

Rare Presentation of Pulmonary Hypertension in a Young Patient

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Received: February 24, 2020

Accepted: April 15, 2020

Published: April 16, 2020

Citation: Jain S, Sengodan P, Donthi D, Majeed M, Williams M, et al. 2020. Rare Presentation of Pulmonary Hypertension in a Young Patient. *J Med Imaging Case Rep* 4(1): 13-15.

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Abstract

Dyspnea on exertion has a broad differential diagnosis which includes pulmonary hypertension (PH). Atrial myxomas are the most common cardiac tumor which presents with pulmonary hypertension. Patient who present with pulmonary hypertension secondary to cardiac lesions such as myxomas might manifest with heart failure symptoms but the etiology of PH as an atrial myxoma is rare. We describe a unique presentation of a 33 year old Hispanic male from Nicaragua with history of hypertension found to have massive atrial myxoma with leading to pulmonary hypertension which was found incidentally on transthoracic echocardiogram. His symptoms resolved after surgical resection of the mass.

Keywords

Atrial myxoma, Pulmonary hypertension, Heart failure, Dyspnea

Introduction

Dyspnea on exertion has a broad differential diagnosis from disorders of the respiratory or cardiovascular system. It can also be multifactorial due to systemic illnesses, renal diseases, endocrinopathies, or malignancy. Cardiac masses are also considered a cause of dyspnea. The most common causes of cardiac masses include myxomas. Severe pulmonary hypertension (PH) can present with shortness of breath and lead to right ventricular failure. The World Health Organization classifies PH into five groups with over a 65% of cases due to left heart disease.

Atrial myxomas are the most common cardiac tumor that are found mostly in the left atrium. Most atrial myxomas tend to be asymptomatic however, given the potential for left ventricular inflow obstruction there has been a case of an atrial myxomas that presented with heart failure [1]. However, in that case the patient had decompensated heart failure with volume overload and pulmonary congestion. Our case is an unusual presentation of pulmonary hypertension due to the presence of a massive atrial myxoma that led to a mitral valve obstruction without evidence of decompensated heart failure.

Case Presentation

A 33 year old Hispanic male with history of hypertension, originally from Nicaragua was evaluated for progressive dyspnea on exertion associated with fatigue after walking four blocks. The patient was asymptomatic and very active

until a year ago. He denied syncope, chest pain, dizziness, palpitations, or edema. He had no previous surgeries, cancers, thyroid or liver diseases, stroke or transient ischemic events. He denied usage of tobacco, alcohol, or drugs.

On initial presentation in the office, he was normotensive with a heart rate of 80 beats per min, blood pressure of 103/70 mm Hg, saturating 100% on room air. Physical exam was significant for clear lung fields, normal first and second heart sounds, without significant murmurs, rubs or gallops, absent jugular venous distention, or peripheral edema. Chest radiography did not demonstrate any pleural effusion or pulmonary congestion. A transthoracic echocardiogram (TTE) found a giant mobile left atrial mass measuring 7 cm x 5 cm x 4.8 cm, filling the entire left atrium which prolapsed through the mitral valve into the left ventricle causing a significant mitral valve obstruction (Figure 1) with moderate eccentric mitral regurgitation. Maximum pressure gradient (PG) across the mitral valve was 19 mmHg and mean PG of 10 mmHg causing functional mitral stenosis at a heart rate of about 80 bpm (Figure 2). The pulmonary artery systolic pressure (PASP) was elevated at 54 mmHg with trace tricuspid regurgitation. Right ventricle was not well visualized. Left ventricular ejection fraction was 55% - 60%.

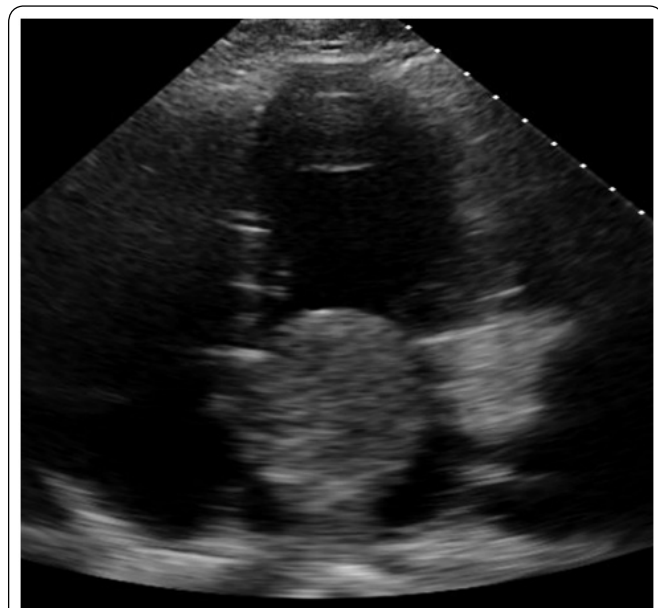


Figure 1: Transthoracic echocardiogram showing giant mobile left atrial mass (white arrow) measuring 7 cm x 5. cm x 4.8 cm prolapsing from the left atrium into the left ventricle.

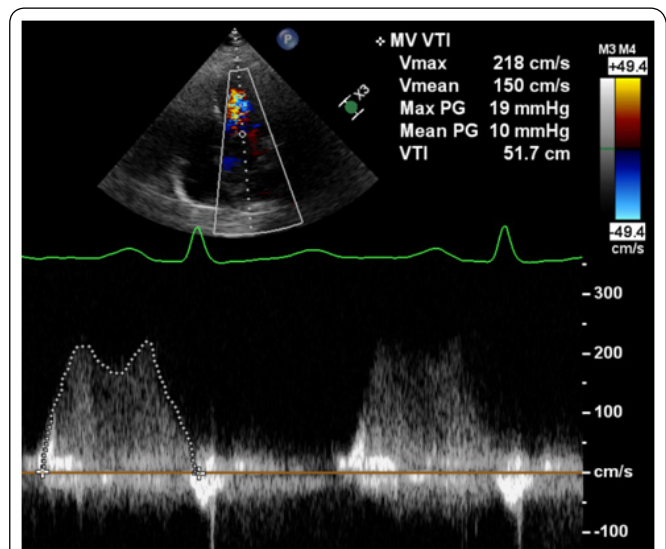


Figure 2: Transthoracic echocardiogram showing maximum pressure gradient (PG) across the mitral valve was between 19 to 22 mmHg and mean PG of 10 mmHg at heart rate of 80 to 86 bpm.



Figure 3: Gross pathological specimen: Red-tan, soft tissue (84.0 grams, 6.6*5.1*4.8 cm). The outer surface is inked blue and the apparent resection margin is inked black. Sectioning reveals a red-yellow, gelatinous and markedly hemorrhagic cut surfaces.

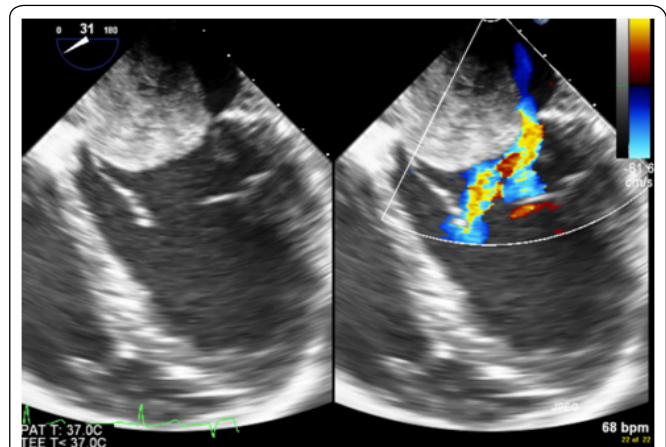


Figure 4: Intra-operative echocardiogram showed eccentric mitral regurgitation (white arrow).

The patient underwent surgery removal of the mass which spanned the entire fossa ovalis along the intra-atrial septum. Standard surgical approach is through the right atrium and the left atrium is opened through the fossa ovalis for tumor removal. In our patient, the tumor was large enough that the incision through the intra atrial septum had to be enlarged over the dome of the left atrium to facilitate removal (Figure 3). Intra-operative echocardiogram showed the eccentric mitral regurgitation jet (Figure 4). Post operatively the patient required atrial pacing due to junctional escape rhythm that was transient and resolved in few hours. Histopathological examination of the mass confirmed the diagnosis of myxoma (Figure 5). At two

month follow-up post operatively, the patient was able to walk up about 7 miles without any symptoms

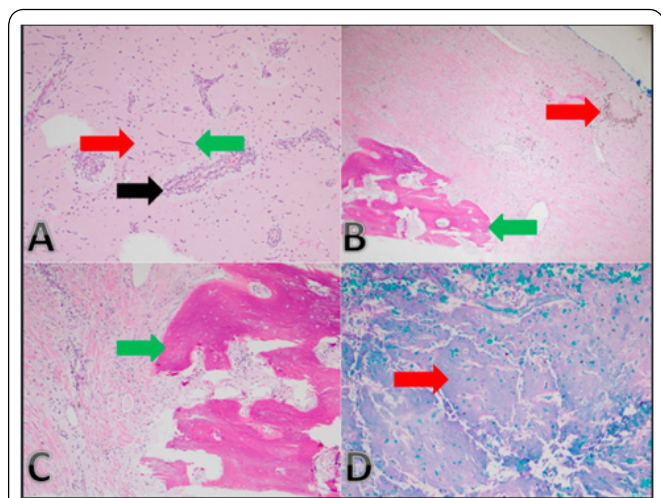


Figure 5: A. (H&E 10X) Numerous myxoma cells (green arrow) characterized by bland, elongated, and fusiform cells with oval nuclei and eosinophilic cytoplasm. The cells rest on a myxoid background (red arrow) and some exhibit perivascular arrangement of cells (black arrow). B. (H&E X4) Osseous metaplasia (green arrow) and numerous hemosiderin-laden macrophages (red arrow), indicating prior intratumoral hemorrhage. C. (H&E X10) Osseous metaplasia (green arrow). D. (H&E X10) Periodic Acid Schiff stain. Myxoid material of the tumor (red arrow).

Discussion

Our case displayed an interesting presentation of atrial myxoma causing significant left ventricular inflow obstruction and pulmonary hypertension. Atrial myxomas are the most common primary cardiac tumors found, presenting as 70–80% of all primary cardiac tumors [2]. These tumors typically present in women and are more commonly found in patient's sixth decade of life [3]. Atrial myxoma clinical presentations vary based on the size, location, and mobility of the tumor itself with the average dimensions of 3.7 cm - 5.6 cm [4]. Our patient is unique with his myxoma measuring 7 cm. Like this patient, most tumors are found in the intra-atrial septum in the fossa ovale region and less commonly in the right atrium and in the ventricles [5].

The most common presentation of patients with atrial myxoma are dyspnea on exertion, dizziness and syncope from mitral valve obstruction. Severely symptomatic patients can have pulmonary edema or heart failure. Dyspnea was the predominant symptom in our patient, however he did not have signs or symptoms of pulmonary edema. His echocardiogram had signs of significant pulmonary hypertension with his elevated PASP. Unlike the more common etiologies of PH such as systolic heart failure, chronic obstructive pulmonary disease, pulmonary emboli, or even mitral stenosis, cardiac myxomas are rare cause of pulmonary hypertension [1]. These patients can sometimes have positional dyspnea due to prolapse of the large myxoma over the mitral valve and "instantaneous mitral stenosis" with cough (pulmonary edema) and near syncope. This notion of the mitral stenosis like phenomena is in the setting of large, mobile obstruction of blood flow from atrium to the ventricle [6]. Stroke is sometimes the presenting

complaint as the myxoma could embolize and it is often found on the echocardiogram done as a part of the stroke work up. The pathophysiology of this phenomena includes the notion that the left atrial myxoma will cause increased pressure within the atrium which causes an overall increase in pressures within the pulmonary vasculature which leads to elevated pulmonary artery pressure as was seen in this patient [7].

Diagnosis of a myxoma is done by imaging where the diagnostic test of choice is usually a TTE, which is usually followed by a transesophageal echocardiogram (TEE). TTE considered the diagnostic of choice since they have a high sensitivity for evaluation of intracavitary and endocardial lesions. TEE are particularly helpful for further evaluation of lesions in atria, interatrial septum, superior vena cava, atrioventricular valves [8]. The echocardiogram was particularly important for diagnosis of this patient since no abnormal heart sounds were observed and the patient initially presented with vague onset of symptoms. Cardiac CT and MRI are becoming more common as it allows assessment of the cardiac tissue and function [9].

Treatment for these cases warrants surgical intervention and excision of these masses. Following removal of these masses, pulmonary hypertension should spontaneously resolve and symptoms should improve as observed in our patient.

Conflict of Interest

The authors declare no conflict of interest.

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