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ADVANCED

CASE REPORT: CLINICAL CASE

Aortic Paraganglioma Masking as Intramural Hematoma

When You Hear Hoofbeats Think Zebras, Not Horses

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ABSTRACT

A 52-year-old woman presented dyspnea and angina. The computed tomography scan indicated an intramural hematoma, and the patient underwent surgery, during which a structure was excised that was identified as aortic paraganglioma. This case report underlines the importance of a multiprofessional interdisciplinary team to diagnose and treat cardiac masses. (**Level of Difficulty: Advanced.**) (J Am Coll Cardiol Case Rep 2023;15:101852)

Cardiac tumors may be classified as primary (originating from the heart) and secondary (ie, metastases) and typically manifest with dyspnea or arrhythmias, the most common type being myxomas. Nevertheless, diagnosis of cardiac tumors comprises a variety of imaging modalities, such as echocardiography, computed tomography (CT), or magnetic resonance imaging. Although the quality of imaging modalities has markedly increased over the past decades, a correct or definitive diagnosis may not be reached through radiological means in every case. This case report illustrates how an initial diagnosis of intramural hematoma prompting open-heart surgery was later revealed to be a rare case of aortic paraganglioma.

LEARNING OBJECTIVES

- To raise awareness for rare differential diagnoses in aortic pathologies.
- To identify treatment options for cardiac tumors causing dyspnea.

HISTORY OF PRESENTATION

A 52-year-old woman presented to the emergency department with dyspnea, New York Heart Association Functional Classification II, and angina. She was normotensive, and the initial electrocardiogram and laboratory results showed no abnormalities. Transthoracic echocardiography showed no valve pathologies but a broadened abnormal structure at the aortic root level, which led to the decision to perform a CT angiogram.

The CT angiogram revealed a 13-mm structure, possibly an aneurysm, right above the right coronary artery (Figure 1). This was diagnosed as an intramural

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ABBREVIATIONS AND ACRONYMS

CT = computed tomography

IMH = intramural hematoma

SDHB = succinate dehydrogenase subunit B

hematoma (IMH). Open-heart surgery was planned to replace the ascending aorta with a Teflon graft. Coronary angiography showed no signs of coronary heart disease but did demonstrate a depot of contrast agent in the ascending aorta, being suspicious for an aortic wall abnormality (Figure 2).

PAST MEDICAL HISTORY

Past medical history revealed a body mass index of 28 kg/m^2 and medications for hypothyroidism and arterial hypertension.

DIFFERENTIAL DIAGNOSIS

The differential diagnoses of IMH include acute aortic dissection and penetrating aortic ulcus. Usually, the diagnosis can be differentiated with an angiography CT scan.

INVESTIGATIONS

The results of complete blood laboratory testing were unremarkable. In addition, coronary angiography ruled out coronary artery disease (Figure 2). CT imaging indicated an aneurysmal structure within the ascending aorta (Figure 1). Preprocedural echocardiography also indicated an aortic aneurysm.

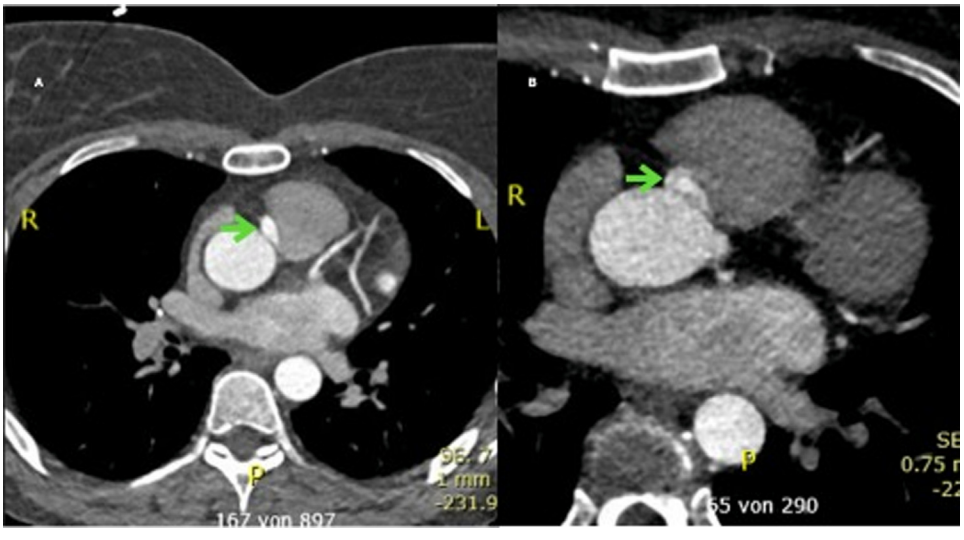
MANAGEMENT

After a diagnostic workup and suspected diagnosis of IMH, urgent cardiac surgery was planned with a potential replacement of the ascending aorta or Bentall's procedure. The procedure was performed with the patient under general anesthesia. Transesophageal echocardiography showed the suspected mass, indicating blood flow between the ascending aorta and the suspected hematoma above the right coronary ostium (Figures 3 and 4). After sternotomy and establishing a cardiopulmonary bypass, and the induction of cardiac arrest using cold blood cardioplegia (Buckberg), the aorta was opened and carefully inspected. The aortic valve was competent, and there were no visible signs of an IMH on the aortic intima or within the aortic wall. Further preparation of the para-aortic tissue revealed a 1.5 × 1.5 × 3.0-cm solid structure on the outer aortic wall that was excised (Figures 5A and 5B). The aorta was closed in the usual fashion, and cardiopulmonary bypass was weaned.

Postoperatively, the patient recovered well and was discharged without any further dyspneic symptoms or angina on the seventh postoperative day.

The mass was sent to the pathology lab, where an extra-adrenal paraganglioma was diagnosed. Performing immunohistochemistry, a loss of the

FIGURE 1 Preoperative Computed Tomography Scan



The **arrow** is pointing to the suspicious mass, later identified as a paraganglioma.

nuclear succinate dehydrogenase subunit B (SDHB) expression was apparent, being a surrogate marker for a loss of function mutation. Subsequent DNA-based parallel sequencing (TSO500 Panel, Illumina) revealed a pathogenic mutation of the SDHB gene (*p.Arg90Ter*). No further pathogenic mutations were found, and the tumor was tested as microsatellite stable with a low tumor mutational burden of 0.8 mutations per megabase (**Figure 6**).

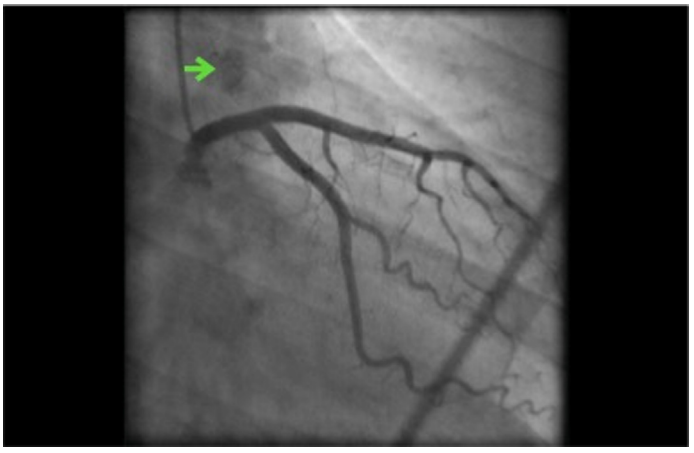
DISCUSSION

Cardiac tumors are rare, with described incidences between 0.17% and 0.19%. Most of these tumors are benign.[1] Paragangliomas arise from extra-adrenal chromaffin tissue and are classified based on histology, location, and innervation. Extra-adrenal manifestation may occur in patients with genetic disorders, such as Hippel-Landay syndrome, Carney-Sratakis syndrome, and specific subtypes of multiple endocrine neoplasia.[2] Paragangliomas located in the aortic body are included in the brachiomeric group. They are usually benign entities, but the presence of a constitutional SDHB mutation is the most potent genetic risk factor for metastasis development. Most of these tumors arise in the retroperitoneum, commonly the adrenal glands. Only 1% to 2% of paragangliomas occur in the thorax, with most occurring in the posterior mediastinum. Cardiac paragangliomas are exceptionally rare and may arise from any heart chamber or periaortic tissue. As such, the differential diagnosis of an aortic paraganglioma tumor adjacent to the aortic wall may include hemangioendothelioma, angiosarcoma, leiomyosarcoma, and sarcoma and may be classified as malignant aortic tumors.[3]

Most patients are asymptomatic, whereas some present with angina or dyspnea, as described in our case. Myocardial infarction is possible when the tumor involves the coronary circulation.[4,5]

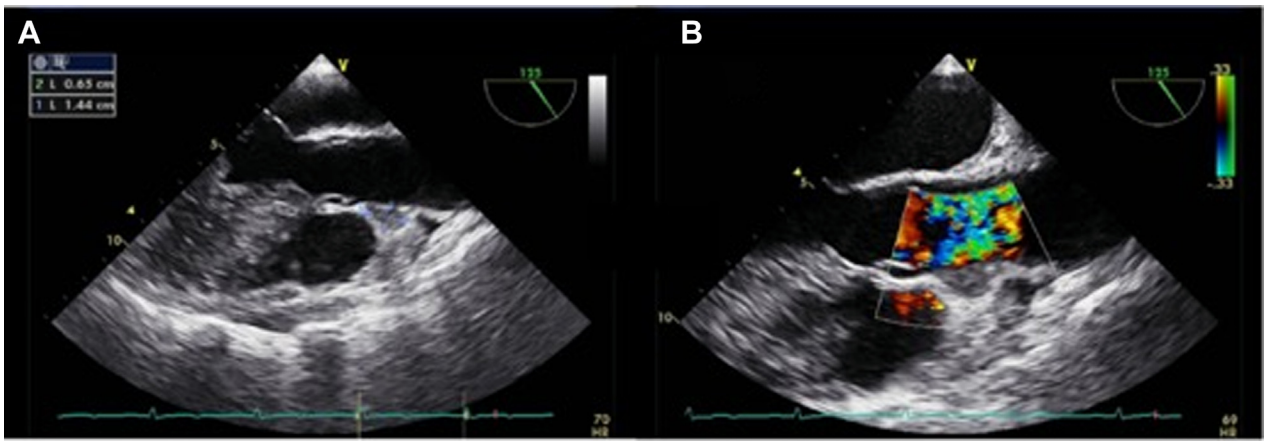
Surgical excision of cardiac tumors is the most effective therapy, and in some cases, may be combined with adjuvant radiation or chemotherapy.[6,7] Concerning neoadjuvant radiation therapy, the elevated risk for radiation-induced cardiac toxicity

FIGURE 2 Coronary Angiography



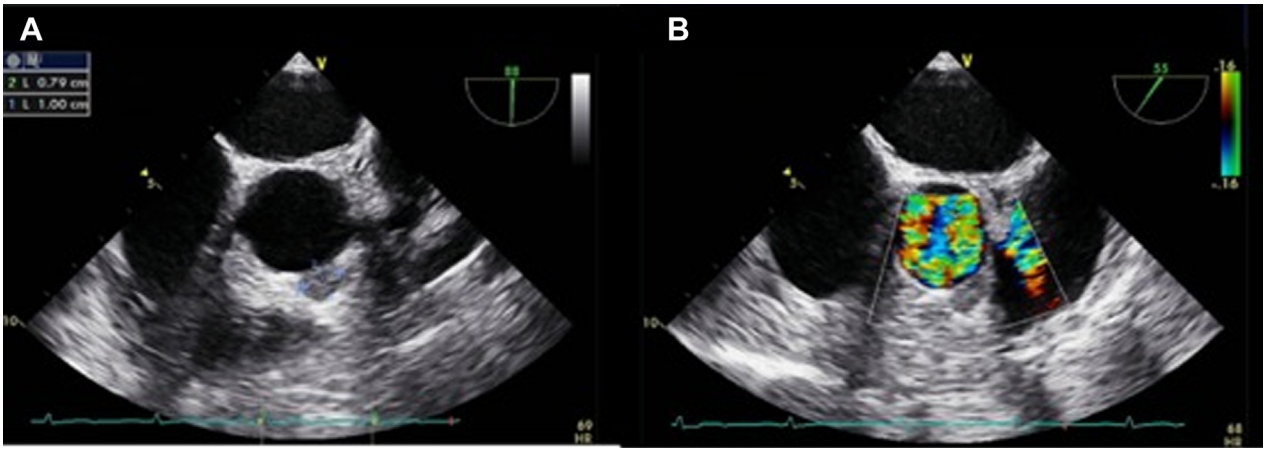
The **arrow** is pointing to the suspicious mass, later identified as a paraganglioma.

FIGURE 3 Preoperative Transesophageal Echocardiography



(A) Mid-esophageal Long axis view. **(B)** Mid-esophageal long axis view, color doppler.

FIGURE 4 Preoperative Transesophageal Echocardiography



(A) Mid-esophageal short axis view, measurements of the suspected mass **(blue marks)**. **(B)** mid-esophageal short axis view color doppler, measurements of the suspected mass **(blue marks)**.

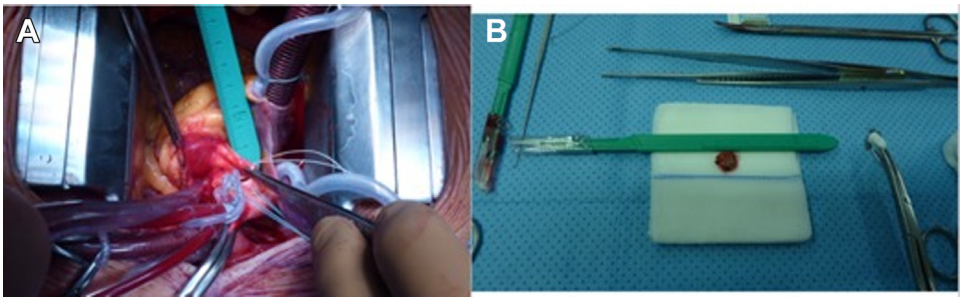
should be considered.[8] Given the rarity of cardiac tumors, particularly paragangliomas, a high degree of suspicion during initial diagnostic testing is needed to detect these tumors.

Only 10% of paragangliomas are considered malignant, but the histological differentiation between a malignant and benign paraganglioma is challenging. As such, the detection of potential metastases is the only reliable way to assess for malignant paragangliomas. Therefore, an additional positron emission tomography-CT was performed postoperatively to exclude potential metastases because the malignancy rate has been reported as high as 50% in some mutations, including the discovered SDHB mutation.[9]

Over the past decades, an increase in the incidence of cardiac tumors has been reported.[10] This may be due to recent improvements in imaging quality, mainly CT and magnetic resonance imaging. Further, advances in ultrasound techniques have led to high sensitivity in detecting even small cardiac masses. As clinical symptoms may be completely absent or inconclusive at best, cardiac tumors may be underdiagnosed or incorrectly attributed to alternative diagnoses (eg, infectious endocarditis), which adds to the importance of adjunct imaging modalities.

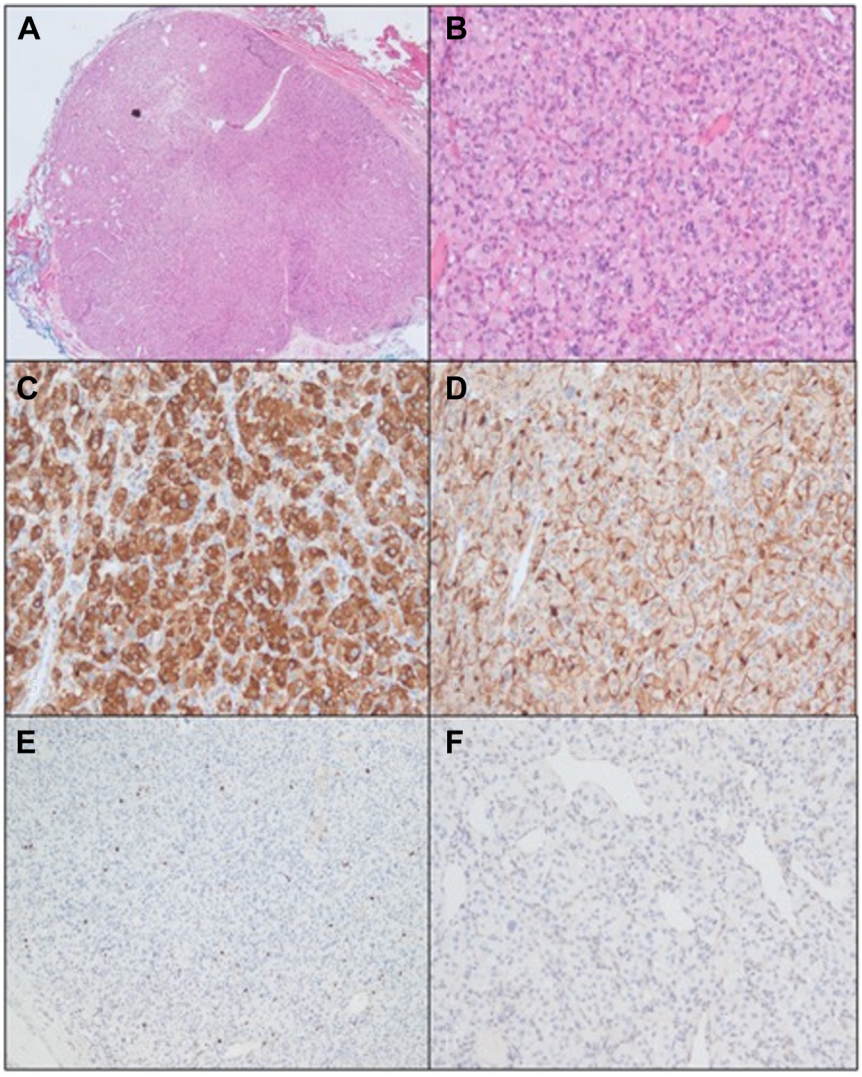
This case highlights several issues in the perioperative management of paragangliomas. First, it should be noted that even the use of several

FIGURE 5 Intraoperative View and Extirpated Paraganglioma



(A) Intraoperative view of the paraganglioma located at the outer aortic wall. **(B)** Excised paraganglioma.

FIGURE 6 **Immunohistochemical Imaging**



(A) Overview of the encapsulated paraganglioma (25x, HE) and **(B)** in more detail the histoarchitechture of the neoplasm (200x, HE). **(C)** Positivity of chromogranin A. **(D)** Strong positivity of S100 for the prominent sustentacular cells. **(E)** Ki67 staining reveals the low proliferative activity. **(F)** Loss of nuclear SDHB expression pointing to a possible mutation.

diagnostic modalities does not preclude a potentially grave misdiagnosis. Second, paragangliomas are often extensively vascularized, and thus biopsies may result in life-threatening bleeding.

Under these conditions, an open approach is indicated, but proper preoperative planning is crucial as resection can be technically challenging. In our case, preparations were made to replace the aortic root. However, the opening of the ascending aorta for inspection could have been avoided with the correct diagnosis and extra-aortic dissection and excision in the first place.

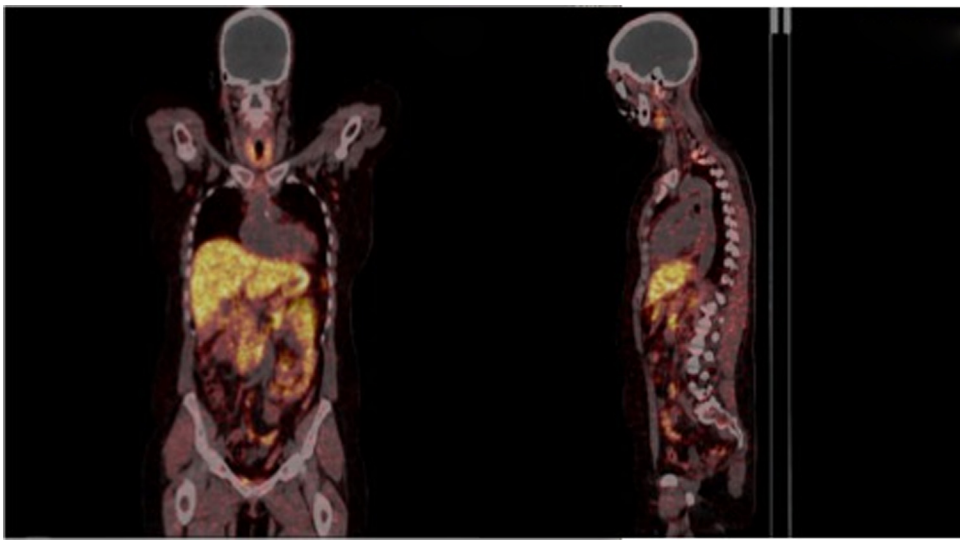
FOLLOW-UP

After surgery, the patient recovered well. A positron emission tomography-CT scan was performed (Figure 7) to evaluate for metastatic disease and showed no abnormalities 2 months after surgery.

CONCLUSIONS

We report a rare case of an aortic paraganglioma that was misdiagnosed as an IMH. Surgical excision of aortic paragangliomas may be challenging because of

FIGURE 7 Postoperative Positron Emission Tomography-Computed Tomography Scan



their vascularity and proximity to critical surrounding structures. Complete surgical excision is the treatment of choice and may benefit from management at experienced centers.

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