

The Role of Three-Dimensional Echocardiography in Interventricular Mass Evaluation

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A young patient underwent a screening electrocardiogram (EKG) that suggested apical hypertrophic cardiomyopathy. Serial investigation with echocardiography showed a well-defined hyperechogenic mass involving the interventricular septal. To better define the lesion extension three-dimensional (3D) echocardiography was done and it demonstrated a mass invading the septal myocardium, involving the major part of the muscular portion. The findings were highly suggestive of a cardiac fibroma. A cardiac magnetic resonance image (MRI) was also compatible with this diagnosis. In our case, 3D echo showed a high accuracy, proving to be a useful tool to determine the anatomy of the lesion, complementary to MRI, guiding best management strategy. (Echocardiography 2013;30:E125-E127)

Key words: cardiac fibroma, cardiac mass, echocardiography, three-dimensional echocardiography, magnetic resonance imaging

Case:

A 21-year-old asymptomatic man presented for a screening evaluation. Twelve-lead electrocardiogram (EKG) (CARDIOVIT AT-2, Schiller, Baar, Switzerland) suggested the apical form of hypertrophic cardiomyopathy (Fig. 1A). He underwent a treadmill exercise test that registered several episodes of nonsustained ventricular tachycardia (VT) during the recovery phase (Fig. 1B). Twenty-four hour EKG detected several episodes of sustained VT. Two-dimensional transthoracic echocardiography (2DE) (Vivid E9 model BT11, General Electric, Horten, Norway) four-chamber apical view revealed a well-defined 6.5×4 cm hyperechogenic mass involving the septal mid and apical segment, without signs of flow obstruction on Doppler evaluation (Fig. 1C and D). Although, borders and grade of myocardial extension were poorly defined in a 2DE short-axis view (Fig. 1E). To establish a better anatomical correlation, a three-dimensional transthoracic echocardiography study (3DE) was also performed. Apical multiple-beat full volume

acquisition, through cropping in multiple views, could characterize this large well-defined hyperechogenic septal mass. This revealed its extension from the septal myocardium, involving the major part of the muscular portion, until to the apex and also a tiny lateral and inferior wall involvement. (Fig. 1F, G and H and movie clips S1–S8). Echocardiographic findings may reasonably predict the specific nature of encountered cardiac masses. Cardiac fibroma usually appears as a solitary intramural homogeneous hyperechogenic lesion, rounded, with a fibrous whorled cut surface.^{1,2} The usual location is in the ventricular septum or left ventricular free wall.² Calcification on the central portion of the tumor is pathognomonic for fibroma reflecting poor blood supply to the mass.¹ Differentiation from other intramural masses may be done. Echocardiography is diagnostic for rhabdomyomas, and shows multiple homogeneous bright intramural masses with luminal extensions.³ Lipomas are encapsulated intracavitary, intramyocardial, or epicardial hypoechoic or isoechoic masses.³

A cardiac magnetic resonance imaging (MRI) (Signa HDxt 3.0T, General Electric, Milwaukee, WI, USA) study confirmed 3DE findings of an intramural aspect mass (Fig. 1L and J and movie clips S9–S10). The mass showed low signal on T₁- and T₂-weighted images, no increase in signal during first-pass perfusion, but diffuse delayed hyperenhancement after gadolinium injection (Fig. 1K and L), a feature typical of cardiac

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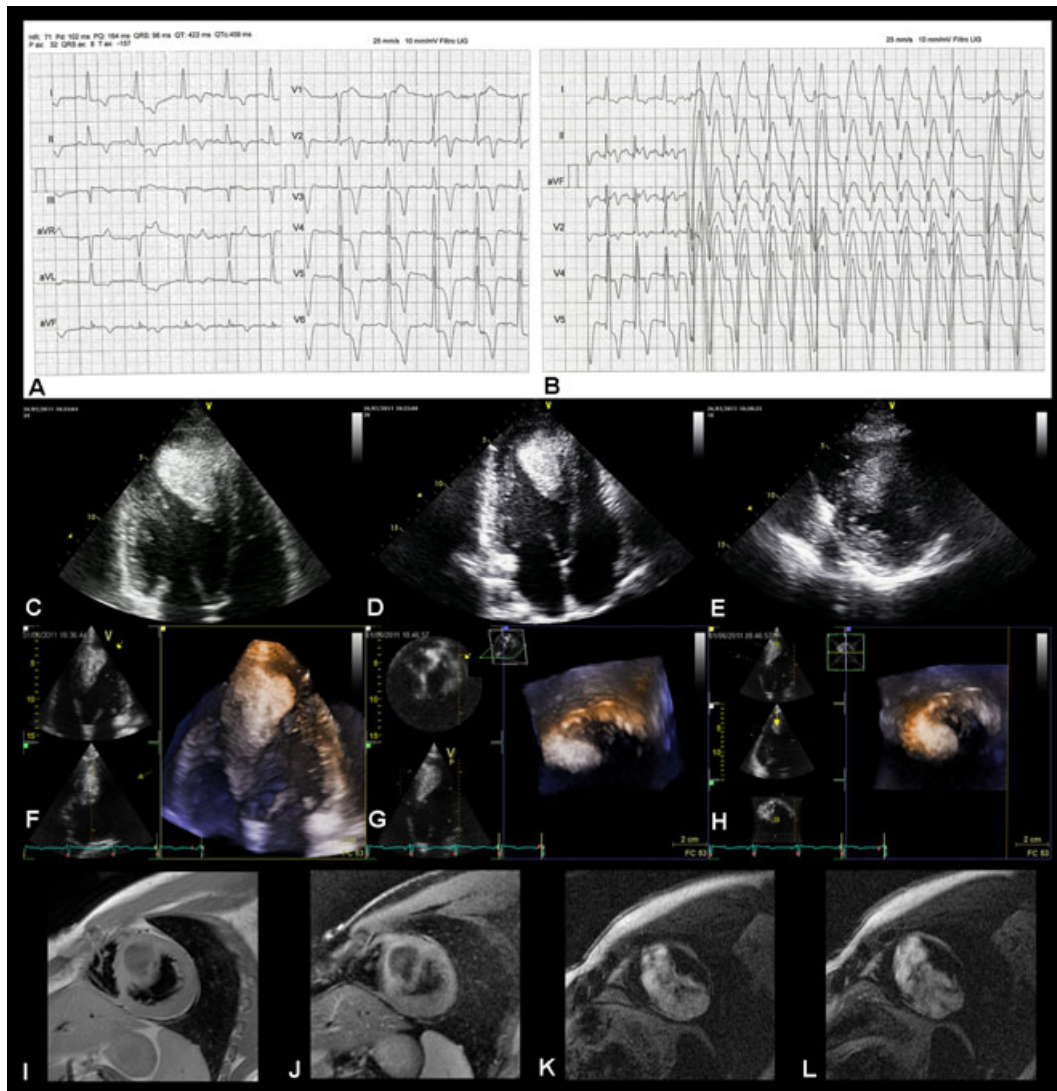


Figure 1. **A.** Rest twelve-lead electrocardiogram (EKG); **B.** Nonsustained broad-complex tachycardia registered during treadmill exercise test at recovery phase; **C.** and **D.** Four-chamber apical view revealing a well-defined hyperechogenic mass involving the septal mid and apical segment – left ventricle on left side; **E.** Short-axis view in a two-dimensional echocardiography evidencing a septal thickness in the same mass topography with poorly edge definition; **F.** Interventricular septum view from left ventricle; **G.** and **H.** In a short-axis cropping view could characterize this large well-defined hyperechogenic septal mass; **I.** and **J.** T₁- and T₂-weighted MRI image; **K.** and **L.** Delayed MRI acquisition images showing an extensive contrast enhancement.

fibroma.^{1,4,5} This feature reflects contrast accumulation within an expanded extracellular compartment by deposition of collagen and elastin fibers.¹ On MRI, fibroma is the only intramural tumor to have a combination of an intermediate-low signal on T₁-weighted images plus a low signal on T₂-weighted images, which reflects small intracellular and vascular space and low water content.^{1,4}

MRI can help to make this differential diagnosis between other cardiac masses based on the presented characteristics. Myxomas can also have an intermediate signal on T₁ and a

low signal on T₂ images, but they are intracavitary in nature.¹ A lipoma has a typically high T₁ signal due to high fat content.^{1,3} Cardiac rhabdomyomas are frequently multiple and return an intermediate-high signal on T₁ and an intermediate signal on T₂ images.¹ Hemangiomas are enhanced with first-pass perfusion and have an increased signal on T₂ images (probably because of slow-flowing blood in the tumor vessels).^{1,3} Finally, malignant tumors tend to return a high signal on T₂-weighted images as a result of increased water content due to cellular necrosis and tissue inflammation.¹

Due to the unique imaging features typical of fibroma, surgical biopsy was not undertaken and due to the size, location, and operative risk a conservative management was decided. Patient was treated with an internal cardioverter defibrillator (ICD) implantation and β blocker prescription. No significant changes in mass features were noted in a follow-up 3DE a year later. ICD readings showed only isolated episodes of nonsustained VT without the need of any therapy. Cardiac fibroma is rare benign connective tissue tumor. Most originated from interventricular septum or left ventricle, is often diagnosed in childhood and rarely observed in adults.^{2,6} Although benign, they are clinically important as they may present with syncope, heart failure, cyanosis, inflow and outflow obstruction, or sudden death. Surgical treatment is advocated even in asymptomatic cases.^{7,8} While not invasive, cardiac fibromas do not have a distinct capsule and they can entrap islands of myocardium, giving the impression of local infiltration.² Often, the complexity of resection and reconstruction procedures seems at high risk or unfeasible and could alter the ventricular geometry or mitral valve function.⁷ For these reasons accurate imaging evaluation is mandatory for determining the lesion topography, myocardium involvement, and surgical treatment feasibility. An integrated image approach with Cardiac MRI and 3DE seems to be the best way to evaluate an interventricular septal mass. Cardiac MRI is usually performed and offers a unique tissue characterization, important to perform the diagnoses, but its availability is usually low and it has several contraindications. Indeed, echocardiography evaluation, easily available, also could suggest the diagnosis, providing information about the anatomic relationship and functional impairment of the mass. In our case, 3D echo showed a high accuracy, proving to be a useful tool to determine the anatomy of the lesion, complementary to MRI, guiding best management strategy.

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Supporting Information

Additional Supporting Information may be found in the online version of this article:

Movie clip S1. Three-dimensional echocardiography full volume acquisition. Left ventricular longitudinal cropping providing a four-chamber view.

Movie clip S2. Three-dimensional echocardiography full volume acquisition. Left ventricular longitudinal cropping providing a four-chamber view.

Movie clip S3. Three-dimensional echocardiography full volume acquisition. Left ventricular longitudinal cropping providing a four-chamber view.

Movie clip S4. Three-dimensional echocardiography full volume acquisition. Left ventricular longitudinal cropping providing a four-chamber view.

Movie clip S5. Three-dimensional echocardiography full volume acquisition. Left ventricular longitudinal cropping providing a short-axis view.

Movie clip S6. Three-dimensional echocardiography full volume acquisition. Left ventricular longitudinal cropping providing a short-axis view.

Movie clip S7. Three-dimensional echocardiography full volume acquisition. Left ventricular longitudinal cropping providing a short-axis view.

Movie clip S8. Three-dimensional echocardiography full volume acquisition. Left ventricular longitudinal cropping providing a short-axis view.

Movie clip S9. Cardiac MRI short-axis view.

Movie clip S10. Cardiac MRI multiple longitudinal views.