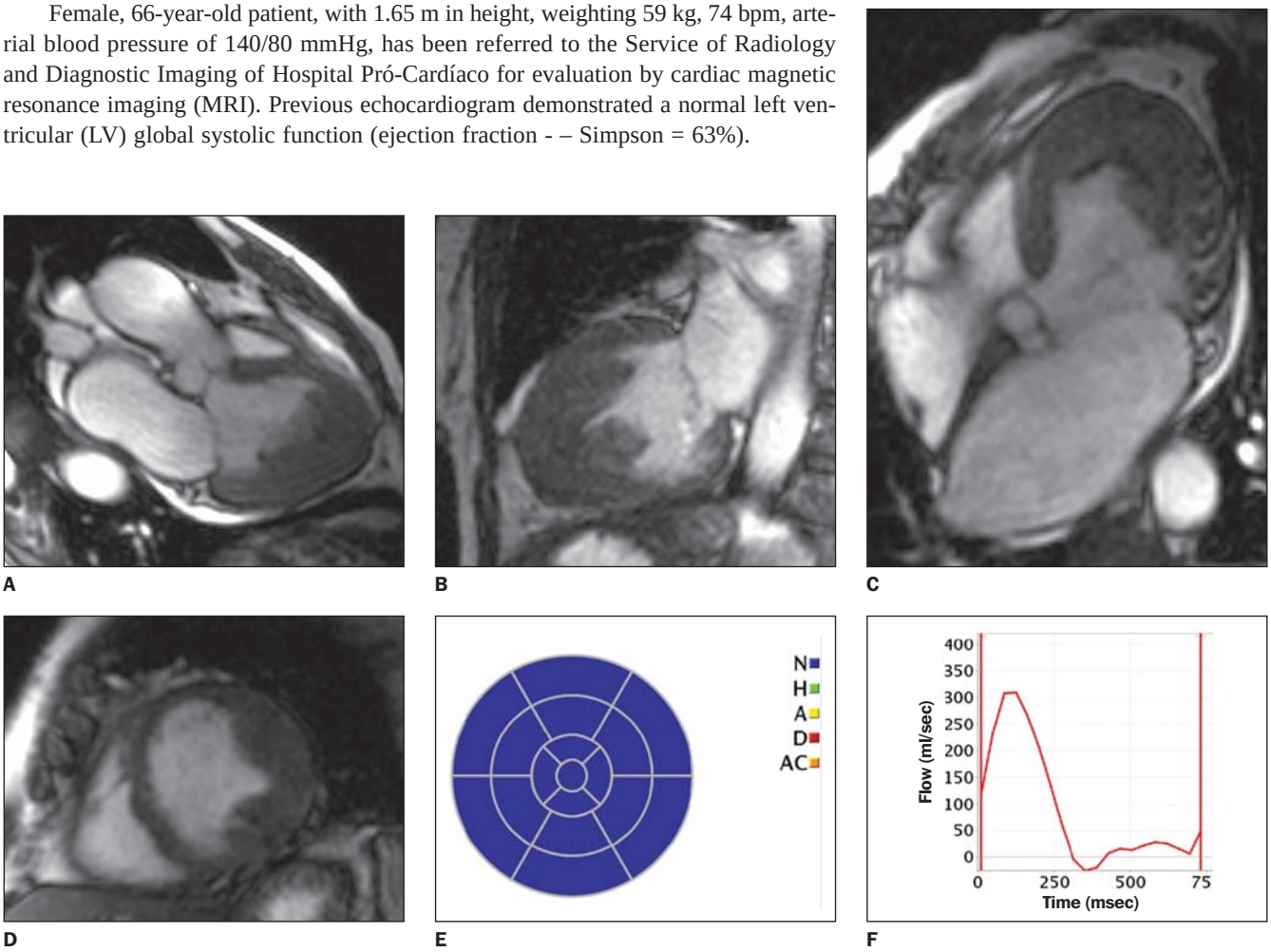
WHICH IS YOUR DIAGNOSIS?

Marcelo Souto Nacif[1,4], Amarino Carvalho de Oliveira Júnior[2], Denise Madeira Moreira[3], Mônica Regina Nagano[4], José Hugo Mendes Luz[4], Mauro Esteves de Oliveira[4], Arnaldo Rabischoffsky[5], José Feldman[6], Carlos Eduardo Rochitte[7]

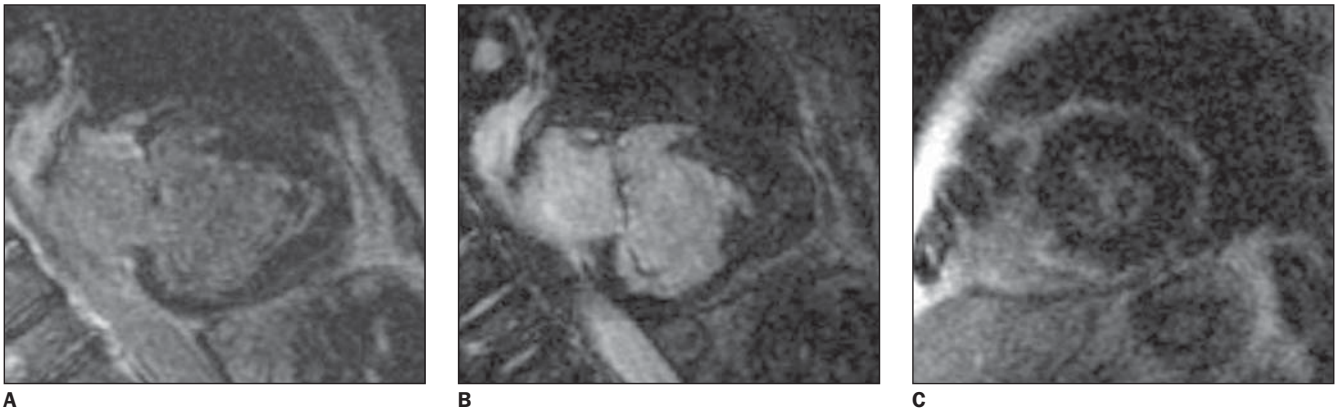
Study developed at Hospital Procardíaco, Rio de Janeiro, RJ, Brazil. 1. Professor at Faculdade de Medicina de Teresópolis and IPGMCC Post-Graduation Course, MD, Radiologist at HUAP-UFF. 2. Coordinator for Service of Radiology and Diagnostic Imaging, Hospital Procardíaco. 3. Sub-Coordinator for Service of Radiology and Diagnostic Imaging, Hospital Procardíaco. 4. MDs, Radiologists at Hospital Procardíaco. 5. Coordinator for Service of Echocardiography, Hospital Procardíaco. 6. MD, Cardiologist. 7. MD, Advisor for Hospital Procardíaco. Mailing address: Prof. Dr. Marcelo Souto Nacif. Rua Álvares de Azevedo, 130, ap. 704/A, Icaraí. Niterói, RJ, Brazil 24220-042. E-mail: msnacif@yahoo.com.br

Female, 66-year-old patient, with 1.65 m in height, weighting 59 kg, 74 bpm, arterial blood pressure of 140/80 mmHg, has been referred to the Service of Radiology and Diagnostic Imaging of Hospital Pró-Cardíaco for evaluation by cardiac magnetic resonance imaging (MRI). Previous echocardiogram demonstrated a normal left ventricular (LV) global systolic function (ejection fraction - – Simpson = 63%).



A B C D E F

Figure 1. Electrocardiogram-gated acquisitions with cine Fiesta (SSFP), T1FGRET and T1FGRIR, at end-diastole, in the following planes: outflow tract (**A**), 2-chamber view (**B**), 4-chamber view (**C**), short-axis (**D**); (**E**) LV function map, and (**F**) aortic flow curve.



A B C

Figure 2. Electrocardiogram-gated acquisitions. Delayed enhanced image: longitudinal (**A,B**) and short axis (**C**).

Images description

Figure 1 – Electrocardiogram-gated acquisitions with cine-Fiesta (SSFP), T1FGRET and T1FGRIR, at end-diastole, in following planes: outflow tract (**A**), 2-chamber (**B**), 4-chamber (**C**) and short-axis (**D**); (**E**) LV function map; and (**F**) aortic flow curve. Observe that the atrium and right ventricle (RV) present with normal size, with preserved segmental and global functions. There is an apical filling of the left ventricular cavity with preserved LV medial and basal segmental functions. Minimum mitral regurgitation and increase in volume of the left atrium, besides mild aortic insufficiency.

Figure 2 – Electrocardiogram-gated acquisitions, longitudinal delayed enhancement with calcification (thrombus) (**A**), longitudinal (**B**) and short-axis (**C**). Subendocardial perfusion defect on the apex and delayed subendocardial enhancement of the LV apical filling surface. Presence of a central hypointense image in this region, which may correspond to an adhered thrombus/calcification.

Diagnostic: Endomyocardial fibrosis with gross calcifications and normal LV function. Mild aortic insufficiency.

COMMENTS

Endomyocardial fibrosis was first described by Davies, in 1948. It is a complex entity of yet undetermined etiology, endemic in several tropical countries, with varied forms of presentation, representing, in some regions, the primary cause of restrictive myocardiopathy. Possible etiologic factors are: serotonin-rich diet, malnutrition, filariosis and viral diseases[1–6].

Endomyocardial fibrosis is characterized (Chart 1) by formation of endocardial fibrous tissue, most frequently located in the apex of the affected ventricle, which, angiographically, characterizes the typical image of LV and RV apical amputation. Eventually, the fibrosis extends to the myocardium, the presence of a massive endocardial calcification, whatever its etiology, being a rare finding in the literature and with possibility of becoming a diagnostic and prognostic marker of the disease[1,6–11].

Chart 1 Endomyocardial fibrosis diagnosis.

- Identification of fibrosis in the ventricular chamber.
- Signs of restricted ventricular filling.
- Signs of cardiac valves involvement.
- Secondary signs.
- It is essential to determine if the involvement is uni- or bilateral, balanced, or predominant in one of the ventricles.

Endomyocardial fibrosis clinical manifestations depend on the form and intensity of cardiac involvement. For difficulting the ventricular relaxation, the fibrosis may cause restrictive cardiac failure. The presence of valvar insufficiency makes such manifestations even more noticeable. Such clinical manifestations differ according to the involvement of the cardiac chambers (predominance in right, left or both chambers)[1,3,9,12–15].

Imaging studies, chest x-ray, echocardiogram, nuclear medicine, magnetic resonance imaging and hemodynamic study, identify ventricular shape changes and allow the recognition and determination of the degree of the chamber involvement[4,6,11,16,17].

The echocardiogram demonstrates signs of restriction associated with a large atrial dilatation and ventricular chambers obstruction, and raises suspicion of endomyocardial fibrosis[4,14,18]. Echocardiogram should be the best propedeutic method for endomyocardial fibrosis diagnostic; however, considering the rarity of this disease, it is not always considered and there are evidences that echocardiography underestimates the fibrosis intensity and does not identify mild forms of the disease[5,6,10–12,19].

The hemodynamic study associated with angiography still is considered the golden standard for the diagnosis of endomyocardial fibrosis due only to its comprehensive use along years (Figure 3). However, it is important to observe that the cardiac MRI is already demonstrating its great diagnostic value[1,4,5,8,13].

The pressoric data with ventricular pressure curves presenting a "square root" pattern identify the cardiac restriction, while angiography demonstrates the apex and inflow tract amputation, outflow tract dilatation, atrial increase and signs of atrioventricular valves insufficiency. The hemodynamic study results also are important for evaluation of the degree of cardiac involvement, this factor being of prognostic importance[6,8,13,17,18].

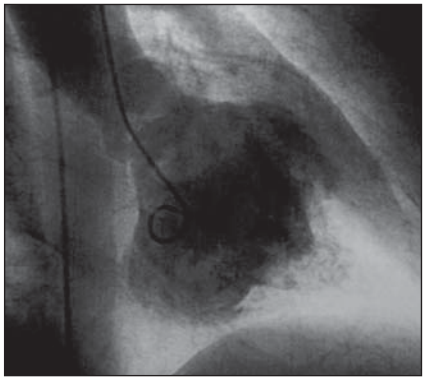


Figure 3. Left ventriculography, 30° right anterior oblique view. Decreased end-diastolic volume, apical obliteration associated with calcification, competent mitral valve.

The relevance of the diagnosis of endomyocardial fibrosis originates from the fact the symptomatic presentation of this disease has a poor prognosis, a fact which has been documented since the first publications on this matter. We have observed that patients with endomyocardial fibrosis confined to LV are most frequently asymptomatic, and that the disease presents a fair evolution, even in those patients with a significant ventricular fibrosis. In these cases, the presence of mitral insufficiency is a determining factor for symptoms onset[11,15,17,18].

The surgery changes the natural history of the disease, reducing symptoms and mortality. Because of their fair evolution, asymptomatic patients - especially those with isolated LV involvement -, just should be maintained on clinical follow-up[10–13,16,18].

Signs of restricted ventricular filling

Alteration of the LV posterior wall motion dynamics – On the LV posterior wall one observes a fast protodiastolic motion followed by rectification in the remaining period, nearly always without the typical telediastolic expansion secondary to the volume resulting from the atrial contraction. This is a sign of diastolic restriction, also frequently found in

constrictive pericarditis. Curiously, contrary to what could be assumed, this LV posterior wall motion is not an exclusive feature of isolated or bilateral left ventricular endomyocardial fibrosis, and may be observed in isolated presentations of right-sided endomyocardial fibrosis[5,11,15,17].

Differential diagnosis

In right-sided endomyocardial fibrosis, a differential diagnosis with constrictive pericarditis and Ebstein's disease should be established. Some signs are common to the three diseases, like alteration in the interventricular septum movement, presystolic opening of the pulmonary vein, anterograde diastolic flow in the right ventricle outflow tract and right atrium dilatation. It is worth to emphasizing that, in constrictive pericarditis, the tricuspid valve is structurally normal, and there is no reason for the left ventricle outflow tract to dilate. In the Ebstein's disease, there is no sign of restriction and the anterior leaflet of the tricuspid valve is redundant. Additionally, in Ebsteins's disease, the tricuspid reflux is situated more apically, contrarily to endomyocardial fibroses, whose reflux is slow and of undefined localization[1,3,8,9,12,19].

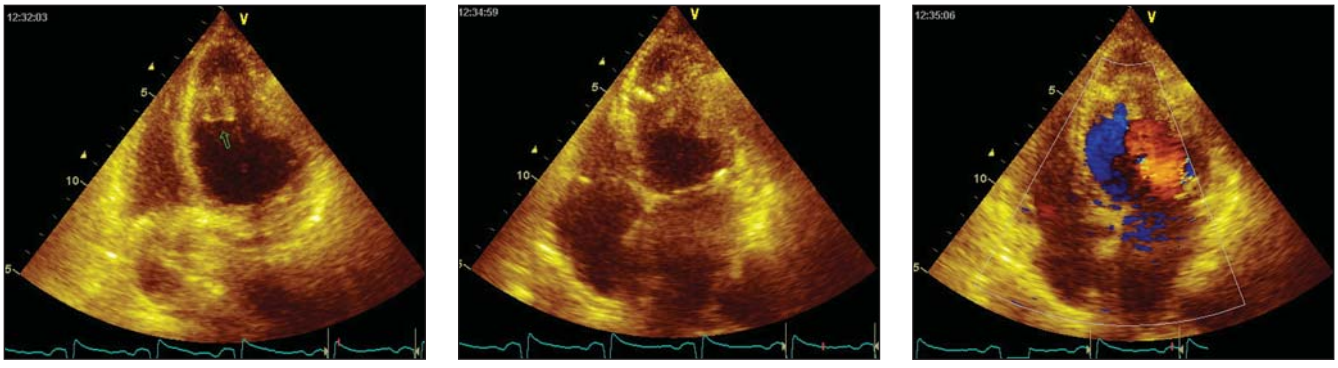
In left-sided endomyocardial fibrosis, the differential diagnosis should be done with other mitral insufficiency etiologies, especially rheumatic fever. In this circumstance, the differentiation is relatively easy because of the presence of mitral valve leaflets thickening and adherence. In dilated cardiomyopathies with functional regurgitation and eventual formation of apical thrombus, the hypocontractility is diffuse and the valve structural aspect is normal. In apical thrombosis of chronic chagasic myocarditis or ischemic cardiopathies, the wall is akinetic or presents dyskinetic movement, which does not occur in endomyocardial fibrosis[1,2,4,5,11,16,18].

In the present case, the echocardiogram (Figure 4) had already demonstrated signs of LV apical filling with preserved contraction and normal systolic function.

Later, we could observe a chest CT of this patient (Figure 5), which demonstrated endocardial gross calcifications.

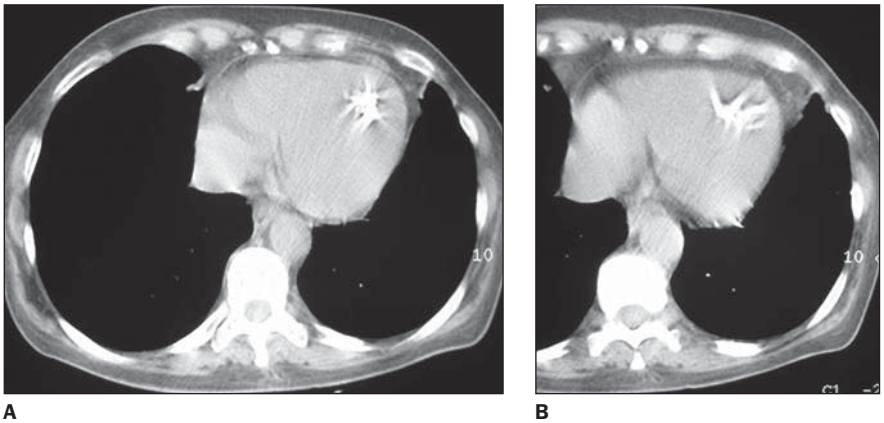
Presently, cardiac MRI is a mainstay in the evaluation of the myocardium thickness and function, as well as the LV perfusion, allowing the assessment of the myocardial enhancement, which is a relevant factor in the diagnosis of several cardiac diseases.

It is important to emphasize that, in our service, the technological development has allowed us to develop a multidisciplinary work, integrating the several examination methods in cardiology, and involving radiologists in active clinical discussions and in the study of heart diseases, bringing an important contribution to the diagnostic and therapeutic decisions making process in this field.



A B C

Figure 4. Echocardiogram with color Doppler demonstrating left atrium dilatation. LV apical filling, with preserved contraction in this region, and endocardial margins calcification. Normal LV systolic function, with decrease in the apical relaxation. Also, mitral ring calcification was found, with mild mitral and aortic insufficiencies.



A B

Figure 5. Chest computed tomography, mediastinal window, where endocardial gross calcifications can be observed.

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