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Left Ventricular Fibroma Mimicking an Acute Coronary Syndrome

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ABSTRACT

Cardiac fibromas are exceedingly rare neoplasms. We report on a 21 year old woman who presented with symptoms that were initially misinterpreted as an acute coronary syndrome. Radical surgical resection was undertaken and was considered curative, as the mass histology was consistent with a benign fibroma.

INTRODUCTION

Cardiac fibromas are predominantly diagnosed in children in whom they follow rhabdomyomas in incidence as the second most common tumor in that age group. Even though they appear well delineated, they are not encapsulated. They typically occur as solitary lesions and do not have a propensity for spontaneous regression, as is the case with rhabdomyomas.

CASE REPORT

We report on a 21 year old woman with recent onset chest pain and no previous cardiac history. Her EKG revealed non-specific ST abnormalities in inferior leads. Multiple episodes of sustained monomorphic ventricular tachycardia were noted. A sestamibi cardiac perfusion scan showed an irreversible defect in the inferior portion of the left ventricle (LV). An echocardiogram showed preserved LV function, absence of valvular abnormalities and a 4x3 cm mass the origin of which could not be clearly identified (Fig. 1). Since it was impossible to distinguish echocardiographically whether the mass was a primary cardiac neoplasm or originated from the pericardium and compressed the LV, the diagnostic algorithm was expanded to include multi-slice computed tomographic imaging (MSCT). The MSCT located the tumor within the left ventricular wall (Fig. 2). No evidence of neoplastic dissemination was evident. Following a left heart catheterization which revealed no pathology of the coronary arteries and no sign of pathologic vascularization, the patient was referred for surgical management. The operation was performed with the aid of cardiopulmonary bypass. The intraoperative findings were consistent with a well delineated tumor encased within the inferior surface of the LV wall (Fig. 3). Even though no clear encapsulation was seen, a dissection plane was developed which made a radical excision of the tumor possible. During the course of the dissection the LV chamber was opened. The proximity of the mitral valve apparatus warranted close inspection of the valve. This was achieved through a separate left atriotomy which facilitated testing the mitral valve for competence with a bulb syringe. With the integrity of the mitral valve preserved, the patient was weaned off cardiopulmonary bypass

without difficulty. Her postoperative recovery had been compromised by a streptococcal line infection which responded favorably to penicillin. Pathohistologic scrutiny revealed elongated fibroblasts interspersed within collagen and elastin fibers, which was consistent with a fibroma.

COMMENT

Cardiac fibromas are primarily diagnosed in children and adolescents. The wide spectrum of clinical manifestations includes chest pain, dyspnea, conduction abnormalities, intracavitary obstructions, congestive heart failure and sudden death secondary to malignant ventricular arrhythmias (1,2). Some tumors, however, remain clinically silent. Cardiac fibromas are solitary lesions, and though circumscribed they lack true encapsulation. They tend to infiltrate the surrounding myocardium and may include calcific deposits (2). Their benign histology correlates well with their clearly defined borders and absence of metastatic potential. These tumors originate from fibroblasts and myofibroblasts, sharing the gross and microscopic appearance of their soft tissue counterparts (2,3). No propensity for spontaneous regression has been demonstrated (2). The diagnostic algorithm begins with echocardiography but may be expanded to include magnetic resonance imaging or computed tomography. Both of the latter imaging methods allow for explicit insights into the regional topography and degree of tumor invasiveness. Cardiac fibromas have a predilection for the left ventricle, which may be reflective of its fibroblast rich interstitium (4). The presentation in the neonate differs in that the tumor more commonly involves the interventricular septum (3). The microscopic appearance of cardiac fibromas is one of uniform fibroblastic cells interspersed within a collagenous matrix (3).

The preferred therapeutic option is radical surgical excision. Optimal long term results have, however, been attained with subtotal resections in certain patients (4). A more radical approach is favored by some authors, and heart transplantation has been

advocated for patients with unresectable cardiac fibromas (5). The patient report on here, presented with symptoms mimicking an acute coronary syndrome. Further diagnostic work up revealed that her symptoms as well as the perfusion defect seen on a sestamibi scan, were related to a left ventricular neoplasm rather than coronary malperfusion. Radical excision of the tumor was feasible and was followed by restoration of the structural integrity of the left ventricle. The proximity of the tumor to the mitral valve apparatus presented a surgical challenge, but its preservation proved to be feasible. The operation was supplemented with intraoperative transesophageal echocardiography which demonstrated a competent mitral valve without evidence of a residual tumor. She continues to do well at her six week follow up examination.

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Fig. 1. Long axis parasternal echocardiographic view showing the tumor compressing the left ventricle and its close proximity to the mitral valve apparatus

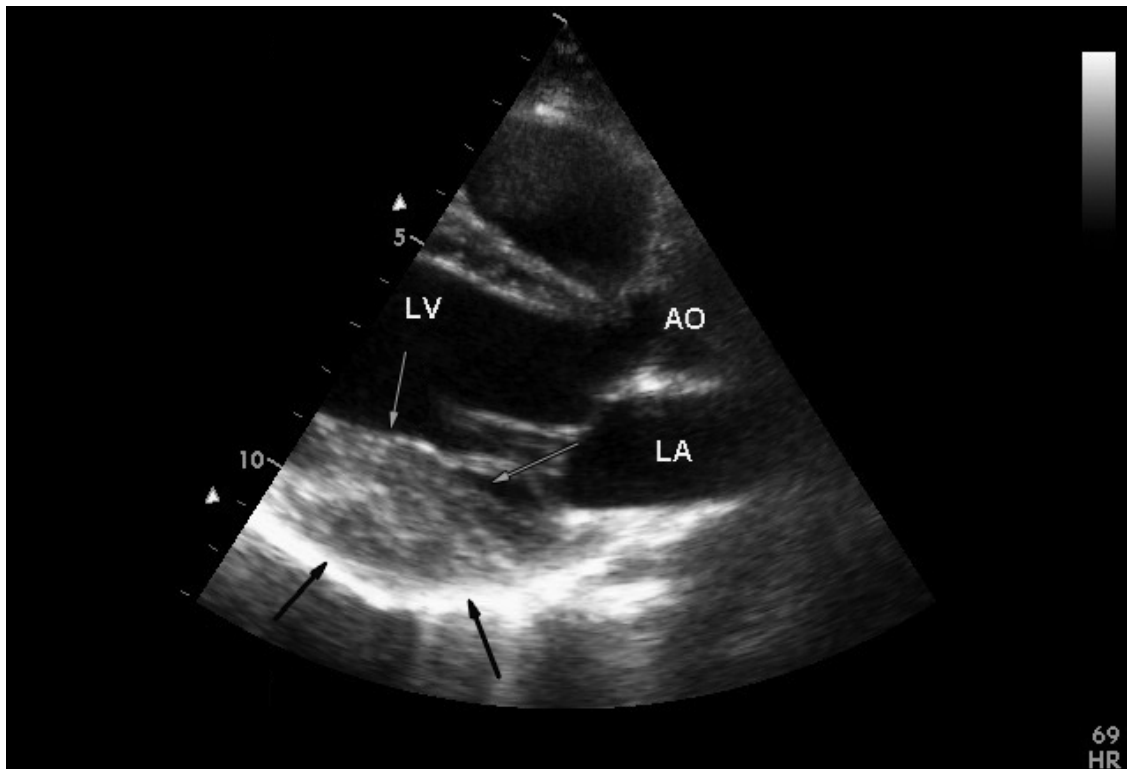


Fig. 2. Sagittal computed tomographic image of the tumor (SVC= superior vena cava, Ao=aorta, LV=left ventricle, Tm=tumor)

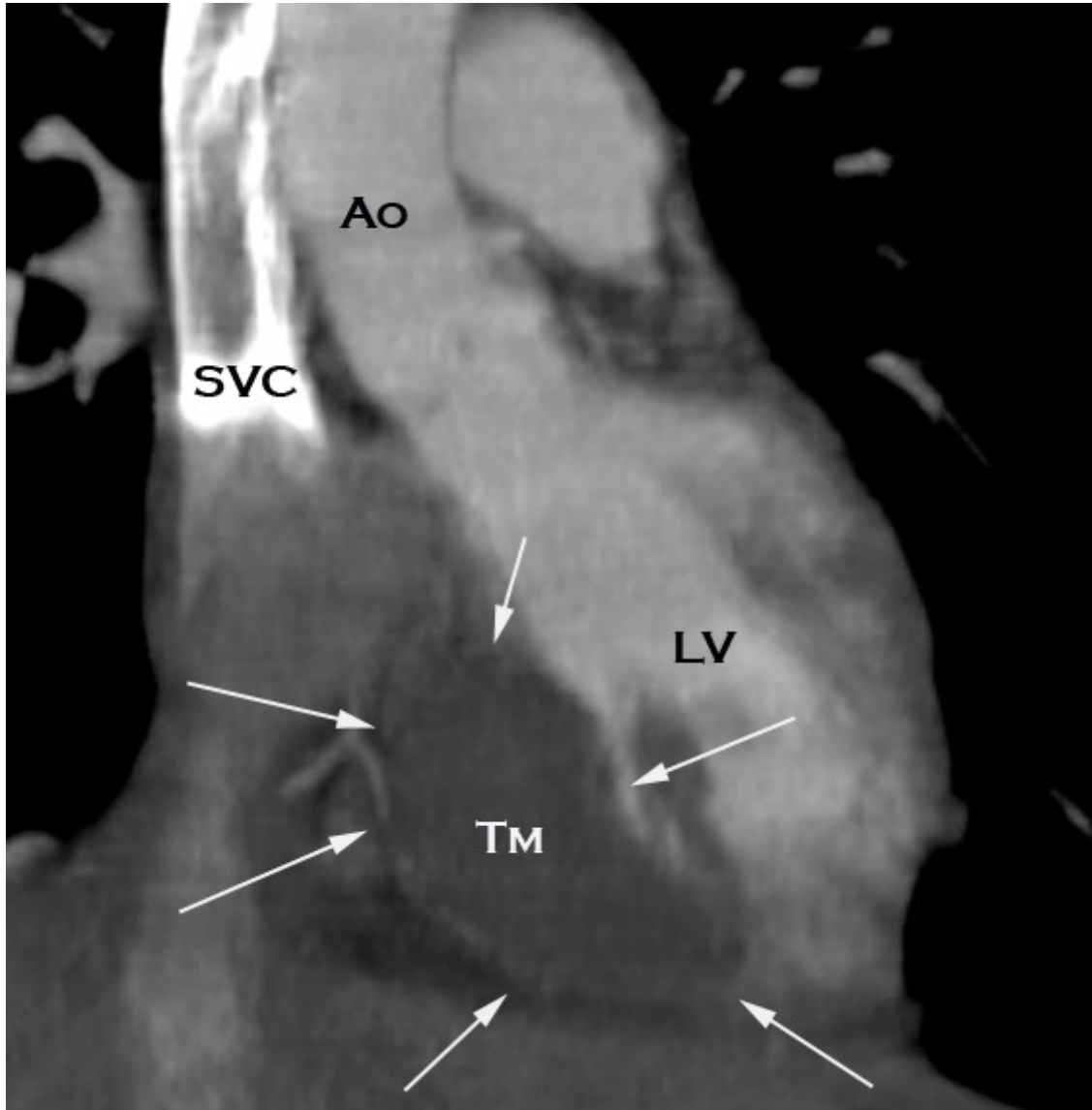


Fig. 3. Intraoperative image of the left ventricular tumor (arrows)

