Intrapericardial Paraganglioma With Intratumoral Coronary Arterial Aneurysm and an Arteriovenous Fistula

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The authors present the case of a 55-year-old woman with a nonfunctioning intrapericardial paraganglioma. The tumor was incidentally found during transthoracic echocardiography that was performed because an enlarged cardiac silhouette had been noted on a chest x-ray. Transesophageal echocardiography further elucidated that the left circumflex coronary artery branched and traversed into the intrapericardial mass. Coronary angiography and cardiac multislice computed tomography both confirmed the presence of a vascular intrapericardial mass being fed by the obtuse marginal artery. Moreover, an arterial aneurysmal formation and a coronary arteriovenous fistula were noted within the tumor. After "en bloc" tumor resection, histopathologic and electron microscopic examinations established the diagnosis of a nonfunctioning intrapericardial paraganglioma. (J Am Soc Echocardiogr 2009;22:211.e1-e3.)

Keywords: Paraganglioma, Intrapericardial tumor, Coronary arterial aneurysm, Coronary arteriovenous fistula

Intrapericardial paraganglioma is a rare type of neural tumor of the extra-adrenal parasympathetic system. Fewer than 50 such cases have been reported thus far, and coronary arteriovenous fistulas (AVFs) within the tumors were not noted in any of these cases. Transesophageal echocardiography (TEE) can depict cardiac masses with superior resolution and can thus improve the preoperative diagnostic accuracy for such tumors. Herein, we report a unique case of an intrapericardial paraganglioma containing an intratumoral coronary arterial aneurysm and a coronary AVF.

CASE REPORT

A 55-year-old woman with a history of diabetes mellitus presented with multiple liver abscesses. Sixteen years earlier, she had undergone emergency pericardial drainage with bloody effusion for cardiac tamponade and had been treated for tuberculous pericarditis.

Clinical examination results were unremarkable, except for the presence of fever and abdominal discomfort in the right upper quadrant. Abdominal sonography revealed the presence of multiple abscesses in both hepatic lobes. An enlarged cardiac silhouette was noted on a chest x-ray. Transthoracic echocardiography detected a large, solid, elastic intrapericardial mass abutting the left lateral LV wall, positioned lateral and adjacent to the atrioventricular groove, left atrium, and basal left ventricular (LV) wall. Further, a small amount of pericardial effusion and 2 intratumoral hyperechoic flecks were noted (Figure 1).

TEE performed in the color Doppler mode displayed a prominent eccentric compression of the coronary artery by the mass. Further, 1 major branch of the artery traversed intramurally, with noticeable rapid turbulent flow. Another dilated vascular structure was observed toward the center of the tumor, along with 2 tiny spots of calcification (Figure 2, Video 1).

Cardiac multislice computed tomography (MSCT) revealed a 6.2 × 5.4 × 5 cm intrapericardial enhanced mass with progressive centripetal fill-in enhancement, a possible AVF between the second obtuse marginal (OM2) artery and the coronary sinus suggested by a high-flow jet of contrast medium into the right atrium, and intratumoral aneurysmal dilatation of the distal OM2 artery. The mass abutted the basal anterolateral wall of the left ventricle. Central necrosis and spotted calcification were observed within the tumor (Figure 3). Coronary angiography disclosed a pericardial mass with a vascular blush; the mass was fed by the dilated OM2 artery branching from the left circumflex coronary artery. The OM2 artery crossed intramurally with distal arterial aneurysm formation that drained into the great cardiac vein. These findings were consistent with those of intratumoral coronary AVF (Figure 4, Video 2).

The liver abscesses were treated with antibiotics and percutaneous drainage. The patient subsequently underwent open-heart surgery. During this surgery, a large, solid, elastic intrapericardial mass abutting the basal lateral LV wall was successfully removed by delicate resection and partial myomectomy. Gross examination revealed that the mass was supplied blood by numerous small feeding vessels and that the engorged OM2 artery cut across the mass and connected to the great cardiac vein. Histologic microscopic examinations revealed that the tumor had polygonal cells with pleomorphic nuclei on hematoxylin-eosin staining. Immunohistochemical staining was positive for S-100; numerous...
dense-core neurosecretory granules and mitochondria were found on electron microscopy, which established the diagnosis of a non-functioning paraganglioma. The patient recovered well postoperatively.

DISCUSSION

Paragangliomas, first described by Kohn in 1900, are histologically classified as pheochromocytomas (secreting chromaffin and catecholamine) and chemodectomas (not secreting chromaffin or catecholamine). They have been reported to occur in a wide variety of locations throughout the body. However, intrapericardial paragangliomas are among the rarest of these tumors, despite the presence of some neuroepithelial cells within the pericardium.

Intrapericardial paragangliomas are usually large (3-8 cm) and commonly localized close to the left atrium. Myocardial invasion, noted in approximately 50% of the reported cases, can be found at the interatrial septum, right atrium, interventricular groove, aortic root, and LV wall. Intrapericardial paragangliomas are generally hypervascular and prone to fatal hemorrhagic complications.

TEE depicted a well-demarcated epicardial mass and provided a better delineation of an engorged eccentrically displaced coronary artery that branched into the tumor associated with intraluminal...
turbulent mosaic color flow. This intratumoral artery corresponded to the OM2 artery with a traversing pattern demonstrated by cardiac MSCT and coronary angiography. In addition, TEE identified the presence of 2 tiny spots of calcification toward the center of the tumor; this detection was congruent with findings on cardiac MSCT. Although calcification may occur in extra-adrenal paragangliomas, no such calcification pattern has been described for intrapericardial paragangliomas.

An intratumoral AVF was identified on coronary angiography but not on echocardiography. Coronary AVF is an uncommon congenital vascular anomaly. However, the coronary AVF in our patient may have been of an acquired nature. The location of the fistula tract suggested that if the fistula were a congenital AVF, it would have been extratumoral rather than intratumoral. Complications of the coronary AVF (eg, myocardial perfusion abnormality, heart failure, and endocarditis) and acquired coronary arterial aneurysm (eg, distal embolization of the clot, resulting in myocardial ischemia or infarction) were not present in this patient. The blood vessels (or coronary AVF) that supply tumor cells commonly appear in the form of highly concentrated, dilated vessels, as seen in our patient. The application of echo contrast enhancement could potentially indicate the vascular nature of the tumor, thereby obviating the radiation dosage inherent in MSCT.

To our knowledge, this is the first report of a nonfunctioning intrapericardial paraganglioma containing a coronary aneurysm and a coronary AVF. The patient underwent open-heart surgery with successful complete tumor resection. Furthermore, the patient exhibited good postoperative recovery.

REFERENCES