

Left Ventricular Myxoma—A Case Report

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Abstract

Left ventricular myxomas are extremely rare and account for 2.5% of all cardiac myxoma cases. A left ventricular myxoma originating from the apical interventricular septum and projecting into the left ventricular cavity was diagnosed by Transthoracic two-dimensional echocardiographic imaging in a 29-year-old male. The presentation of myxoma with symptoms masquerading as infectious hepatitis and dilated cardiomyopathy with a measured ejection fraction of 20% has been described.

Keywords

Myxoma, Interventricular Septum, Left Ventricle, Cardiomyopathy

1. Introduction

Primary intracardiac tumors are rare and a cardiac myxoma is by far the most common type of primary cardiac tumor. Left ventricular localization of a myxoma is accounting for 2.5% - 4% of all cases [1]. The first description of a primary intracardiac tumor was in 1559, located in the left ventricle [2]. In a review of literature, Mazer and Harrigan reported the first case of LV myxoma diagnosed by 2D echocardiography in 1982 [3]. In 1997, Meller *et al.* noted the low prevalence of LV myxoma. At that time only 15 cases have been reported in the English and French literature [4]. In 1992, 30 cases have been reported by Soma, *et al.* [5]. Only 37 cases of left ventricular myxomas have been reported up to 1996 [6] and 72 cases up to 2014 [7].

It is believed that embryonic residues after the in-utero septation of the heart are those that give rise to the myxoma [8]. Thus, cardiac myxomas may originate from anywhere within the cardiac chambers. Actually, majority of cardiac myxomas occur in the atria with only 3% - 10% identified in either the left or right ventricle. These tumors usually project from the endocardium into the cardiac chambers.

Cardiac myxomas have varying clinical presentation, uncertain histogenesis and debatable immunohistochemical profile [9]. These tumors exhibit a heterogeneous phenotype, with adult cells expressing protein antigens specific to various cell lineages, often within the same tumor, including epithelial, endothelial, myogenic, myo-

fibroblast, neural and neuro-endocrine antigens. Cardiac myxomas also show variable response to other antisera, including factor VIII-related antigen, ulex europaeus agglutinin, vimentin, desmin, myoglobin, S-100 and cyto-keratin. Given the variable response to such a broad range of antisera, combined with ultrastructural appearance that commonly suggests an endothelial derivation, but has also been recognized to suggest neural or neuro-endocrine origins. It is most likely that cardiac myxomas are derived from a pluripotent mesenchymal stem cell or sub-endothelial vasiform reserve cell located around the fossa ovalis and surrounding endocardium. These cells persist as embryonic residues during septation of the heart; thus the prevalence of myxoma in the atrial septum is understandable [10]. In cardiac myxoma cells, the phenotypic marker of the embryonic endothelial-to-mesenchymal transformation that precedes terminal differentiation of endocardial cushion, supporting the hypothesis that cardiac myxoma cells may derive from adult developmental remnants [11]. This cell type appears to most commonly following an endothelial lineage, but is capable of differentiation into other cellular phenotypes. The phenotypic expression is variable in these tumors and does not necessarily reflect the tumor origin. It is important to note the difference between cardiac myxomas and Prichard structures, which appear to be minute, age-related, endothelial deformities, and not benign neoplastic growths [12].

Dilated cardiomyopathy occurs when disease affected muscle fibers are damaged or stretched (dilated) in one or more chambers of the heart. As the heart enlarges, it decreases its efficacy in pumping blood and progresses to congestive heart failure. Dilated cardiomyopathy is usually an acquired condition in adults and can be caused by acute myocarditis, an inflammation of heart muscle resulting from infectious, immunological, toxic and nutritional mediators.

A left ventricular myxoma originating from the apical interventricular septum and presented with features of dilated cardiomyopathy is uncommon and so this case had been reported.

2. Case Report

29-year-old, a previously asymptomatic male was admitted in the hospital with a history of sudden onset of non specific constitutional symptoms such as fever, non productive cough and vomiting episodes suggestive of a systemic illness for one week duration. He developed extreme tiredness, lethargy, giddiness and dyspnea on lying down especially during sleep hours. His pulse rate was 110 bpm and blood pressure was 100/70 mmHg. Physical examination revealed enlarged apical impulse on left side of the chest, muffled heart sounds, S₃ gallop and no murmurs. Blood chemistry analysis showed a total leukocyte count of 13,200 cells/mm³ of blood and polymorphs (65%), lymphocytes (33%) and eosiophils (2%). Erythrocyte sedimentation rate was 3 - 9 mm/hour. Blood urea (36 mg/dl), sugar (96 mg/dl) and creatinine (1.4 mg/dl). Haemoglobin level was 13.5 gm%. He was not alcoholic and the total serum bilirubin was elevated to 3.7 mg/dl, suggestive of hepatic infection. ECG was normal. Ultrasound abdomen revealed no abnormality. He was treated with cefotaxime, ranitidine, domperidone, lactulose and multivitamins. His symptoms were worsening. X-ray chest revealed massive cardiomegaly and he was referred to echocardiographic evaluation.

Transthoracic 2D echocardiography revealed a solitary, pedunculated mass with a size of 43.8 mm in length and 16 mm in diameter, attached by a pedicle to the endocardium of apical interventricular septum and projecting into the left ventricular cavity with a typical appearance of “cluster of grapes” suggestive of a myxoma and the features of dilated cardiomyopathy with severe LV (Left Ventricular) dysfunction as shown in the **Figures 1-4**.

The patient developed severe LV dysfunction with features of dilated cardiomyopathy.. He was supported with oxygen, inotropes and diuretics. His condition worsened further and died in the hospital within 10 days of the course of illness. Since his general condition was very poor, surgical removal the tumor was not attempted.

3. Discussion

a) Frequency

Primary cardiac tumors are rare and have an average incidence of 0.02 % [13] [14]. Myxoma is the most prevalent heart tumor and is the most common benign tumor of the heart. A small number (6%) of myxomas are equally split between the left and right ventricles (3%) each [15]. Only 3% - 4% of myxomas are detected in the left ventricle [16].

b) General features

Myxomas occur in all age groups but are particularly frequent between the third and sixth decades of life [17].

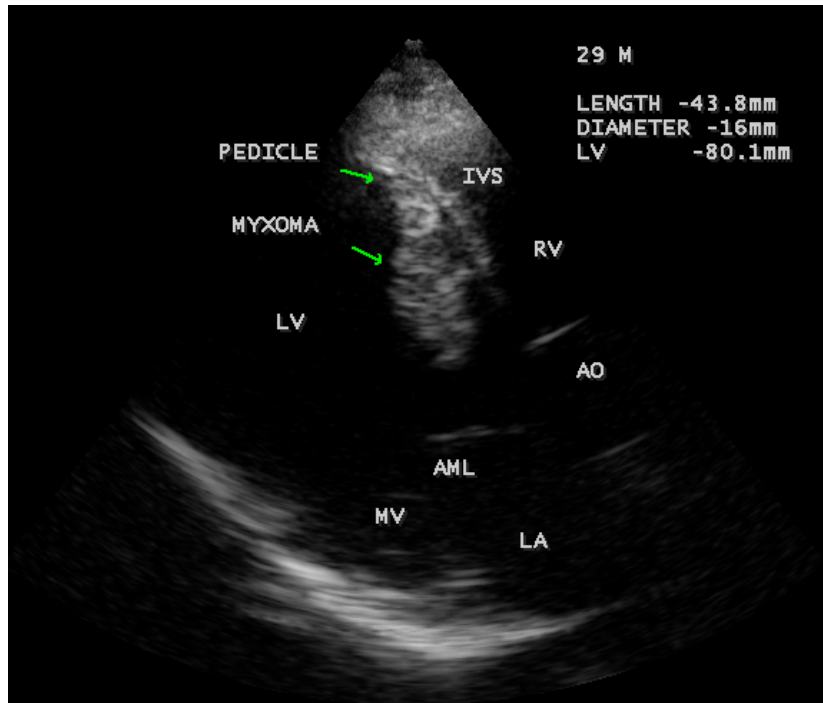


Figure 1. (Parasternal long axis view—showing the myxoma originating from the apical interventricular septum with an attachment by a pedicle to the endocardium and projecting into the left ventricular cavity as “cluster of grapes”).

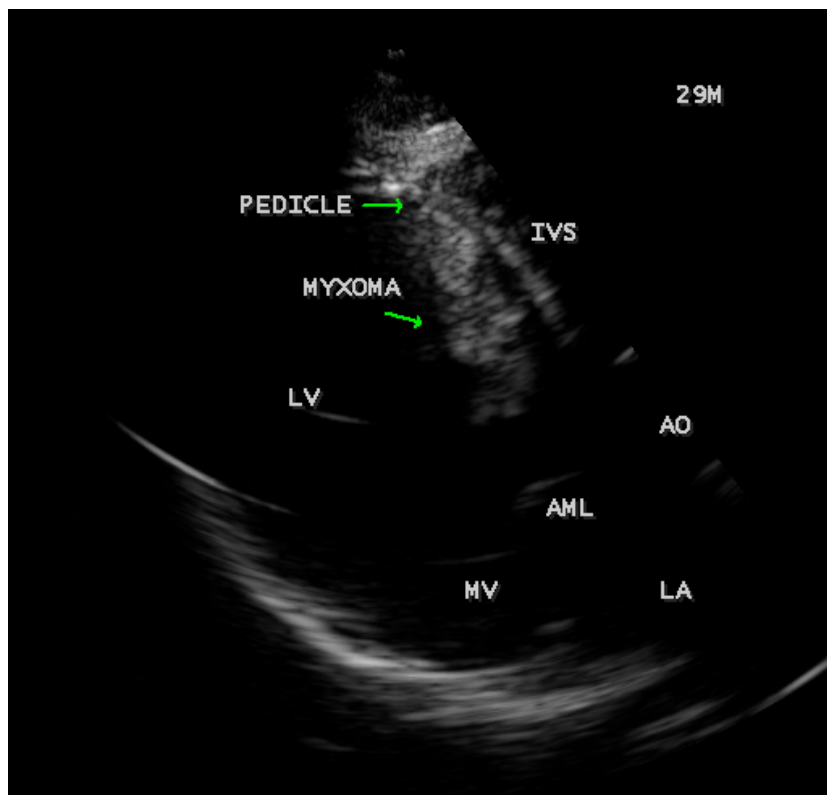


Figure 2. (Parasternal long axis—Tilted view—showing the myxoma progressing towards LV outflow tract).

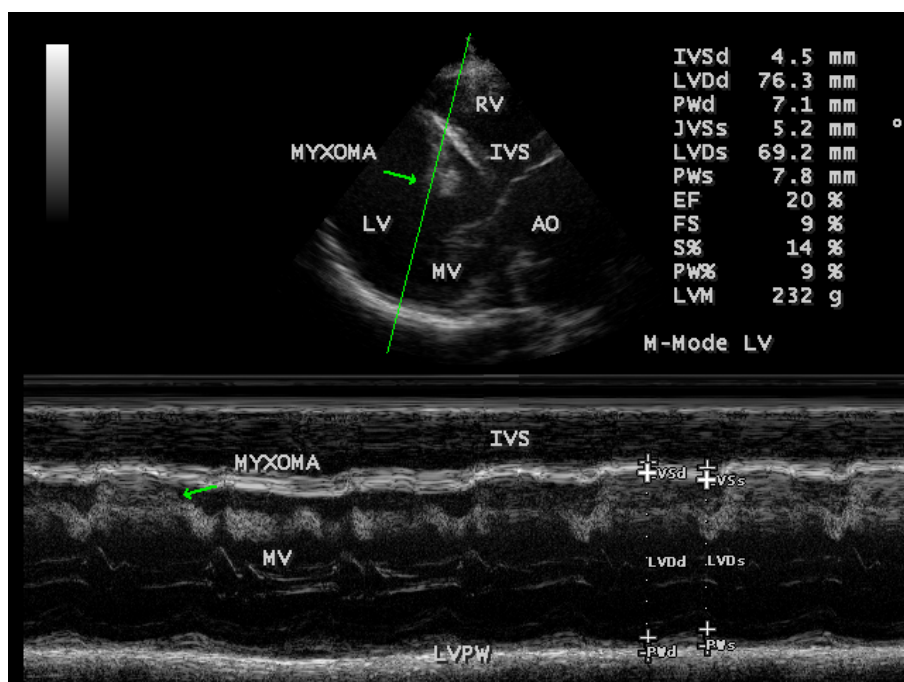


Figure 3. (M-Mode LV function study—showing Global hypokinesia, Severe LV dysfunction with Ejection Fraction (EF)-20% suggesting “Dilated cardiomyopathy”).

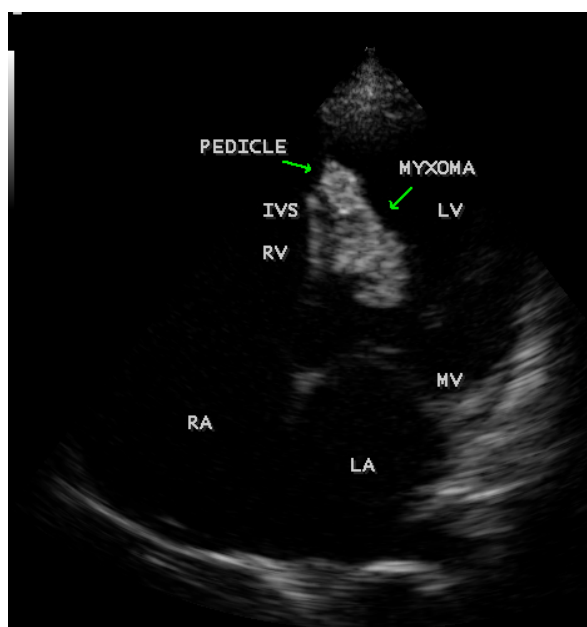


Figure 4. (Apical 4 chamber view—showing the myxoma projecting into the LV cavity with an attachment by a pedicle to the apical interventricular septum and dilated cardiac chambers suggesting “Dilated cardiomyopathy”).

In a series of 66 sporadic myxomas, the female-to-male ratio was 2.7:1 [18] and 3:1 in one series [19]. However, female sex predominance is less pronounced in familial myxomas. The precise rate of growth of cardiac myxoma is unknown, although it is believed to be reasonably fast, an average rate of 0.15 cm per month [20]. Tumors can range in size from 1 - 15 cm in diameter, although most measure approximately 5 - 6 cm across. Myxomas are, indeed, neoplastic and the impetus for this neoplastic transformation is unclear. Although genetic

factors play a role in myxoma syndromes, they do not appear to offer a consistent explanation in sporadic cases. A study by Li *et al.* reported finding evidence for HSV-1 (Herpes simplex virus) infection in 70% of a relatively small cohort ($n = 17$) of surgically resected sporadic cardiac myxomas [21]. Although the majority of cardiac myxomas are histologically benign, due to their strategic location (left or right cardiac chambers) and nature (size, mobility and overall morphology), they may lead to serious consequences for morbidity and mortality of affected patients. The mobility of the tumor within the heart varies according to the amount of collagen they contain, the degree of attachment to the ventricular wall and the length of the stalk attaching them to the heart.

c) Clinical Presentation

The clinical presentation of patients with myxoma can be quite different. Because of nonspecific symptoms, early diagnosis of the myxoma may be a challenge. Cardiac myxomas have no typical presentation. Typically, patients are asymptomatic or present with nonspecific signs and symptoms. Some authors call heart neoplasms the great “masqueraders” [22]. Symptoms of obstruction, embolic and systemic manifestations are components of the classical triad, but rarely are present all. However, at least one of the triad symptoms is present. Systemic reaction due to the products of the degeneration of the tumor leading to fever, weight loss, leucocytosis which may occur especially in the early stage of the illness. Vague constitutional symptoms are due to an inflammatory response that can be associated to many diseases. This patient presented with vomiting episodes, a non-productive cough and mild fever. These symptoms are rather non specific and cardiac myxomas are often misdiagnosed [23]. In rare cases, myxoma can also serve as a nidus for infection [24]. When this occurs, symptoms parallel those of infective endocarditis.

Heart failure is a rare complication of myxoma. Left ventricular myxoma have been reported to lead to silent heart failure, as evidenced by the echocardiographic visualization of a 3.3×1.2 cm size, a gelatinous friable myxoma, hanging as a mass with a stalk (pedicle) on interventricular septum near the anterior mitral valve annulus in a 54 years old male having pneumonitis and severe LV dysfunction with a measured ejection fraction of 22% [25]. In this case, a 4.38×1.6 cm size myxoma, hanging as a mass with a stalk on the apical interventricular septum as visualized by the echocardiography. Initially the patient had non-specific constitutional symptoms masquerading as infectious hepatitis and subsequently developed heart failure with severe LV dysfunction with a measured ejection fraction of 20%. The inflammatory mediators released from the tumor following a systemic illness, producing an immunological damage to the heart muscle fibers. The resultant myocarditis, induced the muscle fibers to stretch and when the disease became progressive, all four chambers of the heart were affected and enlarged with features of dilated cardiomyopathy. Summary of clinical presentation of this case was given in **Table 1**.

d) Echocardiography

Two-dimensional echocardiography is the diagnostic procedure of choice in revealing myxomas [26]. A big (2.5×5 cm) grape-like mass in the left ventricle suggestive of myxoma, attached to the inferior septum and bulging into the left ventricular outflow tract had been described by a routine Transthoracic echocardiography in a 35 year old Asian male [27]. A large left ventricular myxoma, a pedunculated pear-shaped mass (75×45 mm in diameter), attached by a pedicle to the apical interventricular septum and prolapsing through the LV outflow tract and aortic valve, causing a severe obstruction was found by echocardiography in a 60 year old woman [28]. In this patient, 2D echocardiographic **Figure 1** and **Figure 4** illustrate the myxoma attached by a pedicle to the endocardium of apical interventricular septum and projecting into the left ventricular cavity with the appearance of a “cluster of grapes” [29] in parasternal long axis and apical four chamber views. **Figure 2** illustrates the myxoma progressing towards LV outflow tract. **Figure 3** illustrates the M-Mode LV function study suggestive of dilated cardiomyopathy with severe LV dysfunction (EF 20%).

e) Sudden death

Cardiac myxomas elicit a wide variety of symptoms in patients, largely depending on the size and location of the tumor. Individuals with cardiac myxomas can present at any point along the clinical continuum, ranging

Table 1. Summary of clinical presentation.

Course of illness	Symptoms	Duration	Presentation
Early stage	Non-productive cough, mild fever, vomiting episodes	One week	Infectious hepatitis
Late stage	Tiredness, lethargy, giddiness, dyspnea on lying down	Ten days	Heart failure

from complete absence of symptoms (particularly with tumor less than 40 mm in size) to sudden death, usually owing to acute obstruction or embolization [30]. In this case, a 43.8 mm size myxoma produced a systemic illness and sudden death due to severe LV dysfunction in a middle aged male is a rare occurrence.

4. Conclusion

An isolated left ventricular myxoma originating from the apical interventricular septum, masquerading as infectious hepatitis was diagnosed by Transthoracic 2D echocardiography. It presented with features of dilated cardiomyopathy in a 29-year-old male in Tropical Nation such as India at Thoothukudi region in Tamil Nadu State and it is a rare incidence noticed in the year of 2015 at this hospital.

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