Case Report

Left Atrial Myxoma: A Primary Tumor of the Heart

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Abstract

Primary tumors of the heart are rare across all age groups, with a reported prevalence of 0.001 to 0.03% in autopsy series. Secondary involvement of the heart by extra-cardiac tumors is 20-40 times more common than by primary cardiac tumors (PCTs). The first pre-mortem diagnosis of a PCT, a cardiac myxoma, was made by Goldberg in 1952. Despite rarity, there are multiple recognized histologic types of PCTs. The overwhelming majority of PCT are mesenchymal tumors that display the full spectrum of differentiation as do those seen in the soft tissue. About 75 % of all PCT are regarded as benign neoplasms (BN), with cardiac myxoma accounting for at least half of them. Hence, here we report one such rare case of 45 years old admitted with vague complained of breathlessness and bilateral pleural effusion and was subsequently diagnosed to have left atrial myxoma. Sometimes, practicing doctors are unable to detect, and high index of suspicion is needed to arrive at the diagnosis i.e., the cause for breathlessness.

Keywords: Benign neoplasm, Left atrial myxoma, Primary cardiac tumors

INTRODUCTION

Primary tumors of the heart are rare across all age groups, most commonly in the third through sixth decades, with a female predilection, with a reported prevalence of 0.001-0.03% in autopsy series.[1] Approximately, 90% of myxomas are sporadic; the remainder are familial with autosomal dominant transmission.[2] About 75% of all primary cardiac tumors (PCTs)are regarded as benign neoplasms, with cardiac myxoma accounting for at least half of them.[1-3] Of the remaining 25% of PCTs that are considered to be malignant neoplasms, the majority are sarcomas, with lymphomas being the next most common.[4]

The diagnosis of PCTs is frequently challenging. The symptoms associated with most PCTs are nonspecific, and they often mimic far more commonly encountered disease entities.[5] Further, many tumors present with mild and vague symptoms such that most routine work-ups will fail to identify the underlying abnormality. This elusiveness often results in a delay in the diagnosis of disease. Fortunately, the more widespread use of noninvasive and relatively sensitive imaging modalities such as cardiac echocardiography, computed tomography (CT), should facilitate the identification of cardiac lesions.[6]

However, the consideration of PCT in the differential diagnosis combined with a high index of suspicion is also paramount in arriving at the correct diagnosis. In addition to the diagnostic challenges, the management of some PCTs is also not straightforward even when the histologic diagnosis has been made. Many benign PCTs are now found incidentally in asymptomatic individuals. Therefore, the treatment decision requires a thorough analysis of the potential benefits and harms of the surgery versus conservative management.[7] Due to our limited experience with the natural clinical course of many PCTs, this treatment decision may be difficult to reach.

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CASE REPORT

A 45-year-old patient without significant past medical or surgical history presented with 24 weeks of progressive breathlessness associated with decreased appetite and generalized weakness and fatigue.

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On examination, pulse was 64/min, regular and blood pressure was 130/70 mmHg with respiratory rate 38/min. There was pallor present, no icterus, clubbing, raised jugular venous pressure or pedal edema.

Cardiovascular system revealed, the presence of a holosystolic murmur most prominent at the apex with radiation to the axilla, the presence of a diastolic murmur, and the presence of a tumor "plop".

Examination of respiratory system revealed decreased breath sounds, VF and VR in bilateral inframamary axillary and infra-axillary region.

Abdominal and central nervous system examination was unremarkable.

Investigations revealed hemoglobin 9.8 mg/dl, total white cell count 11,000/cmm, with differentials, of P83L15E12, erythrocyte sedimentation rate 45 at 1 h, serum bilirubin 0.6 mg/dl, serum albumin 3.4, serum glutamate pyruvate transaminase 16 U/L, alkaline phosphatase 91 U/L. A random blood sugar level was 142 mg/dl. Immunoassay for HBsAg and ELISA HIV were negative. Serum urea creatinine and urinalysis were within normal limits.

Ultrasonography of the abdomen was normal, and chest PA view showed bilateral pleural effusion. ECG was within normal limits; pleural fluid analysis was transudate with ADA 21.

The patient was subjected to echocardiography that revealed left atrial myxoma (LAM)measuring 42 mm * 32 mm obstructing left atrial outflow. The patient was diagnosed as a case of LAM and ultimately referred to a higher center for surgical excision of the tumour (Figure 1).

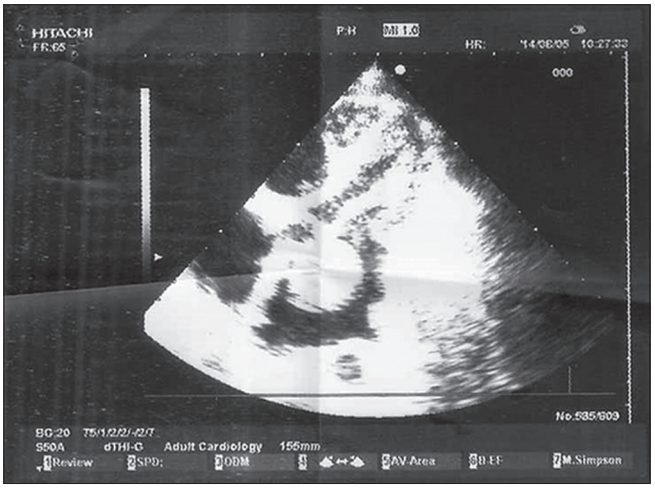

Figure 1: 2D echo 4 chambered view showing left atrial myxoma measuring (42 mm × 32 mm) with stalk attached to interatrial septum

DISCUSSION

Cardiac tumors may present with a wide array of cardiac and non-cardiac manifestations. These manifestations depend in large part on the location and size of the tumor and are often nonspecific features of more common forms of heart disease, such as chest pain, syncope, heart failure, murmurs, arrhythmias, conduction disturbances, and pericardial effusion with or without tamponade.[1,2] In addition, embolic phenomena and constitutional symptoms may occur.

Myxomas commonly present with obstructive signs and symptoms. The most common clinical presentation mimics that of mitral valve disease: either stenosis is owing to tumor prolapse into the mitral orifice or regurgitation resulting from tumor-induced valvular trauma. Ventricular myxomas may cause outflow obstruction similar to that caused by subaortic or subpulmonic stenosis.[4] The symptoms and signs of myxoma may be sudden in onset or positional in nature, owing to the effects of gravity on tumor position. A characteristic low-pitched sound, a "tumor plop," may be appreciated on auscultation during early or mid-diastole and is thought to result from the impact of the tumor against the mitral valve or ventricular wall.[4]

Myxomas also may present with peripheral or pulmonary emboli or with constitutional signs and symptoms, including fever, weight loss, cachexia, malaise, arthralgias, rash, digital clubbing, Raynaud's phenomenon, hypergammaglobulinemia, anemia, polycythemia, leukocytosis, elevated erythrocyte sedimentation rate, thrombocytopenia, and thrombocytosis.[5] These factors account for the frequent misdiagnosis of patients with myxomas as having endocarditis, collagen vascular disease, or a paraneoplastic syndrome.

Two-dimensional transthoracic or omniplanetransesophageal echocardiography is useful in the diagnosis of cardiac myxoma and allows assessment of tumor size and determination of the site of tumor attachment, both of which are important considerations in the planning of surgical excision.[6,7] CT and magnetic resonance imaging may provide important information regarding size, shape, composition, and surface characteristics of the. Obstruction to left atrial outflow was causing raised left atrial pressure and subsequently pulmonary congestion leading to breathlessness that was more on sitting and standing position due gravitional effect causing LA outflow obstruction. Decreased appetite may be explained by systemic effect of the tumor and fatigue due to underlying anemia.

CONCLUSION

Practicing doctors may be advised to look for LAM when patient present with features similar to that of MS and transudate pleural effusion, if not otherwise explained.

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