An unusual cause for syncope: Pericardial paraganglioma causing right ventricular outflow obstruction

Kailyn Mann, Mahek Shah, Naumann Islam, Ronald Freudenberger, Matthew Martinez, Larry Jacobs

ABSTRACT

Introduction: Metastases are the most common cardiac neoplasms with primary cardiac tumors being rare. Cardiac paragangliomas constitute <5% of primary cardiac tumors. They tend to remain asymptomatic until discovered incidentally or grow to a size large enough to cause symptoms. Symptoms of cardiac tumors are generally secondary to local invasion, mass effect or embolization.

Case Report: A case of pericardial paraganglioma leading to right ventricular outflow obstruction and subsequent hypotension and syncope is presented. Due to its large size, slow growth and proximity to large vessels, the tumor considered to be at a very high risk for resection and conservative management was chosen.

Conclusion: Though rare, cardiac tumors must be among the differential for cardiovascular symptoms. Size and location of the tumor may determine the characteristics of symptoms produced, ranging from syncope, angina or dyspnea to cardiovascular collapse.

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CASE REPORT

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Keywords: Cardiac MRI, Cardiac tumor, Compression, Hypotension, Paraganglioma, Pericardial, Syncope

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INTRODUCTION

Primary cardiac tumors may be benign or malignant but are extremely rare with post-mortem studies reporting rates under 0.3% [1–3]. Secondary spread of the tumors to the heart i.e., cardiac metastasis is far more common with reported incidence as high as 18.3% [4, 5]. Most of these cases are asymptomatic and discovered either at autopsy or incidentally while using a myriad of imaging techniques including but not limited to echocardiography, magnetic resonance imaging (MRI) and computed tomography (CT) scans [6, 7]. Considering that the symptoms from tumors usually overlap those with cardiac conditions such as myocardial infarction, heart failure, primary arrhythmias among others, early diagnosis and effective treatment of cardiac tumors remains a clinical challenge [8].

We present a case of cardiac paraganglioma presenting as syncope in a patient from extrinsic compression of the right ventricular outflow tract (RVOT) causing obstruction.

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CASE REPORT

A 54-year-old male with past medical history of hypertension, cryptogenic liver cirrhosis, a benign pericardial mass and prior pheochromocytoma presented to the hospital with weakness and diarrhea. The mass was initially incidentally discovered at another hospital in 2003, when imaging showed the presence of a pericardial and abdominal periaortic mass. High suspicion of a neuroendocrine etiology for the tumors was confirmed by a positive metaiodobenzylguanidine (MIBG) scan and elevated catecholamine levels. The masses were deemed inoperable at the time of diagnosis and a presumed diagnosis of pericardial paraganglioma was made in light of available evidence. The patient failed to manifest any significant symptoms attributable to excessive hormone secretion and underlying hypertension was treated using alpha- and beta-blockers.

At home prior to presentation at current visit, the patient had a single episode of syncope that lasted a few seconds. His blood pressure at presentation was noted to be 60/40 mmHg with significant bradycardia at a heart rate of 40 beats per minute. An electrocardiogram revealed a junctional rhythm. The patient was responsive to two doses of atropine but remained hypotensive requiring admission to the intensive care unit for aggressive fluid resuscitation in combination with pressor support.

Computed tomography scan of the chest demonstrated a large heterogeneously enhancing mass, abutting the main pulmonary artery and ascending aorta measuring up to 7x7.8 cm in the greatest transverse and antero-posterior dimension respectively (Figure 1). 2D echocardiogram showed an estimated left ventricular ejection fraction $\geq 75\%$ and a large vascularized anterior mediastinal mass abutting the aortic root, ascending aorta partially obstructing the right ventricular outflow tract (RVOT) and pulmonary artery (Figure 2). Doppler data was significant for the presence of increased velocities across the RVOT with a peak velocity of 35 mmHg and mean gradient of 17 mmHg. A cardiac MRI scan was then ordered to better assess the anatomy of the mass and its relation to the great vessels.

Cardiac MRI scan (Figure 3) showed a large (65x74 mm) highly vascular anterior mediastinal paraganglioma between the right pulmonary artery and aorta causing extrinsic compression of the RVOT without intracardiac involvement. The patient was considered a poor candidate for surgery due to proximity of the mass to the major vessels of the heart, parasitization of cardiac blood supply and the anatomical challenges it posed for partial or complete resection. With continued supportive treatment, the patient's symptoms improved and his junctional bradycardia resolved. Comparative imaging from 2003 was acquired, which showed that the mass had been stable in size over the years with no evidence of invasion in the absence of related symptoms. The underlying hypertension controlled with cautious reintroduction of anti-hypertensive medications to

Figure 1: (A, B) Computed tomography scan of the chest demonstrated a large heterogeneously enhancing mass (arrow), abutting the main pulmonary artery and ascending aorta measuring up to 7x7.8 cm in the greatest transverse and antero-posterior dimension.

avoid precipitation of hypotension. He was eventually discharged with close outpatient follow-up where he remained asymptomatic.

DISCUSSION

Our case describes a unique case where initial use of an MIBG scan, commonly used for detection of adrenergic tissue such as pheochromocytomas helped in the incidental discovery of a large cardiac mass due to inherent pathological properties of the underlying tumor. Catecholamine-secreting tumors arise from the

Figure 2: Two-dimensional echocardiogram showed an estimated left ventricular ejection fraction $\geq 75\%$ and a large vascularized anterior mediastinal mass abutting the aortic root, ascending aorta partially obstructing the right ventricular outflow tract (RVOT) and pulmonary artery.

Figure 3: Cardiac magnetic resonance imaging showed a large (65x74 mm) highly vascular anterior mediastinal paraganglioma between the right pulmonary artery and aorta causing extrinsic of the RVOT without intra-cardiac involvement.

neural crest cells and have an annual incidence of 1 to 2 per 100,000. Eighty percent arise within the adrenal glands and present as pheochromocytomas with the remaining 20% being extra-adrenal in origin. The majority of the mediastinal paragangliomas are found in the anterior or posterior aorto-pulmonary groove and are largely nonfunctioning [9]. A functional catecholamine-secreting tumor can cause hypertension, diaphoresis or palpitations. Cardiac paragangliomas are rare and constitute $<5\%$ of all primary cardiac tumors. They tend to remain asymptomatic until discovered incidentally or grow to a size large enough to cause symptoms [10]. These tumors also tend to be locally invasive and can affect the cardiac conduction system [11]. When it comes to cardiac tumors, cardiac output can be compromised from several mechanisms contributing to direct flow obstruction (intrinsic or extrinsic), interference with valvular function, myocardial infiltration diminishing contractility, advanced heart block and the development of hemodynamically significant pericardial effusions and/or arrhythmias [12]. Our patient suffered from a paraganglioma led RVOT obstruction, which became clinically relevant due to the presence of significant dehydration resulting in a reduced preload, a drop in cardiac output, hypotension and transient cerebral hypoperfusion with syncope.

Many different imaging modalities can be used when evaluating cardiac tumors. MRI scan is presently the modality of choice in evaluating cardiac tumors, however, its results should be combined with data from coronary angiography. Cardiac MRI scan can assess the extent of myocardial infiltration, pericardial involvement and/or extra cardiac extension. It also allows the differentiation of tumor from other non-tumor masses such as the fibro muscular elements of the posterior wall of the right atrium [13]. On MRI imaging, cardiac paragangliomas are typically iso or hypo intense to myocardium on T1-weighted imaging and hyperintense on T2-weighted imaging. They enhance with contrast given their hypervascularity [13]. Coronary angiography can be useful to help determine the hypervascularization of the tumor by defining its feeding particles. This information is valuable when considering surgical resection and in its preparation [14].

The best therapy for paragangliomas is complete surgical excision, and remains the mainstay of treatment [15]. Surgical resection carries significant risks such as intraoperative hemorrhage given the tumor's hypervascular blood supply. Resection usually requires cardiopulmonary bypass and full thickness tissue resection due to lack of tumor encapsulation [15]. The removal of tumor usually requires reconstruction of the resected sites involving the right atrium, inferior vena cava and RVOT [16]. A surgical approach was abandoned in our case due to its considerably high risk nature resulting from large size of the tumor and its anatomical proximity of the tumor with the large vessels. During our assessment a conservative approach was considered

appropriate, considering the indolent nature of the paraganglioma, transient nature of exacerbation and the improvement in patient condition with simple supportive measures and correction of the underlying hemodynamic abnormalities. Prior reports have documented situations when surgical resection is not feasible. They note that the prognosis is usually guarded in such cases since the course of disease is typically slowly progressive [11].

CONCLUSION

Cardiac paragangliomas account for <5% of all primary cardiac tumors. They tend to remain asymptomatic, however, they can grow to a size large enough to cause symptoms that are generally secondary to local invasion, mass effect or embolization. Different imaging modalities can be used, with magnetic resonance imaging scan the diagnostic modality of choice. The surgical excision remains the mainstay of treatment however, there are cases such as ours that a conservative approach is more appropriate.

Author Contributions

Kailyn Mann – Substantial contributions to conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published

Mahek Shah – Substantial contributions to conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published

Naumann Islam – Substantial contributions to conception and design, Acquisition of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published

Ronald Freudenberg – Substantial contributions to conception and design, Acquisition of data, Drafting the article, revising it critically for important intellectual content, Final approval of the version to be published

Matthew Martinez – Substantial contributions to conception and design, Acquisition of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published

Larry Jacobs – Substantial contributions to conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published

Guarantor

The corresponding author is the guarantor of submission.

Conflict of Interest

Authors declare no conflict of interest.

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