

Giant Cardiac Fibroma in a Completely Asymptomatic Teenager

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Abstract

We report the case of a 19 years old "healthy" and asymptomatic patient accessing the Emergency Room after a car accident trauma. A routine electrocardiogram documented an intraventricular conduction disturbance. The subsequent transthoracic echocardiogram showed the presence of a voluminous heterogeneous intracardiac mass (10 × 10 × 8 cm), localized in the medium-apical cavity of the right ventricle and extended to the outflow tract. A mass debulking intervention was performed and the intraoperative biopsy samples allowed the diagnosis of cardiac fibroma. Because of dimension, intracardiac infiltration and relations, radical surgery was not an option; the patient was candidate for heart transplantation. This unique case highlights the questionable classification of cardiac fibromas as benign and the possibility of a delayed diagnosis because of late clinical presentation.

Keywords: Cardiac fibroma; Benign tumors; Cardiac surgery

Introduction

Primary cardiac tumors are extremely rare, with a reported incidence ranging from 0.0017 to 0.019% [1], most of them are benign and fibroma is the second for prevalence after rhabdomyoma. The most frequent sites of occurrence for cardiac fibroma are the left and right ventricles and the interventricular septum. Clinical presentation is often subtle and is influenced by size and location of the mass, which could determine heart failure, malignant arrhythmias and sudden cardiac death. Echocardiography is essential for the diagnosis, nevertheless computed tomography (CT) and magnetic resonance imaging (MRI) better define size, shape, and mostly the anatomical relationships of the lesion. The therapeutic approach is the "in toto" excision of the mass, when possible according to size and anatomical relationship; conversely as a last choice cardiac transplantation remains the only solution [2].

Case Report

We present the case of a 19 years old "healthy" and asymptomatic patient accessing the Emergency Room after a car accident trauma. In absence of any cardiologic objective sign or symptom, a routine electrocardiogram (EKG) documented an intraventricular conduction disturbance: complete right branch block (RBB) with QRS duration of 160 msec and extreme left axis deviation (Figure 1a). The patient thus underwent a transthoracic echocardiogram with the evidence of a voluminous heterogeneous intracardiac mass (10 × 10 × 8 cm) localized in the medium-apical cavity of the right ventricle (RV) engaging the outflow tract and generating a mean gradient of 20 mmHg (max gradient 41 mmHg). The subsequent trans-esophageal

echocardiogram (TEE) confirmed the same findings in terms of size and obstruction grade (Figure 1b). Presence of metastases was excluded by total body CT. Cardiac CT nevertheless confirmed the anatomical features of the mass and demonstrated an extensive necrotic and heterogeneous component. A mass debulking intervention was performed and several intraoperative biopsy samples for histological diagnosis were obtained. During cardiac surgery a large and hard-elastic mass, originating from the interventricular septum and extending to the left pulmonary artery and left atrium roof, was evident. A transverse incision proximal to the pulmonary valve plane was performed. The oval mass causing obstruction of the RV outflow tract was excised, but the massive involvement of the interventricular septum and the infiltration of the anterior wall of both ventricles did not allow a radical surgical excision. At the end of surgical intervention pulmonary artery and outflow tract of RV were reconstructed with pericardium bovine patch.

Histological examination showed the presence of monomorphic spindle cells arranged in abundant collagen stroma, a pattern suggestive of cardiac fibroma. Because of the massive intracardiac infiltration and relationships, as stated before, a complete radical surgery was not an option and the patient was thus addressed to heart transplantation.

One year after surgery, the patient is still asymptomatic and awaiting heart transplantation; echocardiographic evaluations did not document increase in residual lesions size (Figure 2).

Discussion

Cardiac fibroma usually presents as a solitary lesion and rarely regresses spontaneously, as rhabdomyoma conversely does. It is characterized by slow growth and late development of symptoms

related to the obstruction of the ventricles outflow tracts; nevertheless an earlier onset of malign arrhythmias can occur, potentially leading to sudden cardiac death that can represent the first clinical manifestation of the disease.

This case is quite peculiar because of the complete lack of symptoms or semiological signs in the presence of a considerable intraventricular mass in the right ventricular outflow tract ($10 \times 10 \times 8$ cm), in which symptoms related to pulmonary overload were expected. To the best of authors' knowledge the present case is the biggest asymptomatic cardiac fibroma described in literature. Although one third of cardiac fibromas may produce a specific symptom (such as heart murmurs, palpitations, fatigue) or be an incidental finding, these clinical conditions have been described only in association with significantly smaller lesions [3].

Despite biopsy is essential for definite diagnosis of cardiac fibroma, non-invasive imaging techniques also give crucial information about the nature and the feature of the lesion; mass location, size and anatomic relationships are indeed key prognostic factors and determinants of clinical manifestation and therapeutic planning. For instance, Torimitsu showed that early age presentation and primary involvement of the interventricular septum are negative prognostic factors for the premature involvement of the conduction system [4].

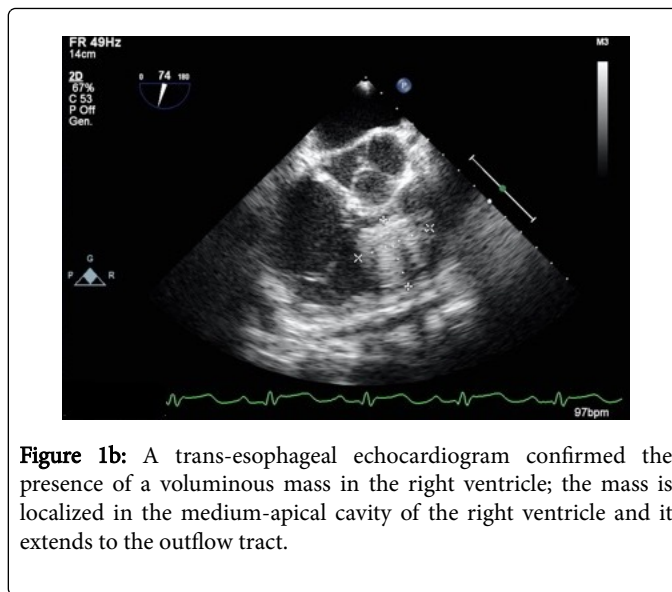


Figure 1b: A trans-esophageal echocardiogram confirmed the presence of a voluminous mass in the right ventricle; the mass is localized in the medium-apical cavity of the right ventricle and it extends to the outflow tract.

This case furthermore arouses some considerations about the prognostic classification of the tumor itself; the definition of "benign" is basically driven by the weak local infiltration tendency and the poor metastatic propensity. From our and other recent reports it can be argued that this definition does not perfectly fit the natural history of cardiac fibroma.

As we showed vast fibromas can be asymptomatic and symptom occurrence can be associated with already severe obstruction of the inflow/outflow tract of the ventricles or, even worse, as previously reported, with malignant arrhythmias and sudden cardiac death [5]. In conclusion according to authors the "take-home" message is that cardiac fibroma, despite a benign tumor, has to be considered a subtle disease because it can be silent up to very significant sizes and trigger fatal arrhythmias as first presentation.

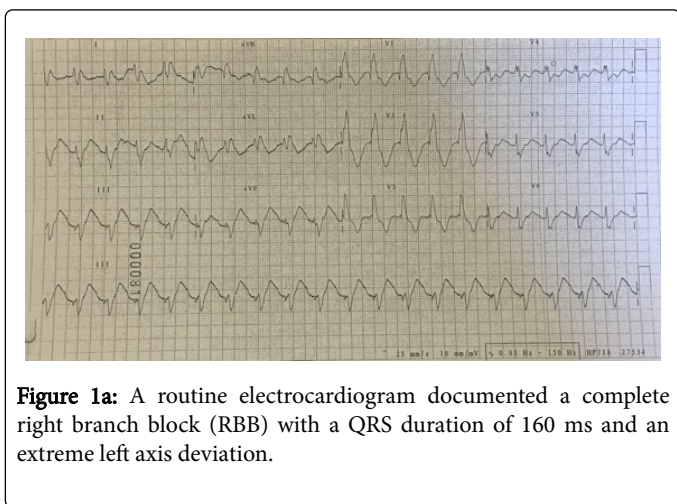


Figure 1a: A routine electrocardiogram documented a complete right branch block (RBB) with a QRS duration of 160 ms and an extreme left axis deviation.

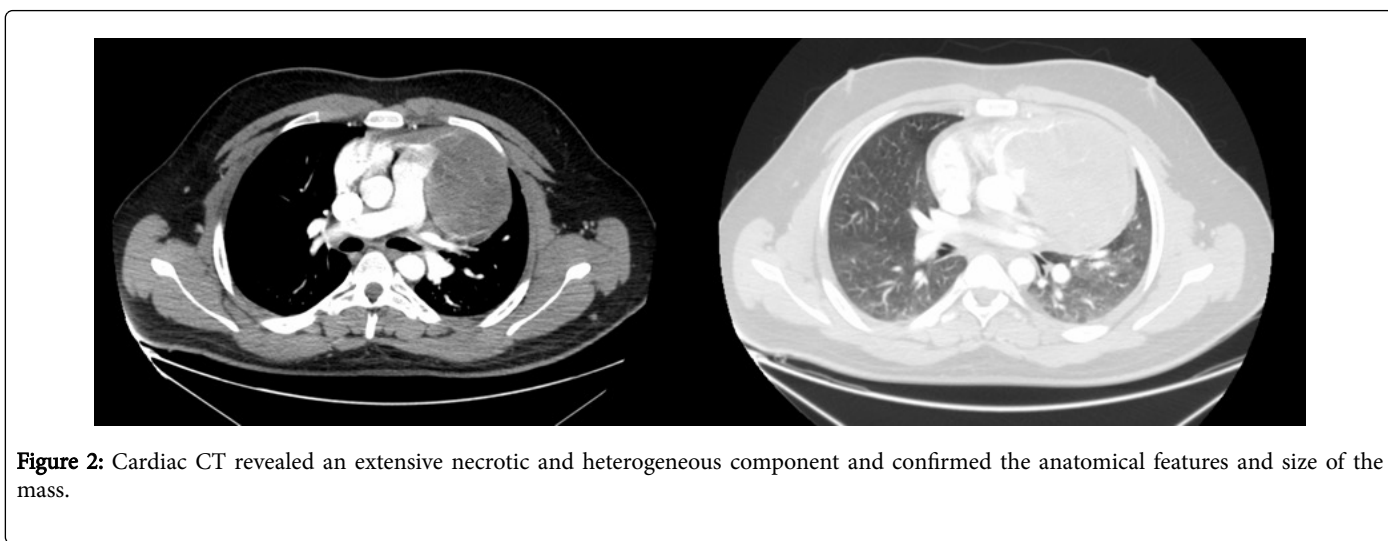


Figure 2: Cardiac CT revealed an extensive necrotic and heterogeneous component and confirmed the anatomical features and size of the mass.

Acknowledgments

Fortunato Iacovelli is currently attending the Cardiopath PhD program.

Conflict of Interest

The authors declare that they have no conflict of interest.

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