of the aorta, and the proximal main pulmonary artery (Fig. 5). Exposure of the tumor required temporary division of the main pulmonary artery and aorta, using cardiopulmonary bypass with femoral arterial and bicaval cannulation, profound hypothermia, and cardioplegia. The tumor was completely excised. Defects in the walls of the left atrium, posterior main pulmonary artery, and posterior ascending aorta were closed with pericardial patches. It was necessary to insert a new segment of saphenous vein into the graft to the left anterior descending artery because of damage during dissection. Finally, the mitral valve had to be replaced because of severe mitral regurgitation, presumably as a result of papillary muscle ischemia. After surgery the patient was supported with norepinephrine (Levophed) for 3 days. She required reoperation on the first postoperative day for a mediastinal hematoma and cardiac tamponade. An intraoperative myocardial infarction was diagnosed by serial enzymes and ECGs. However, her hospital course was characterized by steady improve-

ment. She was discharged on the fourteenth postoperative day in good condition. Urinary catecholamines and blood pressures were normal. A gated blood pool scan showed a left ventricular ejection fraction of 79%. Another chest  $^{131}\text{I}$ -MIBG scan after surgery demonstrated lack of uptake in the region of the left atrium, indicating complete tumor resection. The patient remains well 1 year after surgery.

This case is unique because the presentation was more consistent with ischemic heart disease than with the usual pheochromocytoma complex of hypertension, palpitations, sweating, and headache. There are probably two mechanisms responsible for this. The left main coronary artery was nearly totally occluded by extrinsic compression by the tumor. Second, catecholamine levels were markedly elevated, increasing myocardial oxygen consumption. Of interest is that inverted T waves and ST segment deviation (which may represent myocardial ischemia) have previously been reported in adrenal pheochromocytoma without cardiac involvement.<sup>7</sup> Localization of functioning tumors in unusual locations is greatly facilitated by the  $^{131}\text{I}$ -MIBG scan. This scan will also detect distant metastases that occasionally occur with these tumors. The precise spatial relationships of the tumor to adjacent cardiac structures was best demonstrated by magnetic resonance imaging, which far surpassed two-dimensional echocardiography and angiography. Furthermore, magnetic resonance imaging is far superior to computerized transaxial scans in assessing cardiac and paracardiac tumors because of better contrast resolution and multiplanar imaging capabilities.

When the tumor was evaluated at the initial sternotomy and judged unresectable, biopsy and coronary artery bypass grafting were performed. Although a measure of palliation was achieved, the patient soon redeveloped symptoms as a result of hypertension and angina. Surgical resection and reconstruction are clearly the only means of achieving a cure. There have been 10 previously reported instances of attempted resection of cardiac pheochromocytomas (Table I), with six patients surviving. Two patients have undergone resection without bypass and eight have undergone resection with cardiopulmonary bypass.<sup>3-6</sup> Three patients died of intraoperative hemorrhage, emphasizing the highly vascular nature of these tumors.<sup>1-3</sup> In one of these cases the heart has been explanted, repaired, and autotransplanted.<sup>2</sup> One patient died of sepsis after surgery.<sup>6</sup> Adequate  $\alpha$ -adrenergic blockade is essential in the preparation of these patients for operation to prevent the occurrence of intraoperative hypertensive crises. For this reason we added phenoxybenzamine hydrochloride (Dibenzylin) for two days before surgery and progressively increased the dosage of this medication to the point of producing postural hypotension. Along with this we administered intravenous fluids and plasma to expand the blood volume as vasodilation occurred. Because of these precautions we had no hemodynamic instability in the operating room during the extensive dissection required before cardiopulmonary bypass. In planning the operation it is also important to recognize that these tumors are not well encapsulated so

that, after even a careful resection, the wall of adjacent cardiac structures is likely to be dangerously thinned or even perforated. Thus, cardiopulmonary bypass is usually necessary to permit safe resection and the necessary reconstruction.

#### REFERENCES

1. Awoke S, Perlstein RS. Dopamine and norepinephrine secreting intrapericardial pheochromocytoma in a normotensive patient. *South Med J* 1985;8:994.
2. Cooley DA, Reardon MI, Frazier OH, Angelini P. Human cardiac explantation and autotransplantation in a patient with a large cardiac pheochromocytoma. *Tex Heart Inst J* 1985;12:171.
3. Orringer MB, Sisson JC, Glazier G, et al. Surgical treatment of cardiac pheochromocytoma. *J Thorac Cardiovasc Surg* 1985;89:753.
4. Wilson AC, Bennet RC, Niall JF, Clarebrough JK, Doyle AE, Laus W. An unusual case of intra-thoracic pheochromocytoma. *Aust NZ J Surg* 1974;44:27.
5. Besterman E, Bromley LL, Peart WS. An intrapericardial pheochromocytoma. *Br Heart J* 1984;36:318.
6. Geisler F, Barath G, Jaeck D, et al. Un cas de pheochromocytome a localisation cardiaque. *Presse Med* 1985;14:1024.
7. Sawyer WJ, Moser M, Mattingly TW. Pheochromocytoma and the abnormal electrocardiogram. *AM HEART J* 1954; 48:42.

### Atherosclerotic coronary artery aneurysm progressing to coronary artery fistula: Presentation as myocardial infarction with continuous murmur

Franklin Zalman, M.D., Ana Marie Andia, M.D.,  
Keun-Teh Wu, M.D., William Y. Moores, M.D.,  
Brian Hoit, M.D., and Alan S. Maisel, M.D.  
*San Diego, Calif.*

Aneurysmal coronary artery disease has become a well-recognized clinical entity.<sup>1,2</sup> Although the prognosis is similar to nonaneurysmal obstructive coronary artery disease,<sup>3,4</sup> the pathogenesis and natural history are not well understood. We report a case of spontaneous progression of an atherosclerotic coronary artery aneurysm to a fistula with resultant myocardial infarction. This case illustrates an unusual and previously unreported consequence of aneurysmal coronary artery disease.

A 64-year-old man with prior hypertension and a recent negative thallium treadmill test presented at an outside

From the Department of Medicine, Cardiology Section, San Diego Veterans Administration Medical Center and University of California, San Diego.

Reprint requests: Alan Maisel, M.D., San Diego VA Medical Center, Cardiology Section (111A), 3350 La Jolla Village Drive, San Diego, CA 92161.

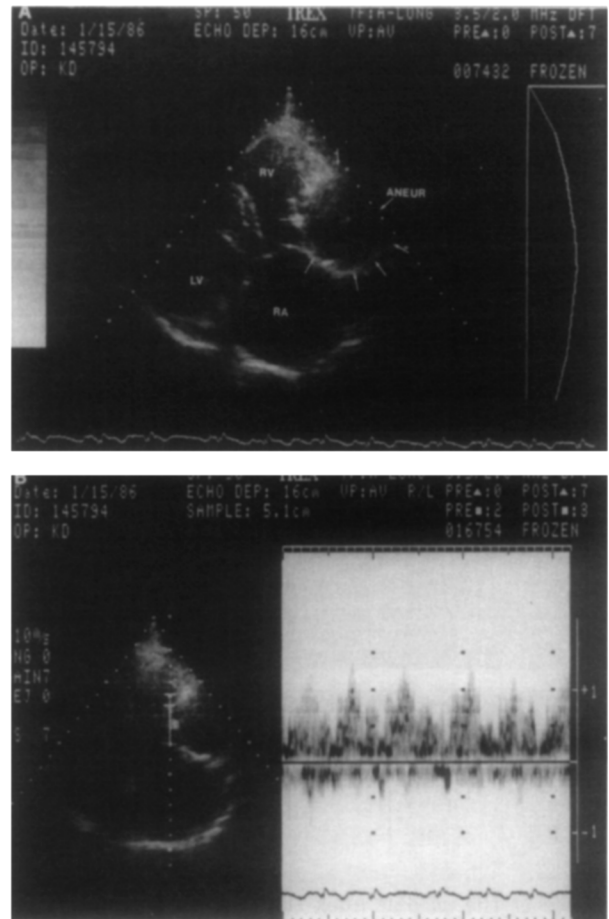


Fig. 1. A, Aneurysm (ANEUR) in the right ventricular long-axis view (arrows). Note the area of echo dropout at approximately 10 o'clock. RA = right atrium; LV = left ventricle; RV = right ventricle. B, Pulsed wave Doppler demonstrating coronary flow entering the right ventricle. Sample volume is on the right ventricular side of the aneurysm. Flow is toward the transducer, and therefore above the baseline. Note continuous flow that peaks in early to mid-diastole.

hospital with an acute myocardial infarction. New systolic and diastolic murmurs were heard on admission. His course was complicated by pulmonary edema, atrial fibrillation, and ventricular tachycardia. Two weeks following discharge he developed nausea and lightheadedness; the ECG was normal except for the presence of atrioventricular junctional rhythm at a rate of 47 bpm and new inferior T wave inversions in leads II, III, aVF. The systolic and diastolic murmurs persisted. He was admitted to our institution, and on physical examination he had a blood pressure of 170/90 mm Hg and a pulse of 90 bpm. Coarse wet rales were audible over the posterior lung bases. The jugular venous pressure was 12 cm. The apex beat was at the anterior axillary line and was sustained. A left parasternal lift was also present. The first and second heart sounds were normal. Third and fourth heart sounds were