Stone heart or apical retraction and calcification in Chagas’ cardiomyopathy

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Case
A 42-year old Caucasian male was found with first-degree atrioventricular block, right bundle branch block, and left anterior hemiblock on electrocardiogram (Panel A) and cardiomegaly on chest X-ray (Panel B). The left ventricle (LV) was severely dilated by echocardiography with a spherical shape (diastolic dimension 75 mm), reduced systolic function (EF 45%), and an akinetic, retracted LV apex with prominent endomyocardial calcification (Panels C and D). Coronary angiography revealed normal coronary arteries (see Supplementary data online, Video S1), and LV angiography confirmed calcification (Panel E), retraction, as well as akinnesia and fixation of the apex (Panel F) (see Supplementary data online, Video S2). Cardiac magnetic resonance imaging showed a spherically dilated LV (see Supplementary data online, Video S3), with late gadolinium enhancement in the LV apex suggestive of focal transmural fibrosis (Panel G).

The patient had consulted as a software specialist, with more than 50 trips to Honduras and Bolivia. This led us to suspect Chagas’ cardiomyopathy. The ELISA serology test was negative, but subsequent polymerase chain reaction (PCR) assay was positive for Trypanosoma cruzi, confirming the diagnosis. Oesophageal X-ray showed normal barium swallow (Panel H). After 2 months of treatment with benznidazole, the PCR had turned negative.

Chagas’ disease is endemic in Latin America, with transmission by triatomine insects, and causes chronic cardiac and gastrointestinal disease (megaoesofagus or megacolon) 10–30 years after initial infection. Apical aneurysm of the LV is the hallmark of Chagas’ cardiomyopathy and found in more than 50% of the cases, and can be associated with thrombo-embolic phenomena, congestive heart failure, ventricular tachycardia, as well as high-degree heart block. Echocardiography studies in Chagas’ cardiomyopathy have also found a significant decrease in apical curvature and disruption of the optimal global prolate-ellipsoid LV shape resulting in a more spherical LV shape, as seen in our patient. However, to our knowledge, apical retraction with endomyocardial calcification has thus far not been reported in Chagas’ patients (Panel I). This case illustrates that a ‘stone heart’ is also part of the clinical cardiac imaging spectrum of Chagas’ disease.

Supplementary data are available at European Heart Journal – Cardiovascular Imaging online.

Panel A. Twelve-lead electrocardiogram with conduction abnormalities in Chagas’ cardiomyopathy: first-degree atrioventricular block, right bundle branch block, and left anterior hemiblock (trifascicular block).
Panel B. Chest X-ray: cardiomegaly without signs of pulmonary oedema.
Panel C. Parasternal long-axis transthoracic echocardiogram showing a mildly dilated, spherically shaped left ventricle with moderate hypokinesis (ejection fraction 45%) and a retracted, akinetic left ventricular apex with calcification.
Panel D. Transeosophageal four-chamber view of a spherically shaped left ventricle with an amputated left ventricular apex and protruding right ventricular apex.
Panel E. Apical calcifications (arrow) on plain chest X-ray.
Panel F. Spherical left ventricular dilatation and apical retraction (arrow) on left ventricular angiography.
Panel G. Dilated spherical left ventricle with delayed gadolinium enhancement in the apex suggestive of transmural fibrosis (arrow) on cardiac magnetic resonance imaging.
Panel H. Normal barium swallow oesophageal X-ray.
Panel I. Diagram depicting the spherical dilatation of the left ventricle (solid line), apical retraction (dashed line), and apical endomyocardial calcification (arrow).