The Intra-Pericardial Paraganglioma Presenting as Ascites and Hemopericardium with Impending Tamponade

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Summary

The intra-pericardial paraganglioma is very rare and most of them present with hypertension or palpitations. Here we reported an extraordinarily rare case of intra-pericardial paraganglioma presenting as faint, pitting edema, abdominal fullness with ascites, and hemopericardium with impending tamponade, which was treated successfully by emergent pericardiocentesis and surgical resection.

Key words: Ascites, Hemopericardium, Paraganglioma, Tamponade

Paraganglioma, also being referred to as pheochromocytoma when functionally active, is a rare neoplasm of neural crest-cell origin. The clinical presentation depends upon the site of origin and functional status of these tumors. Functionally active tumors present with symptoms of catecholamine excess and functionally inactive tumors present with associated symptoms depending on the obstruction of organs. Magnetic resonance imaging (MRI) and computed tomography (CT) provide excellent anatomic information about tumors, and radionuclide imaging has very high specificity and is useful for localization of the functionally-active tumor.

The only curative treatment is complete surgical resection. In the literature, most cardiac paragangliomas are large, ranging in size from 3 to 8 cm and about 20% of them are functionally inactive. Most patients with functionally-inactive cardiac paraganglioma present with chest pain and dysphagia due to obstruction. It is rare to present with symptoms and signs, such as heart failure and hemopericardium. Here we reported a case with intra-pericardial paraganglioma presenting as faint followed by collapse, pitting edema, abdominal fullness with ascites, and hemopericardium with cardiac tamponade.

Case Report

A 63-year-old female presented to the emergency department with faint followed by collapse, progressive dyspnea, pitting edema, abdominal fullness, and 5-kg weight gain in two weeks. Because of faint followed by collapse at home, she visited another hospital where right subphrenic effusion and mild to moderate pericardial effusion were disclosed in abdominal CT. Diuretics were prescribed. Because of persistent symptoms, she was sent to our emergency department (ER) for help. At ER, blood pressure 148/72 mmHg and regular heart beats with tachycardia (110 beats per minute) were noted. Electrocardiogram revealed sinus tachycardia without any ST segment or T-wave abnormalities. Laboratory data showed normocytic anemia (hemoglobin = 10.7 g/dL), CRP 2.27 mg/dL, and NT-pro-BNP 110.2 pg/mL. Chest CT showed that one tortuous and engorged branch from left main coronary artery traveled to a complex vascular structure which was anterior to the aorta with some calcification besides ascites and marked hemopericardium (up to 2 cm in thickness) (Figure 1). Coronary arteriovenous malformation or hypervascular tumor with bloody leakage was highly suspected. Bedside echocardiography showed massive pericardial effusion with compression on the right ventricle and, due to impending cardiac tamponade, emergent therapeutic pericardiocentesis was done and 400 mL bloody pericardial effusion (hematocrit = 20.4%) was drained out. Unfortunately, no catecholamine level was available.

Initially, we highly suspected coronary arteriovenous malformation or hypervascular tumor with bloody leakage. However, due to the abnormal anatomy of feeding artery and hemopericardium, we suspected specific etiologies, and thus we arranged coronary angiography (CAG) and coronary computed tomography angiography (CTA).

The finding of CTA clarified the anatomy of feeding artery and guided our surgical strategies. Coronary angiography showed insignificant coronary artery disease but a large hypervascular mass with a feeding artery from left...
A tortuous and engorged coronary artery traveling to a complex vascular structure with hemopericardium is showed in transverse plane (A-B), coronal plane (C), and 3-D reconstruction of chest CT (D). A hypervascular mass with a feeding artery from left coronary artery is showed in coronary angiogram (E-H).

coronary artery (Figure 1). Gross appearance of specimen showed oozing with bloody leakage from the surface of the hypervascular mass and tortuous feeding vessels. The tumor tight adherent to aortic root, main pulmonary artery, coronary artery, and ascending aorta was removed under cardiopulmonary bypass via the femoral artery and vein (Figure 2). The feeding artery was ligated and ascending aorta replaced with a 26-mm Dacron graft. No significant change of blood pressure was noted intra-operation. The patient was discharged uneventfully several days later. The follow-up CT showed no evidence of residual tumor and pericardial effusion.

A picture of paraganglioma with nesting (zellballen) pattern of cells within a prominent vascular network was noted in the pathological finding. Sustentacular cells are highlighted by S-100, and the tumor cells were diffuse positive for synaptophysin and SDHB/SDHA (succinate dehydrogenase subunit B and A) immunohistochemically (Figure 3). The Ki-67 labeling index was high, but no lympho-vascular or perineural invasion was noted.

Discussion

Primary cardiac tumors are very rare, and cardiac paragangliomas constitute less than 1% of all cardiac tumors. In 2004, WHO defined pheochromocytomas as tumors arising from the adrenal chromaffin cells, and paragangliomas as tumors derived from the extra adrenal sympathetic chromaffin (sympathetic PGLs) and/or parasympathetic tissue (nonchromaffin PGLs) of the head and neck (HNPGLs). Paragangliomas associated with the heart can sometimes be classified as intra-cardiac or intra-pericardial based on their site of origin. Intra-pericardial tumors arise from the ganglia associated with the aorta, pulmonary arteries or coronary arteries, while intra-cardiac tumors arise from the ganglia in the atrial walls.

The clinical presentation depends on the site of origin and the functional status of these tumors. Patients with functionally-active tumors present with the signs and symptoms of catecholamine excess, such as hypertension, headaches, sweating or palpitations. In contrast, functionally-inactive tumors present with the symptoms of heart failure or ischemia due to vascular compression and the obstruction of systemic, pulmonary or coronary blood flow. In the literature, about 10% of reported paraganglioma are clinically silent, and 20% of them are non-functioning and may present as a mass with symptoms related to compression of other organs, such as chest pain, wheezing, pitting edema, and associated compression signs.5,6
It is common for cardiac tamponade to result in heart failure and faint, but it is rare that intra-pericardial paraganglioma results in acute onset of cardiac tamponade, and is followed by the symptoms and signs of heart fail-
ure, ascites and faint. The possible mechanism is that high pressure results in bloody leakage (oozing) in the pericardium leading to impending tamponade, which results in right heart failure, ascites, and decreased venous return. The pathophysiologic factors mentioned above resulted in impaired heart performance and decreased cardiac output. The clinical presentations of this patient, such as pitting edema, abdominal fullness, and faint or syncope followed by collapse, are reasonable.

Regarding excellent imaging tools to diagnose paragangliomas, CT and MRI provide excellent information and, on imaging study with contrast, intra-pericardial paragangliomas have specific imaging features: well-defined; encapsulated heterogeneous masses; and occasionally internal calcification. On MRI, we noted high signal on T2-weighted image and isointense signal on T1-weighted image similar to muscle. In our patient, CTA was a key imaging examination and we believe CTA plays an important role in diagnosis of intra-pericardial paraganglioma.

Radionuclide imaging is an excellent tool for localization of neuroendocrine tumor. We often used I-123, I-131 or Indium-111 octreotide to detect paraganglioma. The sensitivity for intra-pericardial paraganglioma with MIBG screening is 75%, and with In-111 octreotide it is 100%. Therefore, In-111 octreotide is currently the first choice for nuclear medicine imaging of paraganglioma. However, because paraganglioma was not our initial differential diagnosis, therefore the finding of radionuclide imaging and catecholamine level were not available.

The only curative treatment for these tumors is complete surgical resection, which is often complicated by the possibility of serious hemorrhage due to local invasion, rich vascularity, and the proximity of the tumors to the great vessels. For patients with paragangliomas, it is important to recognize that the tumors are not well encapsulated and can be tightly adherent to or infiltrated into the wall of the aorta or cardiac chambers.

Our patient denied familial congenital disease and recent data suggest that familial and genetic mutations constitute up to 25%-50% of the disease population. Various syndromes have been found to be associated with the development of pheochromocytoma and paraganglioma. Immunohistochemistry showed positive for SDHB/SDHA of this patient. The issue of SDHx mutations is popular and germline mutations in the gene of succinate dehydrogenase subunit B (SDHB) are implicated in the development of malignancy (more than 30%) and worst prognosis after metastasis. It implies that the genetic mutations and malignancy are less likely in this patient.

In conclusion, the absence of symptoms of catecholamine excess makes it difficult to diagnose intra-pericardial paraganglioma. Similar to our patient therefore, intra-pericardial paraganglioma without symptoms of catecholamine excess should be considered in the differential diagnosis of a well-defined and hypervascular tumor with initial presentation showing hemopericardium with cardiac tamponade and heart failure.

Disclosures

Conflicts of interest: None.

References