ESC European Society of Cardiology

European Heart Journal - Case Reports (2023) **7**, 1–6
https://doi.org/10.1093/ehjcr/ytac487

CASE REPORT
Cardiovascular imaging

Case report: diagnosis of chronic Chagas cardiomyopathy using a multimodality imaging approach

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Received 6 June 2022; first decision 22 July 2022; accepted 23 December 2022; online publish-ahead-of-print 6 January 2023

Background In the USA, ~300 000 people are affected by Chagas heart disease, a growing, but commonly overlooked, public health issue. Chagas as a potential aetiology of dilated cardiomyopathy remains under-recognized. We present a case where multimodality imaging was essential in the diagnosis and management of Chagas heart disease.

Case summary A 54-year-old man, originally from Mexico, presented to the emergency department with chest pain and recurrent syncopal episodes, found to be in haemodynamically unstable ventricular tachycardia (VT) requiring urgent cardioversion. Urgent coronary angiography revealed no obstructive disease. A transthoracic echocardiogram revealed moderately reduced left ventricular systolic function (left ventricular ejection fraction 35–40%) with apical akinesis and an aneurysm of the apical septum. Cardiac magnetic resonance (CMR) confirmed a prominent apical aneurysm with dyskinesis of the apical septum, with the evidence of transmural myocardial late gadolinium enhancement of the entire left ventricular apex and a small apical thrombus. Serologic testing was positive for *Trypanosoma cruzi* IgG antibody, which was confirmed on repeat testing at the Centers for Disease Control and Prevention. Patient underwent VT ablation and was discharged on guideline-directed medical therapy including a regimen of anticoagulation, beta-blocker, and angiotensin-converting enzyme inhibitor therapies. He has had no subsequent syncope or VT.

Discussion Chagas heart disease remains under-recognized and under-diagnosed despite the growing burden of *T. cruzi* infection in the USA. Suspicion for Chagas heart disease should be considered in patients presenting with heart failure symptoms and ventricular arrhythmias with the right corresponding history and imaging findings on echocardiogram and CMR.

Keywords Chagas heart disease • Dilated cardiomyopathy • Ventricular tachycardia • Left ventricular apical thrombus • Case report

ESC Curriculum 2.1 Imaging modalities • 2.2 Echocardiography • 2.3 Cardiac magnetic resonance • 5.6 Ventricular arrhythmia • 6.5 Cardiomyopathy

Learning points

Case. A patient with dilated cardiomyopathy presenting with chest pain, ventricular tachycardia, and syncope, found to have Chagas cardiomyopathy.

- To be able to recognize the clinical presentation and typical imaging findings of Chagas cardiomyopathy.
- To understand the role and the importance of multimodality imaging in the diagnosis of Chagas cardiomyopathy.

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Handling Editor: Luca Arcari
Peer-reviewers: Chikezie Alvarez; Matthew Williams; Duygu Kocyigit; Sarandeep Kaur Marwaha
Compliance Editor: Nikolaos Spinthakis
Supplementary Material Editor: Michael Waight

Introduction

Chagas disease (CD), caused by the protozoan *Trypanosoma cruzi*, has been traditionally considered as a tropical disease, endemic to Latin America. While nearly 6 million people are diagnosed with CD in Latin America, international migration and globalization have led to growing prevalence in non-endemic areas such as Europe, USA, Canada, Japan, and Australia.[1–3] Yet, CD remains under-recognized and under-diagnosed in the non-endemic areas outside of Latin America. Approximately 50 000 people die annually due to CD; 60% due to sudden cardiac death (SCD), 25% due to heart failure (HF), and 15% due to stroke.[4] Several pathologic features have been noted on autopsy, including cardiomegaly with increased cardiac mass, ventricular wall thinning, aneurysm formation in the ventricles, and intracardiac thrombus.[1,5] Given the growing prevalence of this entity, CD should be considered as a potential aetiology in patients with unexplained non-ischaemic cardiomyopathy associated with arrhythmias, conduction system abnormalities, and left ventricular aneurysm. Multimodality imaging including the use of cardiac magnetic resonance (CMR) and transthoracic echocardiography (TTE) can play a critical role in diagnosing Chagas cardiomyopathy. We present a case where a high index of clinical suspicion and multimodality imaging led to the diagnosis of Chagas cardiomyopathy.

Timeline

Day 1	Presented to the emergency department with chest pain and recurrent syncope, found to be in haemodynamically unstable ventricular tachycardia (VT), requiring urgent cardioversion with successful conversion to normal sinus rhythm. Urgent coronary angiogram did not show any evidence of obstructive coronary artery disease. Transthoracic echocardiography notable for moderately reduced left ventricular systolic function (left ventricular ejection fraction 35–40%) with apical akinesis and an aneurysm of the apical septum
Day 2	Cardiac magnetic resonance (CMR) confirmed a prominent apical aneurysm with dyskinesis of the apical septum. There was transmural myocardial late gadolinium enhancement (LGE) of the entire left ventricular apex, extending into the mid anterolateral wall, indicating extensive fibrosis. Additionally, a small apical thrombus was visualized.
Day 3	Specific serological testing was positive for *Trypanosoma cruzi* IgG antibody. Given his recurrent syncope and clinical VT, the patient subsequently underwent an electrophysiology study which confirmed an apical septal VT origin corresponding to the region of fibrosis seen on CMR. VT mapping and ablation were subsequently done via an epicardial and endocardial approach
Day 6	Patient underwent dual-chamber implantable cardioverter defibrillator (ICD) for secondary prevention
Day 8	Patient discharged home
March 2022 (4 years later)	Admitted for recurrent VT and underwent successful repeat VT ablation
June 2022	Most recent follow-up with heart failure, transplant, and electrophysiology clinic; clinically doing well

Case presentation

A 54-year-old man, originally from Mexico, presented to the emergency department with chest pain and recurrent syncopal episodes. On arrival, he was found to be in haemodynamically unstable ventricular tachycardia (VT), with pulse of 202 b.p.m, requiring emergent cardioversion (*Figure 1*). He had altered mental status and vital signs were notable for blood pressure of 86/53 mmHg, respiratory rate of 20, SpO_2 of 98%, and temperature of 97.8° F (36.6° C). Presenting labs were notable for a peak troponin-I of 3.75 ng/mL (reference range 0.00–0.09 ng/mL). Electrolytes were within normal limits, but creatinine was elevated at 1.53 mg/dL (baseline 0.8–0.9 mg/dL). After restoration of sinus rhythm (*Figure 2*), he underwent urgent coronary angiography, which showed angiographically normal coronary arteries (*Figure 3A–D*, see Supplementary material online, *Video S1A–C*). A TTE revealed moderately reduced left ventricular systolic function [left ventricular ejection fraction (LVEF) 35–40%] with apical akinesis and an aneurysm of the apical septum (*Figure 4*). Cardiac magnetic resonance confirmed a prominent apical aneurysm with dyskinesis of the apical septum (see Supplementary material online, *Video S2*). There was transmural myocardial late gadolinium enhancement (LGE) of the entire left ventricular apex, extending into the mid anterolateral wall, indicating extensive fibrosis. Additionally, a small apical thrombus was visualized (*Figure 5*). Computed topographic brain did not show any evidence of prior or acute infarct concerning for thromboembolic event.

His home medications included aspirin 81 mg daily, hydrochlorothiazide 12.5 mg daily, and lisinopril 20 mg daily. He did not have any history of tobacco, alcohol, and drug use and was not aware of family history of cardiomyopathy or coronary artery disease (CAD).

On further investigation, it was learnt that he had emigrated from Mexico to the USA 20 years ago. He recalled living in a small mud hut in Mexico that was swarming with 'chinches', small nocturnal bugs that routinely bite the sleeping inhabitants.

Given the history and clinical findings, especially in light of the CMR findings and non-obstructive CAD, there was strong suspicion for Chagas heart disease as the explanation for his dilated cardiomyopathy (DCM). Specific serologic testing was positive for *T. cruzi* IgG antibody. Repeat ELISA testing at the Centers for Disease Control and Prevention testing was also positive, confirming the diagnosis of Chagas cardiomyopathy.

Given his recurrent syncope and clinical VT, the patient underwent an electrophysiology study which confirmed an apical septal VT origin corresponding to the region of fibrosis seen on CMR. He underwent VT mapping and ablation via an epicardial and endocardial approach (see Supplementary material online, *Figure S6*). A large area of low voltage corresponding with the apical aneurysm was noted on mapping. The VT was easily induced and upon studying was deemed to be likely intra-septal and micro-reentrant and was successfully ablated on the septal edge of the scar and healthy left ventricular (LV) myocardium. He then underwent loading with oral Sotalol and a dual-chamber defibrillator was placed for secondary prevention of SCD. The patient was discharged on guideline-directed medical therapy

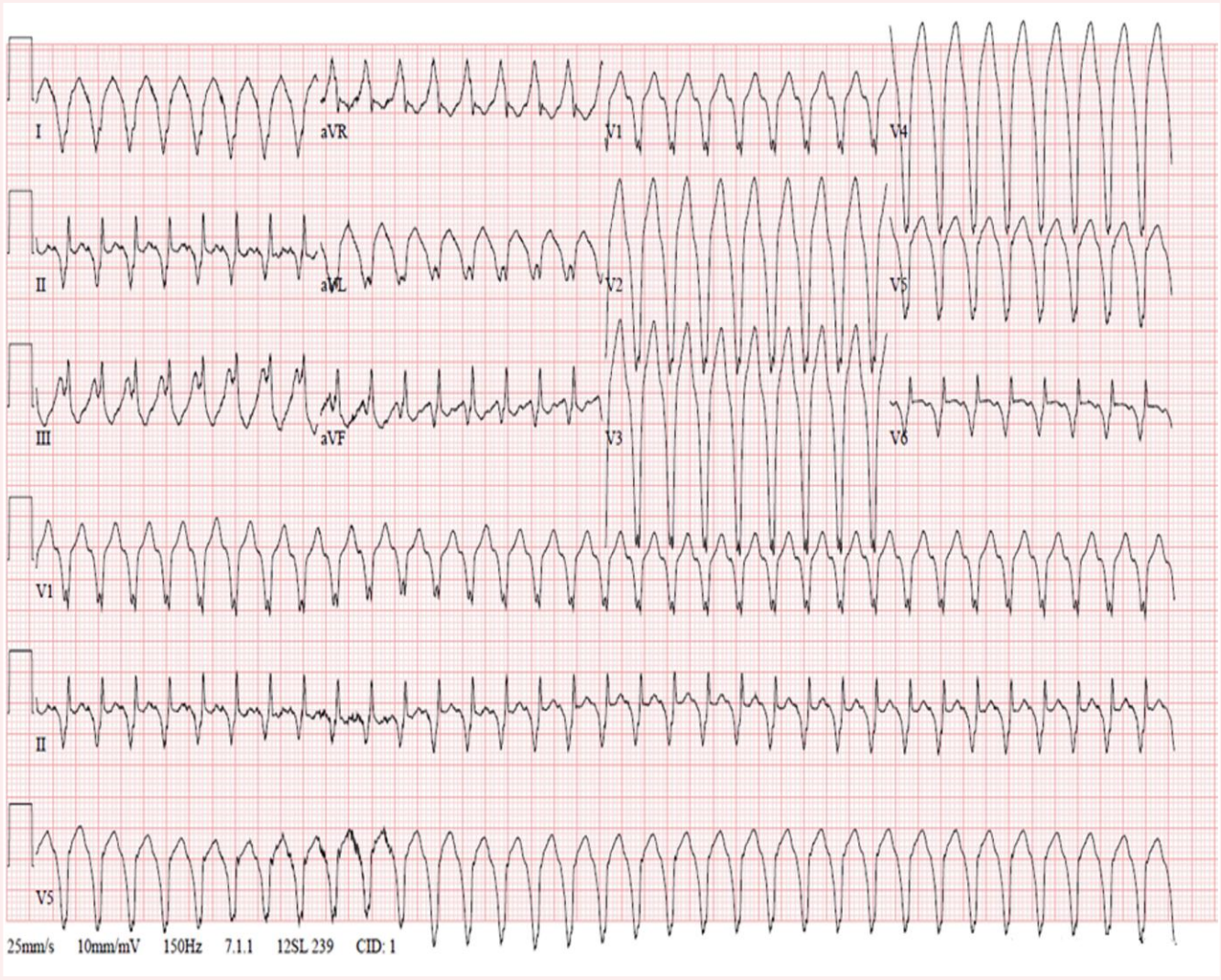

Figure 1 Electrocardiogram (EKG). Ventricular tachycardia with left bundle branch morphology, right inferior axis, possible mid-septal exit site.

(GDMT) including a regimen of anticoagulation, beta-blocker, and angiotensin-converting enzyme inhibitor (ACEI) therapies. Due to recurrence of VT, a repeat ablation was performed 4 years later and he has remained stable on GDMT. As his LVEF remains depressed (25–30%), he is followed closely by the advanced HF and transplant service. He remains clinically stable (New York Heart Association I/II) and is followed with serial TTEs. He was most recently seen in the HF and electrophysiology clinic in June 2022, and noted to be doing well on sacubitril–valsartan 49–51 mg twice daily, metoprolol XL 25 mg daily, and anticoagulation for his apical aneurysm and history of left ventricular thrombus.

Discussion

Trypanosoma cruzi is the protozoan responsible for CD, with transmission occurring through a bite from the triatomine insect, commonly known as the kissing bug. Other methods of transmission include oral intake of contaminated food or drink exposed to secretions from infected mammals or triatomine faeces, vertical transmission from mother to child during pregnancy, blood transfusion, organ transplantation, and unintentional exposure in the laboratory.[6]

There are two phases of CD: acute and chronic. The acute phase period lasts 8–12 weeks after exposure and is commonly undiagnosed as patients are often asymptomatic or have non-specific symptoms of fever, malaise, or hepatomegaly.[1] The chronic phase is divided into indeterminate and determinate phases. Most patients remain asymptomatic and progress into the chronic indeterminate form, which is characterized by positive anti-*T. cruzi* serology and normal electrocardiogram (ECG). Patients may remain asymptomatic for decades in the indeterminate chronic phase before progressing to the determinate phase, which is characterized by the development of cardiomyopathy or digestive disease.[6,7] Our patient was diagnosed at the time of determinate chronic phase with VT and HF. The development of chronic Chagas cardiomyopathy is associated with high morbidity given its association with HF, arrhythmia, and SCD [1,8] The most common cause of death in Chagas cardiomyopathy is SCD (55–60%) followed by HF (25–30%).[1,4]

Both atrial and ventricular arrhythmias can be seen in Chagas heart disease. Typical arrhythmia findings include atrial fibrillation, ventricular arrhythmia arising from the necrotic and fibrotic myocardial lesions, and bradyarrhythmia with conduction abnormalities including development of new right bundle branch block (RBBB) and heart block. Symptoms of palpitations should be further investigated. SCD could

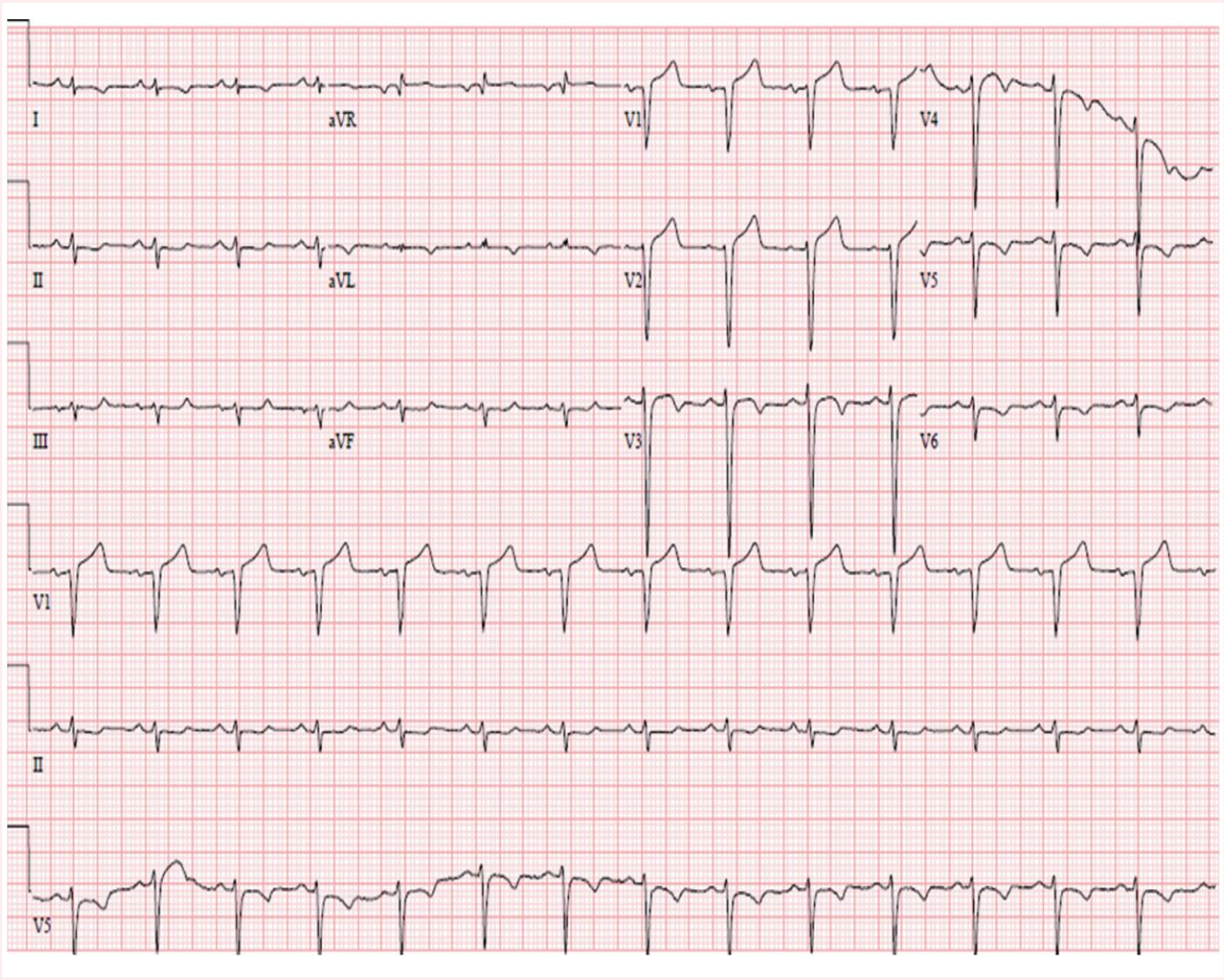

Figure 2 Electrocardiogram. Normal sinus rhythm after cardioversion; of note, no baseline EKGs were available for comparison in our electronic healthcare system.

occur even before the development of HF or other cardiac symptoms.[1] Amiodarone is often used for ventricular arrhythmia; however, it is associated with high toxicity including dermatitis, pulmonary fibrosis, and thyroid dysfunction among many others.[1] Sotalol was likely chosen for our patient given his young age to avoid risk of long-term amiodarone toxicity.

Progressive DCM is seen in Chagas HF, and regional wall motion abnormalities often precede the worsening of LV function. Multimodality imaging including CMR and TTE have become essential in detecting myocardial involvement.[1,9,10] Typical echocardiographic findings in Chagas cardiomyopathy include regional wall motion abnormalities in the inferior-inferolateral wall, LV apex with preserved septal contraction, LV aneurysm, LV diastolic dysfunction, DCM, LV apical thrombus, RV dysfunction, and functional mitral or tricuspid regurgitation [1,11] CMR with LGE are helpful in identifying the pattern of myocardial fibrosis and scar. Ischaemic heart disease is typically associated with a subendocardial or transmural pattern, whereas DCM is typically associated with mid-wall enhancement. Per the European Society of Cardiology guidelines, CMR is a Class I recommendation for further characterization of myocardial tissue in suspected infiltrative disease, CD, inflammatory disease, LV non-compaction, and haemochromatosis.[10]

Thromboembolic event is also commonly associated with CD and is a major cause of stroke in Latin America.[1]

Early diagnosis and recognition of CD is critical, allowing timely initiation of treatment, which may decrease the risk of cardiomyopathy, vertical transmission, and death.[1,8,10] For acute infection, PCR testing is the most sensitive test, but for chronic CD, there is no gold standard test. Typically, at least two different serological tests (e.g. indirect immunofluorescence and ELISA) for detecting *T. cruzi* antibodies are needed to confirm infection.

In patients with DCM, especially of Latin American background and without other risk factors, CD should be strongly suspected. In confirmed CD, ECG and echocardiography should be obtained at time of diagnosis. If normal, ECG can be obtained annually and echocardiography can be obtained every 3–5 years. If abnormal, especially with the presence of RBBB and conduction abnormalities, ECG should be obtained biannually. Electrocardiogram should also be obtained with any new symptoms of palpitations, syncope, or any arrhythmia. CMR is indicated to evaluate the biventricular function, presence of any LV thrombus, and the extent of fibrosis at the time of diagnosis.[1]

Medical therapy for Chagas cardiomyopathy is extrapolated from studies done on other forms of HF and includes the combination of

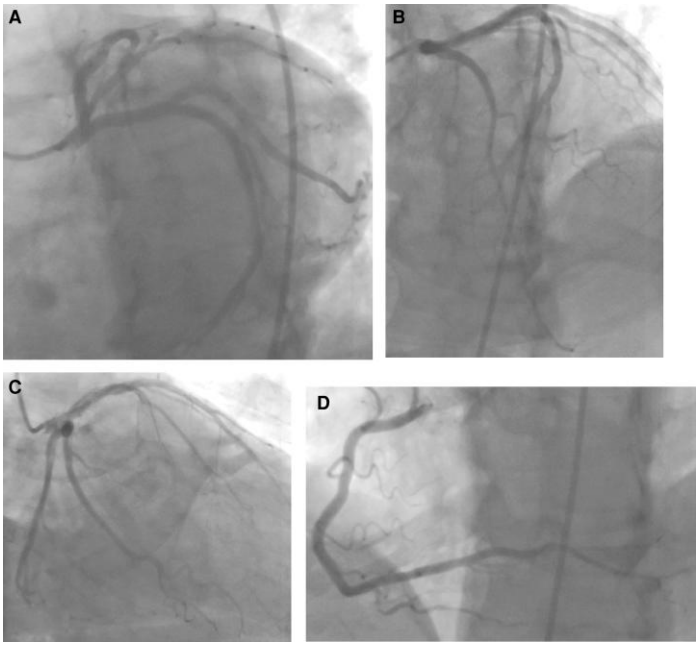

Figure 3 (*A–D*)Coronary angiogram showing angiographically normal coronaries without evidence of obstructive coronary artery disease.

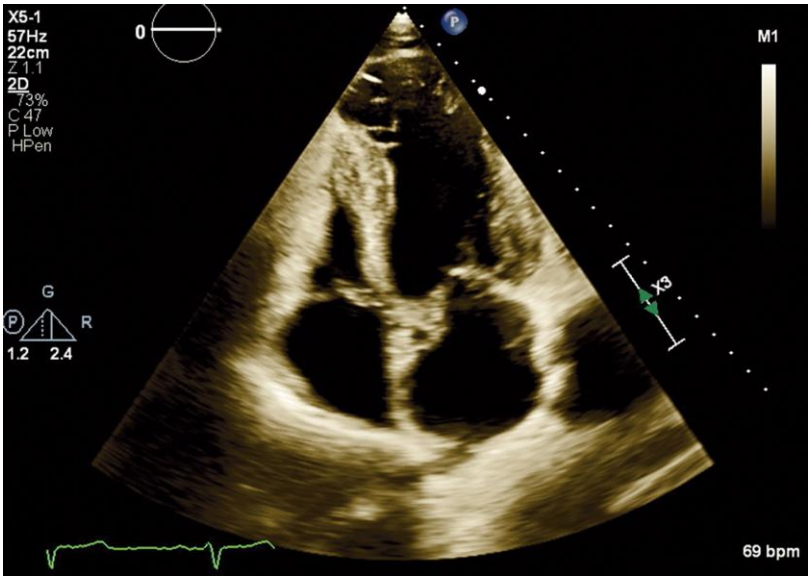

Figure 4 Transthoracic echocardiogram, apical four-chamber view showing a dilated left ventricle with moderately reduced systolic function (ejection fraction 35–40%) and an apical aneurysm. Additional notable findings include markedly dilated left atrium (indexed volume of 51 mL/m^2), left ventricular lateral e′ of 5.66 cm/s and septal e′ of 4.9 cm/s with *E*/e′ of 18. Estimated pulmonary artery systolic pressure of 43 mmHg.

beta-blockers, ACEI/angiotensin receptor blockers/neprilysin inhibitor, and aldosterone receptor antagonists. Diuretics help improve the quality of life in patients with congestive symptoms but has not shown any mortality benefits.

As the disease burden and prevalence of CD increase worldwide, it is imperative for healthcare providers to recognize the manifestations of CD and make timely diagnosis and treatment for the patients. This case highlights the critical role of careful interrogation of demographics and advanced cardiac imaging for the successful diagnosis and treatment of Chagas heart disease. However, limitations in the management of this patient include the lack of 3D-TTE, strain imaging, and T1–T2 mapping on CMR at the time of investigation in 2018.

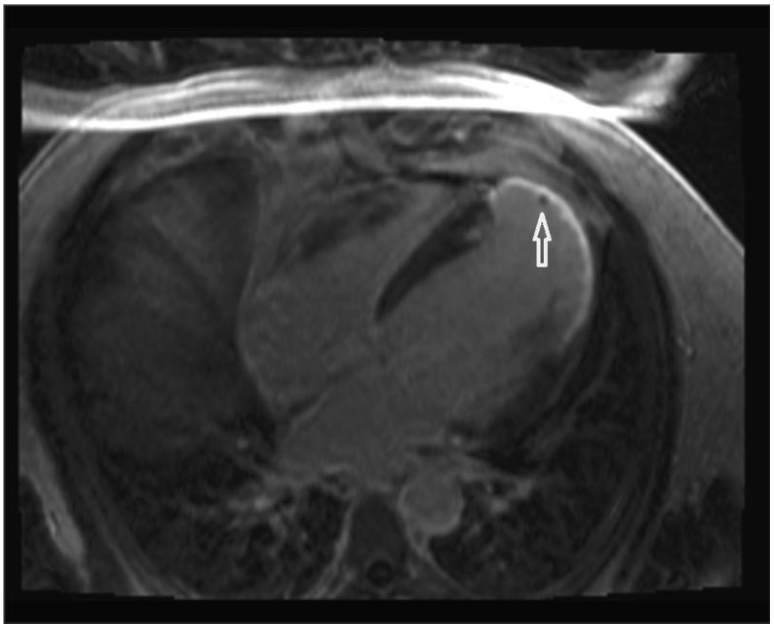

Figure 5 Cardiac magnetic resonance imaging, four-chamber late gadolinium enhancement: rounded density measuring 4 × 5 mm in the left ventricular apex appears to be a small apical thrombus (see arrow). Additional findings notable for severely dilated left ventricular cavity (indexed left ventricular end-diastolic volume of 176 mL/m^2) with moderately to severely reduced systolic function of 35%, normal left ventricular mass index of 78 g/m^2, normal right ventricular end-diastolic volume of 175 mL, and end-systolic volume of 63 mL with the ejection fraction of 63%.

Lead author biography

Dr Shannon Li completed her internal medicine training at the University of Wisconsin and currently undergoing cardiology training at the Rush University Medical Center.

Supplementary material

Supplementary material is available at *European Heart Journal – Case Reports*.

Acknowledgement

None.

Slide sets: A fully edited slide set detailing this case and suitable for local presentation is available online as Supplementary data.

Consent: The authors confirm that written consent for the submission and publication of this case, including images, has been obtained from the patient in line with COPE guidance.

Conflict of interest: P.S.S.: Honoraria: Medtronic. Consulting: St Jude Medical. All other authors none declared.

Funding: None declared.

Data availability

The authors confirm that the data used in this study are available within the article and its Supplementary Material section.

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