

IMAGES IN MEDICINE

Non-Compaction Cardiomyopathy: Echo and Angiographic Imaging

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ABSTRACT

A 35-year-old gentleman was referred to our clinic after two syncopal episodes, and ensuing work-up disclosed a trabeculated left ventricle with blood-filled myocardial recessions on color Doppler echocardiography and myocardial filling defects on left ventriculography, all leading to the diagnosis of noncompaction cardiomyopathy. Ventricular tachyarrhythmias were induced at an electrophysiology study and the patient received an implantable defibrillator.

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KEY WORDS: *noncompaction
cardiomyopathy; echocardiography;
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A 35-year-old construction worker was referred to our clinic due to two syncopal episodes over the last two weeks before admission. The patient was a heavy smoker and drinker. Physical examination including full neurological examination was normal. The electrocardiogram revealed left ventricular hypertrophy, upsloping saddle-shaped ST segment in leads V1 to V3 with negative T waves over all the precordial leads. An echocardiogram revealed trabeculations and deep recessions filling with blood in the apex and inferior wall and mild hypertrophy of the interventricular septum (Fig. 1). Left and right ventricular dimensions were normal; ejection fractions of both ventricles were also normal. Coronary angiography revealed normal coronary arteries; the right ventriculogram was normal, while the left ventriculogram showed numerous filling defects and a non-smooth contour (Fig. 2). At an electrophysiological study, performed to further investigate the cause of syncope, ventricular fibrillation was reproducibly

ABBREVIATIONS

ICD = implantable cardioverter
defibrillator

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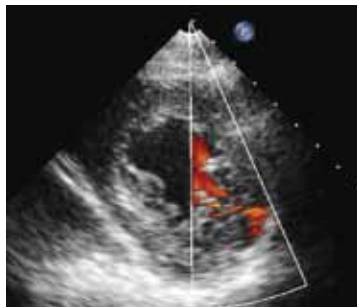


FIGURE 1.

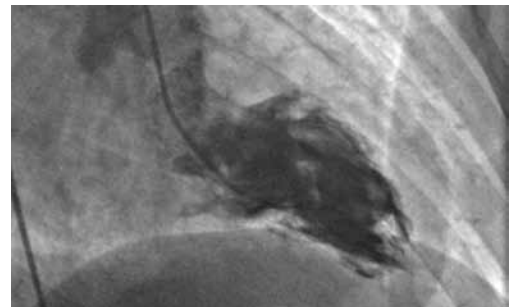


FIGURE 2.

Conflict of interest: none declared

inducible with 3 ventricular extrastimuli at the apex and the outflow tract of the right ventricle. The diagnosis of noncompaction cardiomyopathy was made and the patient received an implantable cardioverter defibrillator (ICD).



Echocardiography is the imaging modality of choice for the diagnosis of noncompaction cardiomyopathy, although cardiac magnetic resonance imaging is a new promising modality, and specific diagnostic criteria have been suggested for both modalities.¹⁻⁴ Patients with noncompaction cardiomyopathy present with the triad of heart failure symptoms, embolic episodes or arrhythmias.¹ Ventricular tachyarrhythmias have been reported in 47% of patients and sudden cardiac death in 50% of deaths in a large series of patients with noncompaction cardiomyopathy.⁵ An association with facial dysmorphisms and neuromuscular disorders has been described, as well as familial association. Prognosis may seem better than previously thought,⁶ albeit nonuniformly.^{7,8} In a recent analysis, survival or freedom from heart transplantation at 3 and 5 years were 85% and 75% respectively.⁶ However, in another study from a French Registry,⁸ among 105 patients with noncompaction cardiomyopathy, a rather high complication and mortality rate was recorded during an average follow-up of 2.33 years; 33 patients developed severe heart failure, 9 patients underwent cardiac transplantation, 7 patients had ventricular arrhythmias, 9 patients had embolic events, and 12 patients died. Our patient had syncope presumed to be due to ventricular arrhythmias (inducible at the electrophysiology study), for which he received an ICD, but has had no heart failure symptoms or

embolic events as yet. Diagnosis was confirmed by both echo and angiographic imaging (Fig. 1 & 2).

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