Cardiac pheochromocytoma

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Thoracic pheochromocytomas account for less than 1% of reported cases and are usually located in the posterior mediastinum when they occur in the chest. Primary cardiac pheochromocytomas (PCTs) are rare, with fewer than 50 cases reported in the literature. The following case study illustrates the presentation of a patient with a PCT.

Clinical Summary

A 25-year-old man arrived at the hospital with dyspnea and symptoms consistent with New York Heart Association class II heart failure. He denied chest pain. His medical history was significant for hypertension, for which he was receiving β -blockers. Physical examination was remarkable for an S3 gallop murmur. The blood pressure was 213/150 mm Hg by means of cuff bilaterally.

Findings on chest radiography were consistent with interstitial edema and cardiomegaly. An echocardiogram showed a right atrial mass, a dilated and hypertrophied left ventricle, and severe mitral regurgitation. He was admitted for blood pressure control and evaluation.

A contrast computed tomographic scan of the chest showed the mass to be abutting the right atrium, although it was unclear whether the mass was extrinsic to the myocardium. A gated magnetic resonance study was performed in the axial and coronal planes through the right atrial region. The T1-weighted images showed an oval mass along the wall of the right atrium. The lesion indented the wall and was associated with the interatrial septum (Figure 1); it appeared fairly well demarcated from the adjacent tissues and did not appear to be invading the right atrium. The lesion demonstrated slightly increased signal relative to the myocardium on the T1-weighted images and a further increase in signal on the T2-weighted images (Figures 1 and 2). The signal characteristics of the mass favored a vascular lesion, making PCT the likely diagnosis on the basis of the clinical presentation and imaging features, and this was further confirmed by increased 24-hour urine norepinephrine and normetanephrines and plasma-free normetanephrines.

Cardiac catheterization demonstrated the mass with feeding vessels from the right coronary artery. Once the patient's blood pressure was controlled with α - and β -blockade, he consented to

Figure 1. T1-weighted axial image through the somewhat triangular mass (*asterisk*) demonstrates its signal to be slightly higher than myocardium or skeletal muscle. Note the relationship of the mass to the interatrial septum (*arrows*). RA, Right atrium; LA, left atrium; RV, right ventricle.

Figure 2. T2-weighted axial image at the same level as Figure 1. A. Mass (*asterisk*) now shows significantly greater signal than myocardium and skeletal muscle. RA, Right atrium; LA, left atrium; RV, right ventricle; LV, left ventricle.

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mediastinal exploration and excision of a cardiac tumor on cardiopulmonary bypass.

The patient was started on bypass with bicaval cannulation. The mass was densely adherent to the right atrium. Its borders grossly were the superior vena cava, the right atrium–inferior vena caval junction, and the interatrial septum. With gentle manipulation of the

mass, the patient's mean arterial pressure increased to 140 mm Hg. The aorta was crossclamped, and the heart was arrested and emptied. The mass was resected, and the defect in the right atrium was closed with a pericardial patch. The patient was weaned easily from cardiopulmonary bypass. Norepinephrine levels taken from a coronary sinus catheter before and after resection went from 859 to 249 pg/mL.

The patient's course was uncomplicated, and he was discharged on postoperative day 5. His only antihypertensive medication at discharge was an angiotensin-converting enzyme inhibitor. Histopathologic examination demonstrated a 5 cm × 5 cm × 3 cm PCT.

Discussion

PCTs are tumors of the sympathetic nervous system arising from chromaffin cells. In adults the majority of these masses are located in the adrenal medulla, where chromaffin cells are concentrated. Ten percent occur in extra-adrenal locations, with the organ of Zuckerkandl being the most common. Chromaffin cells can also be found in the walls of blood vessels, along the aorta, and in the heart, prostate, and ovaries. PCTs produce large amounts of catecholamines, particularly norepinephrine. These tumors occur in less than 0.1% of hypertensive patients.

In a review of 30 cases,¹ the most common presentation of PCT was hypertension, and dyspnea accounted for only 3 cases. Although our patient was hypertensive at the time of presentation, it was his symptoms of heart failure that prompted the echocardiogram. On magnetic resonance imaging, its signal characteristics suggested a vascular lesion, such as angiosarcoma, hemangioma, or PCT.^{2,3}

Coronary angiography is recommended before resection. At least one case reported reimplantation of a coronary artery.¹

Most cardiac PCTs are densely adherent to underlying tissues. We and others recommend cardiopulmonary bypass and aortic cross-clamping for resection.⁴ Manipulation of the tumor can cause marked hypertension, even if preoperative α - and β -blockade is performed. Wilson and colleagues⁵ described systolic blood pressures of greater than 300 mm Hg when a PCT was resected through a right thoracotomy with no bypass. Four deaths have been described in a review of 25 patients, all resulting from massive hemorrhage.

Conclusion

Cardiac PCTs represent interesting diagnostic and treatment dilemmas. Although rare, they can be treated for cure in more than 90% of cases. Magnetic resonance imaging is the noninvasive test of choice. Cardiac catheterization is recommended and aids in surgical management. These authors advocate excision with cardiopulmonary bypass and cardioplegic arrest.

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Internal thoracic artery injury after transvenous pacemaker implantation

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Injuries associated with transvenous insertion of pacemaker leads occur infrequently. Common complications include pneumothorax-hemothorax, subclavian vein and artery injury, and myocardial perforation.¹ We describe, for the first time, a laceration of the left internal thoracic artery (LITA) diagnosed 2 days after pacemaker implantation.

Clinical Summary

A 71-year-old man with a medical history significant for sick sinus syndrome and atrioventricular nodal conduction system disease underwent an insertion of a dual-chambered pacemaker. Screw-in pacing leads were placed transvenously and fixed in the right atrial appendage and right midventricular septum. The generator was implanted in a subcutaneous pocket in the left side of the chest. Pacing thresholds were adequate. Chest radiography revealed both leads to be in good position. The patient was discharged home the next day.

Thirty-six hours later, the patient presented in respiratory distress with left-sided chest pain. Initial systolic blood pressure was 70 mm Hg. Hemoglobin was 10.1 g/dL. Cardiac enzymes were within normal limits. Chest radiography showed opacification of the left hemithorax. Echocardiography showed no pericardial effusion. Computed tomography demonstrated a left hemothorax with extrusion of the pacing lead into the left hemithorax (Figure 1).

Tube thoracostomy was performed, and 1.5 L of blood was drained. Subsequently, the patient bled about 400 mL/h and therefore underwent emergency thoracotomy. An actively bleeding

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