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Mediastinal Pheochromocytoma with Single Coronary Blood Supply: A Case Report

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ABSTRACT

Primary pheochromocytomas located outside the adrenal glands account for only 10% of all pheochromocytomas. Mediastinal pheochromocytomas are even rarer and usually represent a therapeutic challenge as they often infiltrate adjacent structures. We report the case of a large primary mediastinal pheochromocytoma in a 65-year-old patient presenting with a sudden angina-like chest pain and dyspnea. Thoracic multislice computed tomography showed an 8 × 5 × 6–cm retrocardiac mass causing compression of both atria and infiltrating the left superior pulmonary vein. The tumor was highly vascularized and presented a blood supply derived from the circumflex artery. The mass was successfully removed by open heart surgery, and the patient was discharged 10 days postoperatively.

BACKGROUND

Primary cardiac tumors are rare clinical entities, accounting for 0.0017% to 0.28% of cases in autopsy series. The majority of these tumors are benign, and atrial myxoma is the most frequent [Gowdamarajan 2000]. Pheochromocytomas are catecholamine secreting tumors arising from chromaffin cells usually located in the abdomen. These tumors are rare, occurring only in 0.3% to 0.95% of cases in autopsy series [Atiyeh 1997]. Pheochromocytomas located outside the abdomen are even rarer, constituting 1% to 2% of cases; they are often located in the posterior mediastinum and can be named paragangliomas [Pickering 2000]. We present a successfully treated case of a mediastinal paraganglioma compressing both atria and pulmonary arteries as well as infiltrating the pulmonary veins; the patient initially presented with atypical chest pain.

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CASE REPORT

A 65-year-old male patient was referred to a local cardiology department with angina-like chest pain and effort dyspnea. The patient presented no previous clinical record. Physical examination was unremarkable. Resting 12-lead electrocardiography showed sinus tachycardia without ST segment elevation; symmetric T-wave inversion in leads V3 through V5 was observed. Laboratory parameters showed a maximum creatinine kinase of 831 UI/L, leading to a clinical diagnosis of a non–ST elevation myocardial infarction. Transthoracic echocardiography showed a retrocardiac mass of approximately 6 × 4 cm compressing the left atrium as well as causing a light hypokinesia of the apical anterior and posterior wall; the ejection fraction was preserved (60%). The coronary angiogram showed no intrinsic obstruction of the coronary circulation. An aberrant vessel arising from the circumflex artery to a highly vascular retrocardiac mass was observed (Figure 1). The patient was transferred to our department for further assessment. Hematological and biochemical investigations including neuroendocrine tumor markers and urinary catecholamine levels were performed. Values of chromogranine A (1018 ng/mL; normal, <98 ng/mL) and neuron-specific enolase (NSE) (18.1 ng/mL; normal, 0-16.3 ng/mL) were elevated. The urinary catecholamines and their metabolites (vanillin mandelic acid, 16.2 mg/24 h; normal, <7.5 mg/24 h) were markedly elevated, suggesting a pheochromocytoma.

Thoracic examination by multislice computed tomography (MSCT) showed an 8.5 × 5.5–cm retrocardiac mass compressing the left atrium and infiltrating the left inferior pulmonary vein (Figure 2A). However, the relationship between the tumor and the pericardium was unclear. Magnetic resonance imaging (MRI) showed compression of both atria, infiltration of the pericardium and left pulmonary veins, and contact with the pulmonary arteries and trachea. T2-weighted sequences showed mild signal intensity, which increased in the T1-weighted sequences; the signal was more elevated in the tumor perfusion (Figure 2B).

Once the patient was prophylactically controlled with α- and β-blockade, surgical excision of the tumor through median sternotomy was performed. The patient was put on

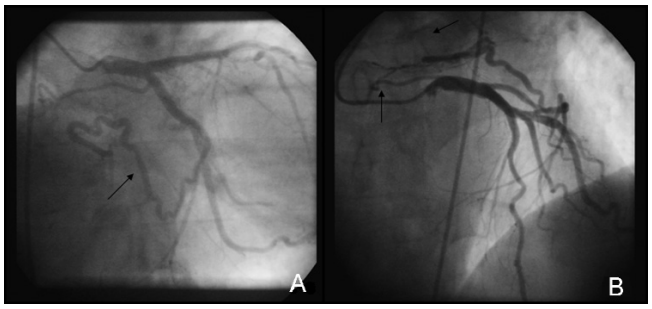

Figure 1. A, Zero degree left anterior oblique projection demonstrating the aberrant vessel arising from the circumflex artery (arrow). B, Thirty degree left anterior oblique projection depicting the highly vascular mass with its "tumor blush" phenomenon (arrows).

cardiopulmonary bypass (CPB) with bicaval cannulation, the aorta cross clamped, systemic cooling to 30° instituted, and the heart arrested with cold cardioplegia. Macroscopically, the tumor adhered to the roof of the left atrium and touched the right atrium and the pulmonary arteries. The aorta, main pulmonary artery, and superior vena cava were cross sectioned to gain access to the posterior mediastinum (Figure 3A). The 8.5 × 5.5–cm tumor was then meticulously dissected from the pericardium, pulmonary vessels, left atrium, esophagus, and trachea. After ligation of the vascular coronary supply, the tumor was resected en bloc (Figure 3B). After restoration of the sectioned vessels, the cross clamp was removed and the patient uneventfully weaned from CPB. Histopathological examination of the resected mass showed macroscopically a highly vascular tumor with a cystic core (Figure 3B). Microscopically, the tumor showed irregular cells of variable size with an eosinophilic cytoplasma and pleomorphic nuclei. The cells were grouped in nests divided by thin trabecular septa without increased mytotic activity. Immunhistochemical study showed a positive reaction to NSE, synaptophysin, and chromogranin.

The postoperative course was uncomplicated and the patient was discharged on the tenth day after his operation. At 6 months, he remains normotensive and symptom free.

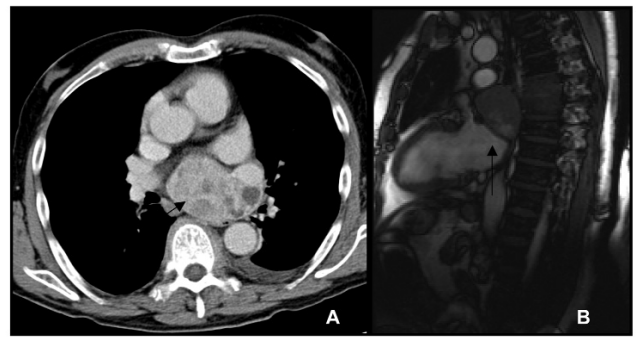

Figure 2. A, Thoracic multislice computed tomography showing a heterogenic retrocardiac mass in contact with the left atrium and the descending aorta. B, Lateral projection of the mediastinum by magnetic resonance imaging. The left atrium is compressed by the tumor.

DISCUSSION

Pheochromocytoma is a catecholamine-producing tumor arising from the sympathetic nervous system [Atiyeh 1997]. Pheochromocytomas are located in the adrenal medulla in 95% of cases and only 1.5% to 2% are situated above the diaphragm [Fitzgerald 1995]. Pheochromocytomas of the mediastinum occur in 75% of cases and are located in the posterior mediastinum, usually arising from the paravertebral sympathetic ganglia. Tumors from the middle mediastinum are rare and arise either from the aorto-pulmonary paraganglia or from the visceral autonomic paraganglia of the heart. Pheochromocytomas can present with constitutional symptoms, weight loss, and fever; however, the classical presentation includes hypertension, sweating, headache and palpitations. Our patient presented in an unusual way with chest pain and a non–ST elevation myocardial infarction. The tumor's feeding vessel arose from the circumflex artery, probably causing a transient ischemia of its territory because of a stealing phenomenon. Although the catecholamine and their metabolite levels in urine were elevated, none of the significant symptoms of increased production of catecholamines were detected. Echocardiography helped us with the preoper-

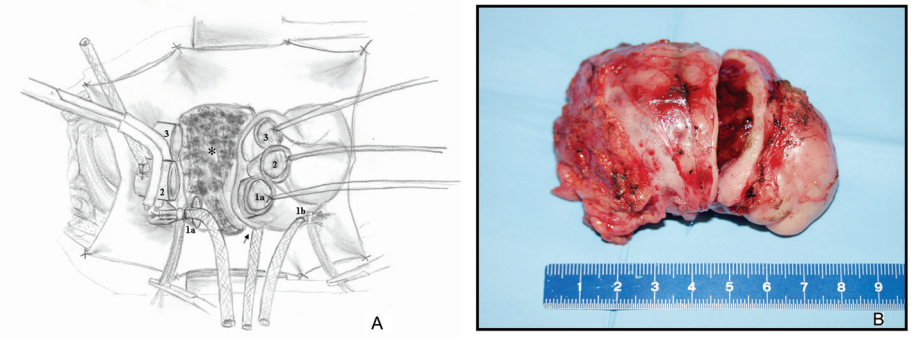

Figure 3. A, Intraoperative view of the retrocardiac tumor (*). The vessels were dissected and the heart was positioned forward for optimal visualization of the mass. 1a indicates the superior cava vein; 1b, inferior cava vein; 2, aorta; 3, pulmonary artery. B, Macroscopic image of the 8.5 × 5.5–cm explanted tumor.

ative assessment of the patient by providing us with information about the localization of the tumor and the ejection fraction of the patient. However, MSCT and MRI allowed us to differentiate between the structures adjacent to the tumor. These imaging techniques are useful in the management of such a patient, as they allow planning of the surgical strategy [Hartgrink 2001]. Although we employed MSCT to localize the mass, MRI was found to be more sensitive as it gave more information about the vascular status of the tumor and it permitted a better differentiation of the soft tissues because of its greater resolution. Arteriography demonstrated the feeding vessel and the relationship between the tumor and the coronary arteries. The absence of coronary disease also suggested the relationship of the tumor vascularization and the ischemia in the circumflex artery territory.

Complete surgical excision is the treatment of choice, although mediastinal pheochromocytomas are often difficult to remove because of the infiltration of adjacent structures. Cardiac reconstruction or even orthotopic transplantation may be necessary after removal [Turley 2005]. Furthermore, a rigorous management is necessary to prevent intraoperative hypertension. The patients should undergo an adequate α- and β-blockade before manipulation of these tumors. Our patient did not present serious intraoperative hypertension after α- and β-blockade. Resection was performed with safety surgical margins under CPB. Cross sections of the aorta, the pulmonary artery, and the superior vena cava were performed to access the posterior mediastinum, permitting the safe dissection of the tumor with its capsule. Such a procedure was necessary to avoid damage to vital structures and to minimize the intraoperative risk.

Pheochromocytomas are able to synthesize different peptides, including synaptophysin, chromogranin A, and NSE. Synaptophysin is the best neuroendocrine marker for this type of tumor; however, the presence of NSE suggests its benign origin [Moreno 1999]. Our patient presented elevated plasma values of chromogranin A and NSE. Furthermore, all of these peptides were detected in tumor biopsies by means of immunohistochemistry.

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