Primary Cardiac Paraganglioma With Intratumoral Coronary Artery Aneurysm

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We report here the surgical treatment of a rare case of primary cardiac paraganglioma with an intratumoral coronary artery aneurysm.

Primary cardiac tumors are very rare, with a reported prevalence of less than 0.05% in patients who die of malignancy; and cardiac paragangliomas account for less than 1% of cardiac tumors [1]. Fewer than 100 cases of cardiac paraganglioma have been reported in the literature to date, and only one case of cardiac paraganglioma associated with a coronary artery aneurysm and arteriovenous fistula has been reported. Primary cardiac paraganglioma with an intratumoral coronary artery aneurysm but no arteriovenous fistula has not previously been reported.

A 42-year-old Chinese woman was referred to our center with shortness of breath after exertion, a nonproductive cough, and atypical chest discomfort. She had no history of hypertension or hyperhidrosis. Her vital signs and the results of physical examination were normal. A 12-lead electrocardiogram showed normal sinus rhythm with no ST segment or T wave abnormalities. Transthoracic and transesophageal echocardiography showed a heterogeneous, medium echogenicity, well-vascularized, clearly demarcated mass measuring 10 \times 8 \times 6 cm in the left atrioventricular groove. Computed tomography (CT) angiography of the chest showed a heterogeneous, medium echogenicity, well-vascularized, clearly demarcated mass measuring 10 \times 8 \times 6 cm in the left atrioventricular groove. Computed tomography (CT) angiography of the chest showed a heterogeneous mass with a low-density center attached to the left atrioventricular wall. The left coronary artery (LCA) was significantly dilated and supplied several small branches to the mass. There was a 3 \times 1.5 \times 1.5 cm aneurysm of one of the branches of the LCA in the center of the mass (Fig 1). Coronary angiography confirmed a vascular pericardial mass supplied by the circumflex branch of the LCA. Magnetic resonance imaging (MRI) of the chest showed a fibrous capsule surrounding the tumor and a tiny connection of fatty tissue between the mass and the left atrioventricular wall (Fig 2). CT of the abdomen, pelvis, and adrenal glands did not show any abnormalities. The operation was performed through a median sternotomy with the patient under hypothermic cardiopulmonary bypass. After incision of the pericardium, a dark red, encapsulated, intrapericardial tumor measuring 10 \times 8 \times 6 cm was observed on the outer aspect of the left atrioventricular wall (Fig 3). The entire tumor was carefully resected, including the coronary artery aneurysm. Histologic examination of the specimen with hematoxylin and eosin staining revealed polygonal cells with pleomorphic nuclei (Fig 4). Immunohistochemical staining showed that the sustentacular cells were positive for S-100 protein (Fig 5). The tumor cells were strongly positive for chromogranin (Fig 6). These findings were consistent with primary cardiac paraganglioma. After the operation, the patient recovered well and returned to her normal activities, with no evidence of abnormal blood pressure.

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Comment

Paragangliomas are rare tumors originating from the neural crest cells, most commonly in the adrenal medulla. Only 1% to 2% of these tumors occur in the chest, most commonly in the posterior mediastinum. Cardiac paragangliomas are extremely rare. The most common site of cardiac paraganglioma is the roof of the left atrium, possibly explained by the close proximity of paraganglionic cells to the left atrium. However, paragangliomas can develop in any of the cardiac chambers, or at the base of the aortic root or pulmonary artery [2]. They occur mainly in adults aged between 30 and 60 years and are more common in women than in men [3]. Cardiac paragangliomas are classified histologically as chromaffin (catecholamine secreting) or nonchromaffin (non–catecholamine-secreting) tumors [4]. Patients with the former type may present with hypertension, sweating, and palpitations; those with the latter type may present with cardiac tamponade, superior vena cava syndrome, intracardiac flow obstruction, dyspnea, hoarseness, dry cough, or dysphagia because of the local mass effect or invasion. Rarely, paragangliomas are asymptomatic and are detected incidentally on cardiac imaging examinations. Cardiac paragangliomas are highly vascular tumors and are commonly fed by branches of the LCA, although...
dual vascularization from the right and left coronary systems may also occur. Echocardiography is an important modality for detecting cardiac tumors. CT angiography of the chest and coronary angiography are useful for investigation of these highly vascularized tumors because they enable analysis of the feeding vessels of the tumor and their relationships with the epicardial arteries, which is important for operative planning. MRI is also important for operative planning. Most paragangliomas are hyperintense on T2-weighted images, which helps to differentiate them from the surrounding cardiovascular structures. Benign and malignant paragangliomas are histologically indistinguishable. Most authors agree that malignancy can be confirmed only by the demonstration of metastasis to the lymph nodes, lungs, or bones. However, there is a relationship between tumor size and malignant behavior. There is consensus in the literature that cardiac paragangliomas should be resected through a median sternotomy with the support of extracorporeal circulation. Successful management depends on complete resection of the tumor because incomplete resection leads to recurrence and metastasis. Removal of the tumor requires reconstruction of local anatomic structures. Radiation therapy and chemotherapy are not useful alternatives to complete resection. After experiencing this case, we conclude that the use of multiple imaging modalities, including echocardiography, CT angiography, coronary angiography, and MRI, is important for operative planning in patients with primary cardiac paraganglioma and that the best treatment is en bloc resection.

References