WHICH IS YOUR DIAGNOSIS?

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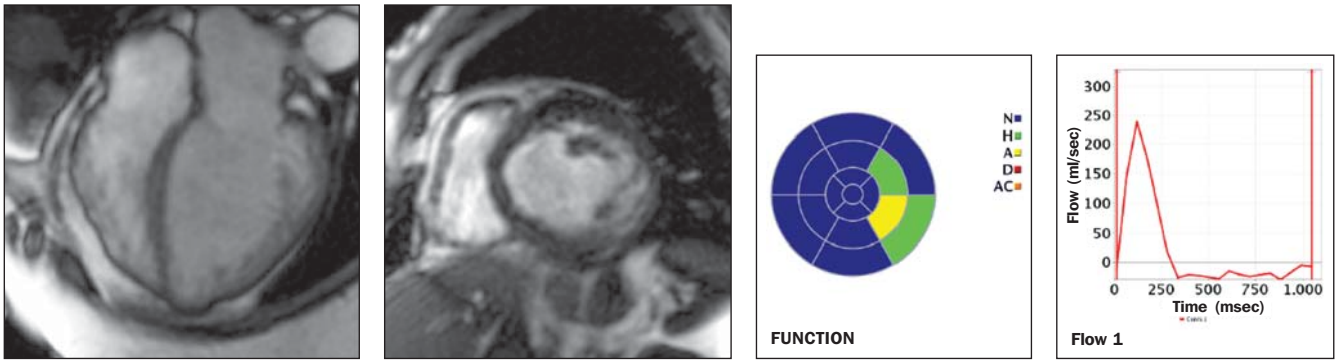
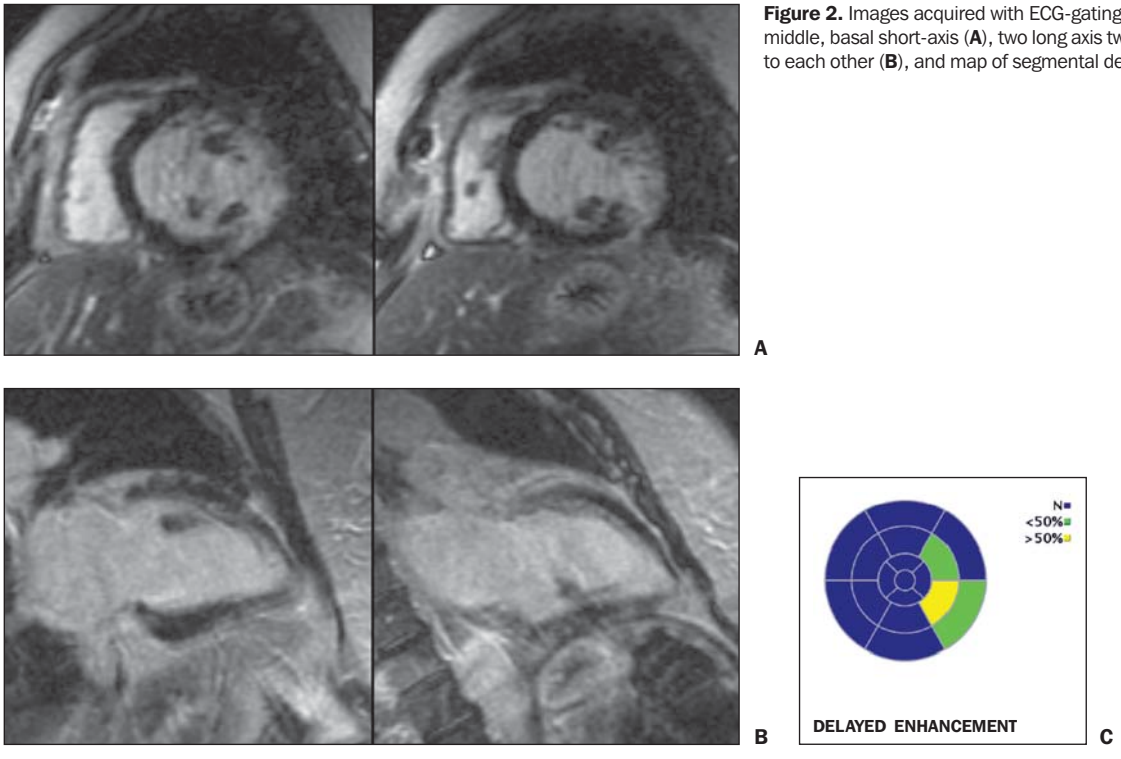
A female, 67-year old patient weighting 80 kg, with 1.53 m in height, coming from Minas Gerais state, has been referred to the Service of Radiology and Diagnostic Imaging at Hospital Pró-Cardíaco to be submitted to magnetic resonance imaging (MRI) of the heart.



A B C D

Figure 1. Images acquisition with ECG-gating, in cine-Fiesta sequence (SSFP) at end-diastole, in the following planes: four-chambers (**A**), middle, basal short-axis (**B**), map of segmental function (**C**) aortic flow curve (**D**).



A

B C

Figure 2. Images acquired with ECG-gating. Delayed enhancement – middle, basal short-axis (**A**), two long axis two-chamber perpendicular to each other (**B**), and map of segmental delayed enhancement (**C**).

Description of images

Figure 1. Images acquisition with ECG-gating, in cine-Fiesta sequence (SSFP), at end-diastole, in the following planes: four-chamber (**A**), middle, basal short axis (**B**), map of segmental function (**C**) and aortic flow curve (**D**). Observe normal-sized atria, normal right-ventricular volume with preserved ventricular function. There is a mild dilatation of the left ventricle, with thinning of the medial and basal, infero-lateral walls, a slight global left-ventricle (LV) dysfunction, estimated ejection fraction of 47.1%. Also, akinesia was identified in the medial, infero-lateral segment, hypokinesia in the basal, infero-lateral and medial antero-lateral segments, with normokinesia in the other segments. Note the typical aspect of apical aneurysm and mild aortic insufficiency.

Figure 2. Images acquired with ECG-gating. Delayed enhancement, middle basal short-axis (**A**), two long axis two-chamber perpendicular to each other (**B**), and map of segmental delayed enhancement (**C**). Observe that there was a delayed myocardial enhancement (< 50% of segmental area) in the medial antero-lateral and basal infero-lateral segments, and delayed intramural enhancement (> 75% of the segmental area) in the medial, infero-lateral segment.

Diagnostic: Chagasic cardiomyopathy.

COMMENTS

Firstly described by Carlos Chagas in 1909[1], American trypanosomiasis or Chagas disease — a condition caused by the Trypanosoma cruzi, a protozoan parasite of the Mastigophora class, order Kinetoplatida and Trypanosomatidae family — is a highly prevalent antropozoonosis, with high morbidity in the Latin America, and is considered as one of the parasitic infectious diseases with higher incidence in the continent[2,3]. As a matter of fact, overall prevalence of Chagas disease in Latin American countries is estimated at 16 to 18 million cases, and about 100 million people at risk of contracting the infection[4,5].

The pathological findings are highly suggestive of the disease. Macroscopically, chronic chagasic cardiopathy is characterized by a progressive, chronic myocarditis with increase in the myocardial muscle— which may reach more than 1,000 grams —, leading to the four cavities dilatation, and giving the heart a globoid appearance[6,7]. Usually, the myocardium presents softened, with irregular thickness and areas where the thickness of the wall is decreased, and others were the wall is hypertrophic. The pericardium may present smooth, bright, transparent, or thickened. Frequently, the valvular endocardium is not involved, except when there is a remarkable dilatation of the ring with dysfunction and insufficiency, originating valvular fibrous thickening. Microscopically, the presence of chronic myocarditis in association with fibrosis is observed, in some cases with a granulomatous component (separation of the fibers with formation of pseudofollicular structures with epithelioid cells and giant, multinuclear, non-parasited cells), amastigote nests being rare. Cardiac cells demonstrate several alterations, such as vacuolation, accumulation of lipofuscin granules, hyaline degeneration, intracellular edema and myofibrilar disorganization[6,7].

Chronic chagasic cardiopathy manifestations will depend on the degree of myocardial function involvement. So, clinical-pathological presentations are the following:

1) Cardiac failure syndrome (CFS). One of the lesions typically related to CFS — but also to thromboembolic alterations — is the apical aneurysm that may range from a fibrous thinning to true aneurysmal formations resulting from sacular dilatation with few millimeters in diameter (up to 5 cm), whose wall is some times constituted uniquely by the union between endocardium and pericardium (Figure 3).

2) Arrhythmic syndrome whose pathological substrate includes involvement of the sinoatrial node, atrioventricular node and bundle of His, with the following main alterations: a) disorders of atrio- and intra-ventricular conduction; b) functional disorder of the sinusal node; c) primary and secondary ventricular repolarization abnormalities; d) fibrosis and inflammation; e) autonomic dysfunction; f) endothelial and coronary dysfunction. All of these disorders may lead to difficult-to-manage arrhythmias[8]. Sudden death predominates as frequent cardiac cause of death in all presentations of the disease in endemic areas, and is erroneously diagnosed as acute coronary syndrome.

3) Thromboembolic syndrome, a frequent finding in patients with CFS, although rarely diagnosed, is a factor responsible for both the refractoriness to treatment and a higher risk for death. This syndrome probably is a result of: a) stasis secondary to cardiac dilatation; 2) arrhythmias; 3) endocardial fibrosis; 4) mural endocarditis. Most affected systemic organs in decreasing frequency

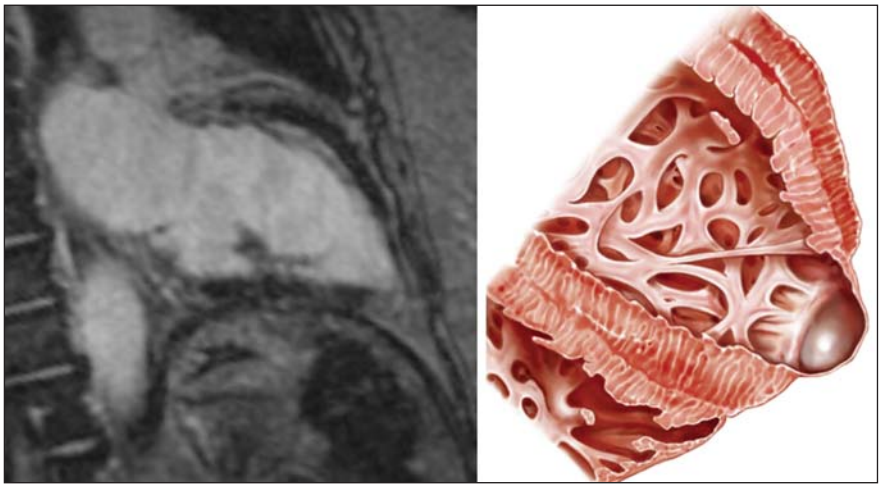


Figure 3. Apical aneurysm in the chronic chagasic cardiopathy.

order are the kidneys, the spleen and the brain[9,10].

Cardiac magnetic resonance imaging contribution in Chagas disease

Cardiac involvement is a determining factor in the Chagas disease prognosis. Therefore, an appropriate evaluation of the heart is essential in the management of the patient. Approximately 30% to 40% of infected peoples will develop cardiac abnormalities during their lives, but only 10% to 20% will present the symptomatic form of the disease.

Histopathological studies indicate the presence of mild chronic myocarditis manifested by the scattered mononuclear cell infiltrate with the surrounding myocytes undergoing several degrees of degeneration and necrosis. The focal and diffuse fibrosis is prominent in the myocardium and conduction system. Myocardial fibrosis has been detected in several anatomopathological studies for Chagas disease where the prevalent sites for fibrosis and left ventricular (LV) microcirculation abnormalities were the LV apex and the basal, infero-lateral segment (Figure 4).

Similarly to data found in the literature, the segmental analysis by MRI and the delayed myocardial enhancement clearly demonstrate *in vivo* that the LV apical and infero-lateral regions are the most frequent sites for myocardial fibrosis in patients with Chagas disease[11,12]. This corroborates the concept that in the Chagas disease, myocardial fibrosis is frequently related to regions of terminal circulation like the apex (terminal circulation between the anterior descendent coronary artery and descendent posterior) and the basal, infero-lateral segment (terminal circulation between the right coronary artery and the left circumflex artery)[11,12].

Future prospects

Recent technological developments in cardiac magnetic resonance imaging have been utilized to detect and quantify early signs of cardiac involvement in Chagas disease. These signs may constitute significant prognostic information. This has already been routinely made by means of a detailed evaluation of the cardiac function with the myocardial tagging technique, High-resolution cine-MRI, and delayed enhancement for detection of myocardial fibrosis. Cardiac MRI study in Chagas disease will include acquisitions in a single breath-hold with cine-3D, the techniques of myocardial perfusion, edema and myocardial inflammation detection, and monitoring of intramyocardial stem-cell injections for treatment of the chagasic cardiomyopathy.

Our view is that cardiac MRI may be utilized in the future for selection of patients with very early myocardial involvement by the Chagas disease. With this data, we can develop new therapeutic methods to change the natural history of the Chagas disease.

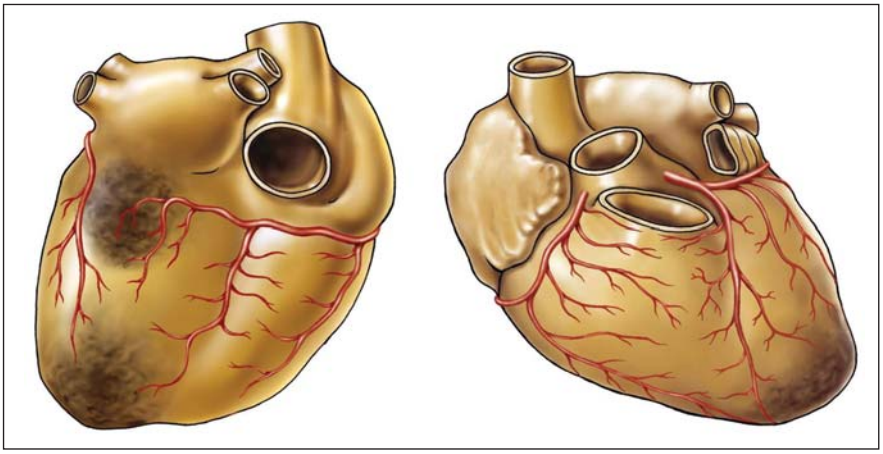


Figure 4. Posterior and latero-apical views of LV, demonstrating with a black stain the regions with more extensive myocardial involvement in Chagas disease.

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