Young Patient with Chagas Disease Presenting Initially as Acute Chest Pain and Treated for Acute Coronary Syndrome

Introduction

Chagas disease, a chronic and endemic condition in 21 countries within the Americas, affects approximately 6 million people. The northeast region has always been particularly important for the disease. According to data from the Mortality Information System (Sistema de Informação sobre Mortalidade), between 1980 and 2007, Brazil registered 156,224 deaths from Chagas disease; of them, 20,472 were in the northeast region and 3,144 occurred in Pernambuco.

Classic symptoms of Chagas disease can include chest pain, commonly typical for angina. Records of Chagas patients usually relate typical acute anginal pain to superimposed coronary artery disease. However, microcirculation disease has also been suggested as a chest pain trigger. Despite the high prevalence of Chagas heart disease in inland northeastern Brazil, coronary artery disease remains the main cause of heart failure and acute coronary syndromes.

This report describes a rare presentation of a neglected disease with a high regional prevalence. Thus, it reports the clinical condition and complementary tests of a young patient who presented with typical and acute anginal chest pain who entered the line of care for acute coronary syndrome.

Case report

Our patient was a 38-year-old man, a high-voltage electrician commonly working in rural areas inland within the northeast region and living in an urban area with good sanitary conditions, with hypertension and type 2 diabetes. The patient was admitted to the emergency room complaining of severe oppressive retrosternal pain triggered by exertion and associated with vertigo that started seven hours before admission. He reported experiencing a similar condition in the previous week with lower intensity that was triggered by exertion and improved with rest. He presented no significant changes on clinical examination.

A care line for acute coronary syndrome was defined, with an electrocardiogram (ECG) on admission showing no changes and high-sensitivity troponin testing negative (Figure 1). Transthoracic echocardiography showed a slightly increased left atrial volume (36 mL/m²), preserved right ventricular (RV) size and systolic function, tricuspid annular plane systolic excursion (TAPSE) of 22 mm, borderline left ventricular (LV) size and function (diastolic, 56 mm; systolic, 39 mm; indexed LV mass of 104 g/m²; and left ventricular ejection fraction [LVEF], 56%), and no identifiable contractility changes.

Cardiac magnetic resonance (CMR) (Figure 3 and Video 3) showed left atrial enlargement, an increased LV size, and mild global dysfunction (LVEF of 48%) with an area of dyskinesia in the apex and diffuse hypokinesia predominantly in the inferolateral wall. The presence of trabeculation was noted in the apical region with an aspect of non-compacted myocardium (NCM) and an NCM/compacted myocardium (MC) ratio of 3.04 in the lateral (apical) segment. Perfusion changes were noted at rest in the inferolateral wall along with an endocardial pattern of delayed enhancement occupying about 50% of the lateral segment (apical) and > 50% of the inferolateral segments (mid and basal).

Serology for Chagas disease by the chemiluminescence method was positive and confirmed by indirect immunofluorescence at a titration of 1:80. Finally, the patient was diagnosed with chest pain due to Chagas heart disease. Medical anamnesis usually attributes epidemiologically suspected Chagas disease to living in a mud house. However, in endemic areas, the risk of exposure can be occupational, as in the case described here.

Discussion

Diabetes and hypertension, as in the case reported here, significantly increase a patient’s risk of coronary artery disease due to its potential severity and high prevalence. In Brazil, ischemic heart disease is the leading cause of death, accounting for 31% of cardiovascular deaths. Typical acute pain as the first presentation of Chagas heart disease is rare.
Figure 1 – Electrocardiogram performed during emergency care.

Figure 2 – Myocardial scintigram showing perfusion at rest versus physical stress. Reduced marker uptake is evident on the anterior, lower, and left ventricular apex segments.

Video 1 – Right Coronary Artery.
Video 2 – Left Coronary Artery.

Figure 3 – Cardiac magnetic resonance image. (A) Two-chamber steady-state free precession cine image showing trabeculation and apical aneurysm (with a glove finger appearance). (B) Three-chamber steady-state free precession cine image showing trabeculation in the inferolateral myocardium. (C) Steady-state free precession cine imaging of the left and right ventricular short axis (apical section) showing trabeculation with a non-compacted myocardium/compacted myocardium ratio > 3.0; (D) short-axis delayed enhancement showing an area of fibrosis.

Video 3 – Cardiac Magnetic Resonance Imaging (CMR) showing an enlarged LA, increased LV dimensions, mild global dysfunction, with an area of dyskinesia in the apex and diffuse hypokinesia.
Cardiac changes in Chagas disease include tapered ventricular walls, biventricular enlargement, apical aneurysm, and mural thrombi, which may progress with signs of perfusion defects. The findings are possibly explained by the occurrence of microthrombus formation associated with microcirculatory spasms, endothelial dysfunction, and increased platelet activity.

Chest pain, commonly atypical, may be present in patients with Chagas disease. Conditions similar to those of angina pectoris can occasionally occur, being apparently related to microvascular changes including microthrombus formation, microcirculatory spasm, endothelial dysfunction, and increased platelet activity. Microcirculatory changes seem to amplify the inflammatory process and cause myocardial ischemia, although the subepicardial coronary arteries usually show no significant obstructions as reported here. Microcirculation changes commonly cause myocardial hypoperfusion in some areas naturally irrigated by fewer coronary branches, which are prone to aneurysm formation in the apical and posterior-basal LV walls.

CMR enables an accurate morphofunctional analysis that can be very informative, even in cases in which the echocardiographic image is inconclusive. It has great value in the detection of changes in ventricular geometry, such as the glove finger aneurysm (apical ventricular), a typical finding of Chagas cardiomyopathy. More recent studies highlight the potential of CMR to detect myocardial fibrosis regions in patients with Chagas cardiomyopathy and be a potentially valuable tool for these patients in the noninvasive risk prediction of sudden death, even in those with a preserved LVEF. The pattern of fibrotic involvement varies, with the presence of diffusely distributed, focal, and even transmural involvement simulating an area of fibrosis usually seen in myocardial infarction due to obstructive coronary disease as reported here.

A pattern similar to that of NCM has been reported for Chagas heart disease. Some characteristics are important to differentiating the etiology of myocardial involvement, including the presence of apical aneurysm (common in Chagas cardiomyopathy but unlikely in non-compaction) and the presence of transmural or epicardial fibrosis (more common in NCM, while in Chagas disease, the fibrosis tends to be subendocardial and adjacent to the trabeculation).

In the case reported here, the presence of this pattern of fibrosis that lacked a direct correlation with the typical acute chest pain on admission but was associated with the other CMR findings (more typical of Chagas heart disease) suggests that the myriad of findings are, in fact, related to changes resulting from Chagas disease.

In this case report, a young patient with previously unknown Chagas heart disease presented with unusual acute coronary syndrome. These findings confirm the need to consider the diagnosis of Chagas disease in cases of chest pain with consideration of the epidemiological profile of the region.

Authors’ contributions

Data collection: DRV Lima, JF Neto, JHC Silva, and PVAM Patriota; manuscript writing: JF Neto; critical review of the manuscript for important intellectual content: AML Silva and AC Armstrong.

Conflict of interest

The authors have declared that they have no conflict of interest.

References