Right Atrial Paraganglioma: An Extremely Rare Primary Cardiac Neoplasm Mimicking Myxoma

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In this report, we present a case of 35-year-old lady who had presented with atypical chest pain and exertional breathlessness for past six months. Transthoracic and transesophageal echocardiograms showed a well-circumscribed, echo-dense mass in the right atrium, attached to the interatrial septum at the level of atrioventricular junction and in the vicinity of coronary sinus ostium. She underwent successful resection of the cardiac mass. Histopathology revealed paraganglioma, which was reconfirmed by immunohistochemistry study. This represents an extremely rare presentation as primary cardiac tumors are 20-times less common than metastatic tumors and paraganglioma is one of the rarest primary cardiac tumors, accounting for <1% of all cases.

KEY WORDS: Cardiac mass · Primary cardiac tumor · Neural crest tumor · Pheochromocytoma.
are primary cardiac neoplasms. Myxomas are the most common primary cardiac tumors whereas paragangliomas are amongst the rarest, accounting for <1% of all cases.\(^\text{12}\)

Paraganglioma is a tumor of neural crest origin and is similar to a pheochromocytoma. For this reason, paragangliomas are also known as 'extra-adrenal pheochromocytoma'. Mediastinal paragangliomas can be either intracardiac or extracardiac and arise from autonomic neural tissue in these organs. Intracardiac paragangliomas are primary cardiac neoplasms. Myxomas are the most common primary cardiac tumors whereas paragangliomas are amongst the rarest, accounting for <1% of all cases.\(^\text{12}\)

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cardiac paragangliomas have been found mostly in the left atrium and less commonly in the interatrial septum, left ventricle, anterior surface of the heart, right ventricular outflow tract, or very rarely, right atrium.\(^5\) To our knowledge, only few cases of right atrial paraganglioma have been described in the world medical literature so far.\(^{14,9}\)

Clinically, cardiac paragangliomas can present either with symptoms of adrenergic excess or with mass effect.\(^{4,7,9}\) Mass effect depends on the location and the size of the tumor in relation to cardiac structures. Thus, the patients can present with acute coronary syndrome (compression of coronary ostium), heart failure (valvular obstruction), syncope (valvular obstruction, compression of great vessels), etc.\(^{4,5}\) Rarely, these tumors may also remain completely asymptomatic.

Diagnosis of paragangliomas is most often established after surgical resection only as there are no specific echocardiographic features. However, in catecholamine secreting tumors, preoperative diagnosis is important because surgical resection of such tumors without prior pharmacological conditioning can lead to disastrous consequences during the immediate postoperative period. Measurement of plasma metanephrine levels may help in preoperative diagnosis of such tumors.

Surgical resection is the primary therapeutic approach for paragangliomas. Surgery can be quite challenging in these cases due to vascularity of these tumors and their proximity with vital structures. For obvious reasons, mediastinal and cardiac paragangliomas are among the most technically demanding to resect.\(^{10}\) After surgical resection, long-term follow-up is essential, as paragangliomas can recur many years after initial surgery.\(^{10}\)

Our patient did not have any significant mass effect due to the tumor. Further, she probably had a non-secreting tumor because she was not complaining of any symptoms suggestive of catecholamine excess such as hot flush, sweating, headache or intermittent palpitation. Unfortunately, as we did not suspect paraganglioma in this case, we did not check for urine and serum metanephrine levels. However, in hindsight, we realized that measurement of urine and serum catecholamines should be performed in all primary cardiac tumors, particularly when tumor presents with atypical features and the diagnosis is not readily apparent.

**SUPPLEMENTARY MOVIE LEGENDS**

Movie 1. Transthoracic echocardiogram showing a large, echo-dense, sessile mass in the right atrium, attached to the interatrial septum at the level of atrioventricular junction.

Movie 2. Transesophageal echocardiogram showing a large, echo-dense, sessile mass seen attached to the interatrial septum near coronary sinus ostium. A small central area of echolucency is also seen.

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