Rare association of endomyocardial fibrosis and Chagas heart disease

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Chagas heart disease (CD) and endomyocardial fibrosis (EMF) are distinct and uncommon cardiomyopathies that can lead to a very poor prognosis. Both diseases are mostly found in African and South- and Central American countries. The patient presented in this case was from an area in Brazil endemic for Chagas disease. CD results from infection by the protozoan Trypanosoma cruzi and is associated with typical electrocardiographic and echocardiographic findings, including posteroinferior wall motion abnormalities in the left ventricle, as well as apical aneurysm. On the other hand, the underlying cause and mechanisms of EMF remain unclear. Obliteration of one or both ventricular apices might be seen. The left ventricular (LV) ejection fraction is usually preserved.

In this case, a 63-year-old female patient from an area in Brazil endemic for CD was referred for cardiologic evaluation after an ischaemic stroke. The patient was asymptomatic, and the diagnosis of the chronic phase of Chagas disease was suspected based on the electrocardiographic and echocardiographic findings and confirmed by antibody detection using two tests: indirect immunofluorescence and chemiluminescence. The electrocardiogram showed sinus rhythm, right bundle-branch block, and left anterior hemiblock, which are common CD findings. On Holter monitoring, the patient also presented with several episodes of non-sustained ventricular tachycardia and no heart blockage.

No specific laboratory test can diagnose EMF. The electrocardiogram is neither sensitive enough nor specific enough to rule in or rule out EMF, and meticulous echocardiographic examination is fundamental to detect milder forms of EMF. The criteria often used for diagnosis of right ventricular (RV) EMF include apical obliteration, cleavage plane between the fibrous tissue and myocardium, parietal thrombosis, dilatation of the right atrium as evidenced by echocardiography, or cardiac magnetic resonance imaging, which is considered the gold standard technique for diagnosis of EMF.

The transthoracic echocardiogram showed hypokinesia of the lateral wall, preserved LV ejection fraction, discrete obliteration of the LV apex, signs of obliteration and akinesia of the RV apex, and a small pericardial effusion related to the inferior wall (A–F). Speckle-tracking echocardiography strain analysis confirmed the abnormalities in the lateral wall (G and H). Three-dimensional echo demonstrated apical RV hyperechogenicity (I–L and see Supplementary data online, VideoS1), and the cardiac catheterization revealed the absence of coronary artery lesions and apical amputation (see Supplementary data online, VideoS2). Cardiac magnetic resonance imaging with tissue characterization by late gadolinium enhancement showed heterogeneous abnormalities in LV lateral segments and a distinct homogeneous pattern,
exclusively endomyocardial, with biventricular apex involvement, associated with dark areas, representing thrombus formation, which have been described as a 'V signal' and strongly suggest EMF (M–P and see Supplementary data online, VideoS3).

Despite the occurrence of non-sustained ventricular tachycardia, and typical abnormalities of CD and EMF on cardiac imaging, the patient had no cardiovascular symptoms. This case illustrates a very rare association of two uncommon diseases and, to the best of our knowledge, is the first case reported in the literature. This case also reinforces the value of using new techniques for the proper diagnosis of this unusual association.

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

Conflict of interest: None declared.