

Saw tooth cardiomyopathy: a case report

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Abstract

Saw tooth cardiomyopathy is an unusual and rare type of left ventricular dysplasia that is characterized by multiple projections of compacted myocardium that makes the appearance of ‘saw tooth’ in noninvasive imaging. We present a young man with signs and symptoms of heart failure and reduced left ventricular function in echocardiography who showed distinctive left ventricle features of saw tooth cardiomyopathy (saw tooth appearance of myocardium in basal inferolateral and basal to mid lateral segments) in cardiac magnetic resonance imaging.

Keywords cardiomyopathy, saw tooth, left ventricular non compaction

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Introduction

A few cases of saw tooth cardiomyopathy have been reported in the literature. This type of left ventricular dysplasia has considered as a variant of left ventricular (LV) noncompaction (NCLV) by some authors. We present the case of a 32-year-old male patient, referred to our department due to heart failure sign and symptoms.

Case presentation

The present case is a 32-year-old man with history of dyspnoea on exertion for 5 years. He was referred to our outpatient heart failure clinic for further evaluation after primary assessment. He had history of dyspnoea on exertion with a New York Heart Association functional class of II. His past medical and family history was unremarkable, and there was not any history of drug abuse. The electrocardiography showed no remarkable findings (Figure 1). Transthoracic echocardiography showed mild LV enlargement with an ejection fraction of 35%, mild LV diastolic dysfunction, normal right ventricle (RV) size and function, and trivial mitral regurgitation.

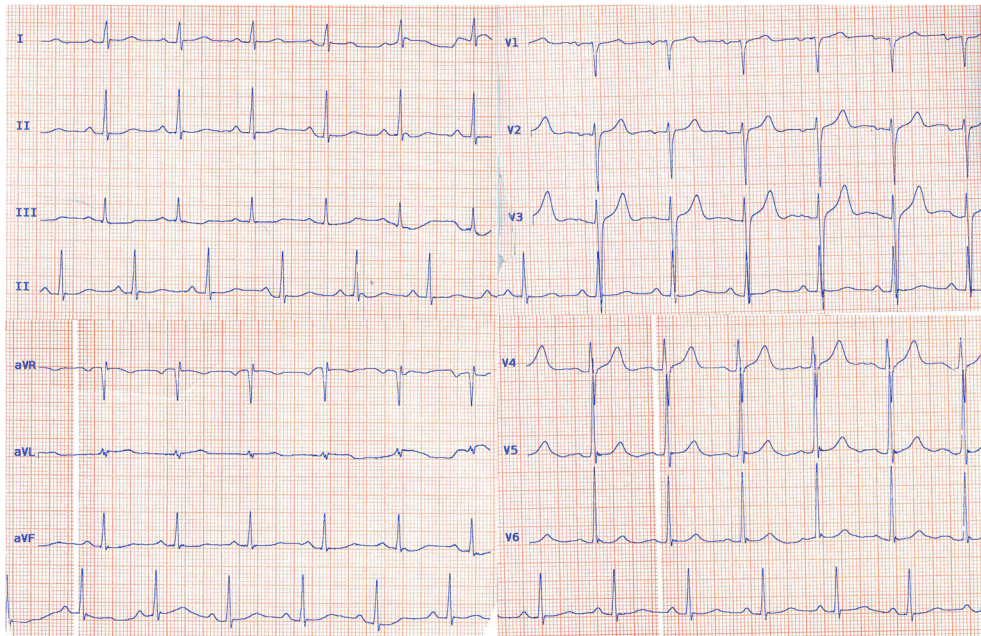
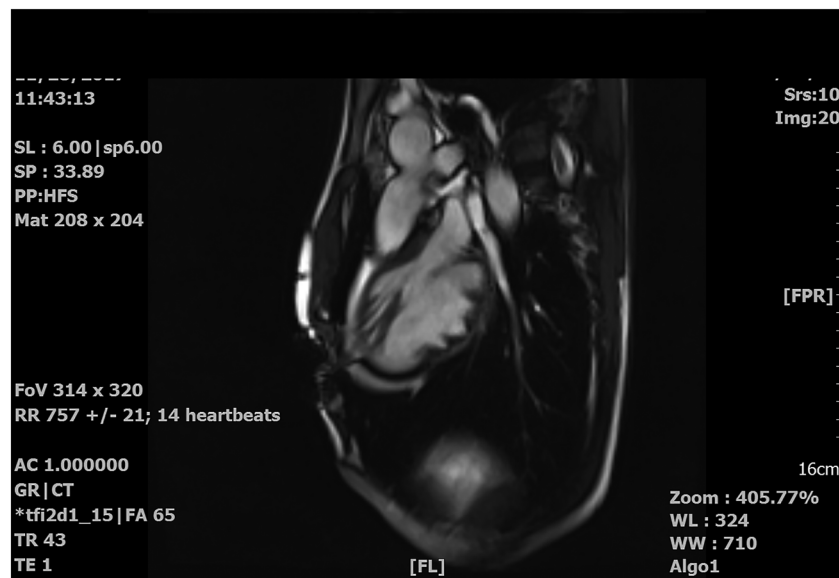
He had also a report of coronary computed tomography angiography that was unremarkable in terms of coronary artery disease except for a muscle bridge segment at the mid

portion of left anterior descending artery. Guideline-directed medical therapy for heart failure was started for him, and he was referred for cardiac magnetic resonance (CMR) examination for better assessment of cardiac morphology and function.

CMR examination was performed about 3 months after initiation of heart failure therapies on a 1.5 Tesla Avanto magnetic resonance imaging system (Siemens, Erlangen, Germany), and a 0.2 mmol/kg of gadolinium-diethylenetriamine penta-acetic acid was administered intravenously, and delayed enhancement short axis and long axis images were recorded 10–15 min later.

His CMR exam demonstrated normal LV size with end diastolic volume index of 92.5 ml/m², mild systolic dysfunction with LV ejection fraction of 45%. Normal thickness of apical segments with normal pattern of basal to apical tapering was also noted. Saw tooth appearance of myocardium in basal inferolateral and basal to mid lateral segments was also seen. (Figures 2, 3, and 4)

Right ventricular size was normal (right ventricular end diastolic volume indexed to body surface area measured 73 ml/m²). Mild right ventricle systolic dysfunction was evident by right ventricular ejection fraction of 48%. No evidence of myocardial delayed hyper enhancement was seen in delayed post contrast images suggesting no evidence of identifiable

Figure 1 Twelve leads electrocardiogram of patient**Figure 2** Long axis view of left ventricle, showing distinct projections of myocardium

replacement fibrosis in the myocardium. His dyspnoea was improved (New York Heart Association functional class of I) after heart failure therapies, and in his last clinic visit, the clinical condition was acceptable. To screen the presence of similar condition in his family, the first degree relatives of patient were advised to performed electrocardiography and transthoracic echocardiography which showed no abnormalities.

Discussion

Saw tooth cardiomyopathy is an unusual type of left ventricular dysplasia that is characterized by multiple projections of compacted myocardium¹ that makes the appearance of saw tooth in noninvasive imaging. Detection of this abnormality in an infant with heart failure

Figure 3 Short axis view of left ventricle

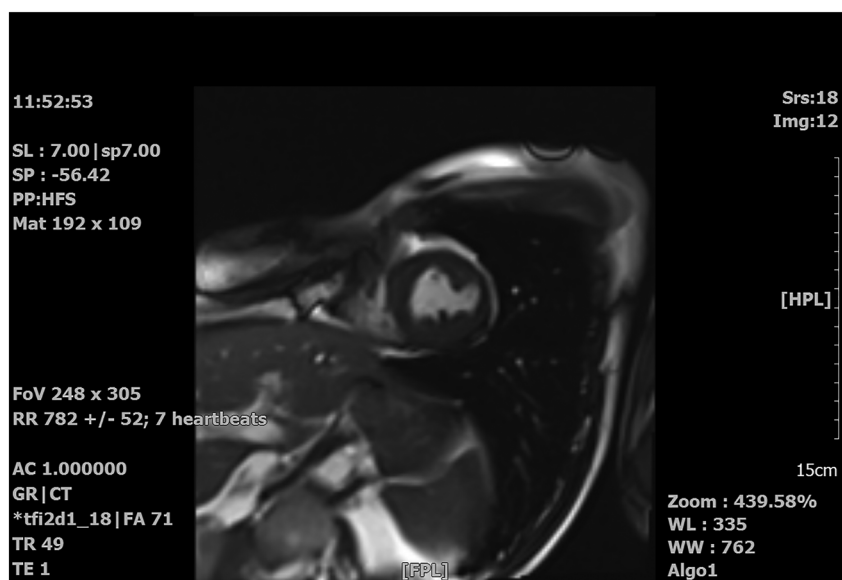
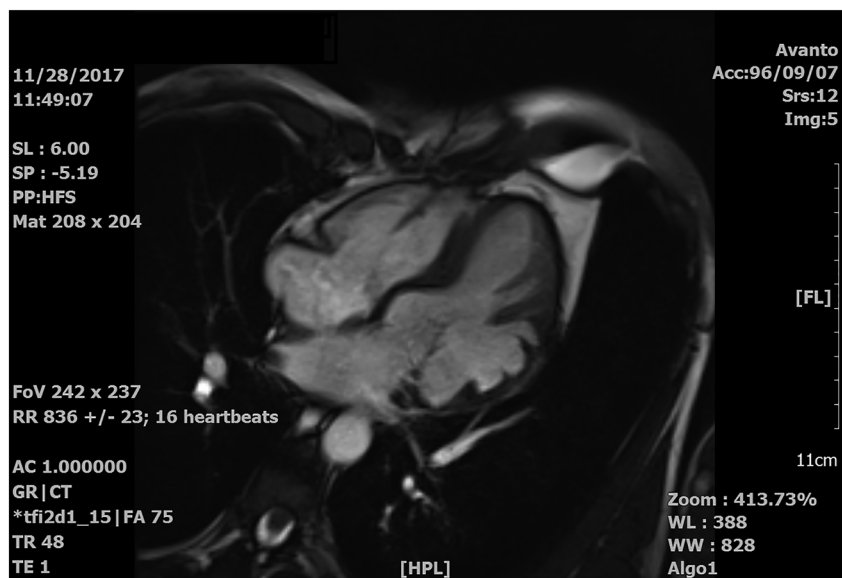


Figure 4 Four chamber view



symptoms² raises the possibility of genetic basis as well as aggressive form of this cardiomyopathy; however, similar imaging findings in an elderly patient may be indicative of more benign course of this disease or presumably an associated abnormality³, like NCLV. Only a few cases of this cardiomyopathy has been reported, and it could be an incidental finding or in the setting of heart failure symptoms.

Whereas there is a slight resemblance of this cardiomyopathy with excess trabeculation cardiomyopathy (NCLV), the differences between these two cardiomyopathies are remarkable. While in saw tooth cardiomyopathy, numerous projection of compacted myocardium is noticeable, and in NCLV, prominent trabeculated meshwork can be seen⁴. All of the reported cases were male participant that raises the possibility of X-linked disorder. Regarding few data about this

cardiomyopathy, further evaluation of these patients like genetic studies may be helpful.

Our patient has stable clinical condition, and his heart failure symptoms have completely improved after the initiation of standard heart failure therapies. Besides, CMR study showed a better LV function about 3 months after the onset of the treatment. Considering the improvement in clinical and imaging findings of patient, we can say that our patient may have a benign form of saw tooth cardiomyopathy. Anyhow, guideline directed medical therapy for heart failure will be continued for him, and he was scheduled to have an outpatient follow up visit each 3 months and/or whenever his signs or symptoms were aggravated

Article history

This case report is about a case with a diagnosis of saw tooth cardiomyopathy, which was approved to be reported by the

ethics committee of Rajaie Cardiovascular Medical and Research Center, and the patient gave the informed consent.

Each author has made an independent material contribution to the work, this paper has not been published or submitted in any other journal, and all authors have read and approved the manuscript to be published in ESC heart failure and have no disclosures to admit.

Conflict of interest

The authors disclose that this study has no any relationship with industry and financial associations, and no competing interest has been declared.

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