

Original Article

Left ventricular noncompaction cardiomyopathy: a case report and literature review

Ying-Nan Lin^{1*}, Yu-Qiang Wang^{1*}, Yi Yu^{2*}, Qing Cao¹, Fei Wang¹, Shu-Yan Chen¹

¹Department of Geriatrics, Xinhua Hospital Affiliated to Shanghai Jiao Tong University School of Medicine, Shanghai 200092, China; ²Departments of Ultrasonography, Xinhua Hospital Affiliated to Shanghai Jiao Tong University School of Medicine, Shanghai 200092, China. *Equal contributors.

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Abstract: Left ventricular noncompaction cardiomyopathy (LVNC) is a relatively rare congenital disorder prominently characterized by prominent trabeculations and intertrabecular recesses that communicate with the ventricular cavity rather than the coronary circulation. LVNC can occur in isolation or coexist with other cardiac and/or systemic anomalies, in especial neuromuscular disorders. The clinical presentation varies ranging from asymptomatic patients to patients who develop ventricular arrhythmias, thromboembolism, heart failure and sudden cardiac death. Although LVNC is commonly diagnosed by echocardiography, there are also other useful diagnostic techniques, including contrast ventriculography, CT and MRI. Now, it is being diagnosed more frequently in patients of all ages because of increased awareness and improvements in imaging methods. We described the case of a woman who presented with heart failure for the first time at 62 years of age. The diagnosis was LVNC. Transthoracic echocardiography showed a trabeculated, sponge-like appearance of the ventricular apical and inferolateral segments. After medical management, the patient was asymptomatic at the 1-month follow-up examination. Now we discussed the diagnosis of this case and reviewed the medical literature that pertained to LVNC.

Keywords: Trabeculations, left ventricular noncompaction cardiomyopathy, congestive heart failure

Introduction

LVNC is a relatively rare congenital disorder prominently characterized by prominent trabeculations and intertrabecular recesses that communicate with the ventricular cavity rather than the coronary circulation [1, 2]. It is thought to be caused by intrauterine arrest of compaction of the myocardial fibres and meshwork-an important process in myocardial development [3], although there is some controversy as to whether the condition can also be acquired [4, 5]. The American Heart Association classified it as a primary genetic cardiomyopathy. In a recent analysis [6], it was determined that 18% to 42% of left ventricular noncompaction cases were familial. However, the phenotype of patients with noncompaction can vary even within familial cases and range from clinically benign to fatal [7]. Genetic and imaging studies are justified in first-degree relatives of affected individuals, to identify potential risks and the

possibly causative genes [5, 6]. The clinical presentations varied ranging from asymptomatic patients to patients who develop ventricular arrhythmias, thromboembolism, heart failure, and sudden cardiac death [8]. They are being recognized more often in the differential diagnosis of patients with heart failure.

Here, we described the case of a woman who presented with heart failure for the first time at 62 years of age. We discussed the diagnosis of this case and reviewed the medical literature that pertained to LVNC.

Case report

In February 2014, a 62-year-old woman was admitted to our hospital because of congestive heart failure during the previous twelve days. She had no chest pain, palpitations, or syncope. Physical examination revealed bilateral crackles in the lower-lung fields and edema of lower extremity. Neurologic findings were unre-

Left ventricular noncompaction cardiomyopathy

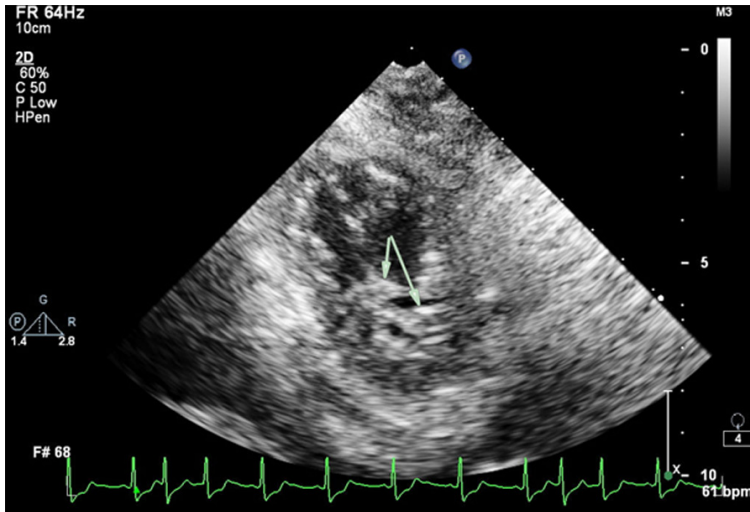


Figure 1. Left ventricular with more than 3 trabeculations protruding from the posterolateral wall (arrows) visualized by two-dimensional echocardiography.

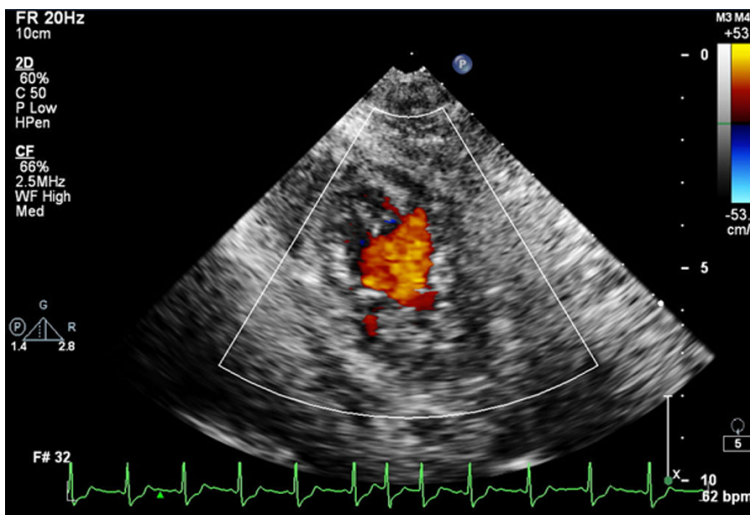


Figure 2. Left ventricular trabeculations and blood flow into the intertrabecular recesses (parasternal short-axis view) visualized by Color Doppler flow imaging.

markable. An electrocardiogram showed ST-T changes. The results of routine biochemical tests were within normal limits. The trabeculae were located on the posterolateral wall of the left ventricle (Figure 1). The blood flow into the intertrabecular recesses could be visualized by color Doppler (Figure 2). The ratio of non-compacted myocardium to compact myocardium at the end of systole was $> 2:1$ (Figure 3). The patient also had mild systolic dysfunction and an ejection fraction of 0.33 (Figure 4). Moderate mitral and tricuspid regurgitation and pulmo-

nary hypertension were noted (peak systolic pulmonary artery pressure, 65 mmHg). Morphologically, the heart valves were normal, and no coexisting congenital anomaly was found. These findings led to the diagnosis of LVNC. The patient had no significant arrhythmias during her hospital stay. After medical management with β -blockers, ACEI, a loop diuretic, spiro lactone and digoxin, she was asymptomatic at the 1-month follow-up examination.

Discussion

We have described the diagnosis of LVNC in a 62-year-old woman. The patient had been asymptomatic until mild symptoms of congestive heart failure developed during twelve days before initial presentation. She responded well to medical management of this relatively rare cause of heart failure.

Tools for the diagnosis of LVNC are echocardiography, contrast ventriculography, computed tomography (CT) and magnetic resonance imaging (MRI) [9]. Although Petersen and colleagues [10] reported that CMR showed the extent of involvement and the exact location of noncompacted segments, echocardiography was crucial. Widely accepted diagnostic criteria by Jenni et al. [11, 12] were as follows: (1) Absence of coexisting cardiac abnormalities. (2) Segmental thickening of myocardial wall of left ventricle with two layers: a thin epicardial layer and a thick endocardial layer with prominent trabeculations and deep recesses. The ratio of non-compacted myocardium to compact myocardium at the end of systole is $> 2:1$. (3) The trabeculae are usually located on the apical/lateral, middle/bottom walls of the left ventricle. Most non-compacted segments are hypoki-

Left ventricular noncompaction cardiomyopathy

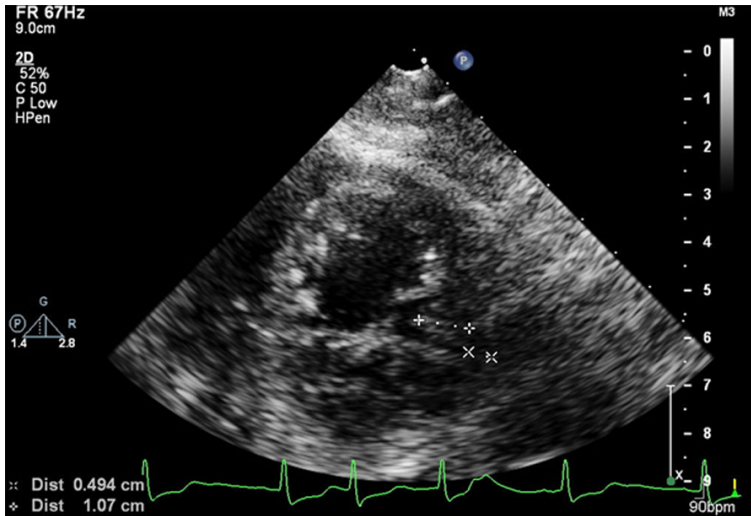


Figure 3. The ratio between noncompacted and compacted wall was more than 2 in the diastole.

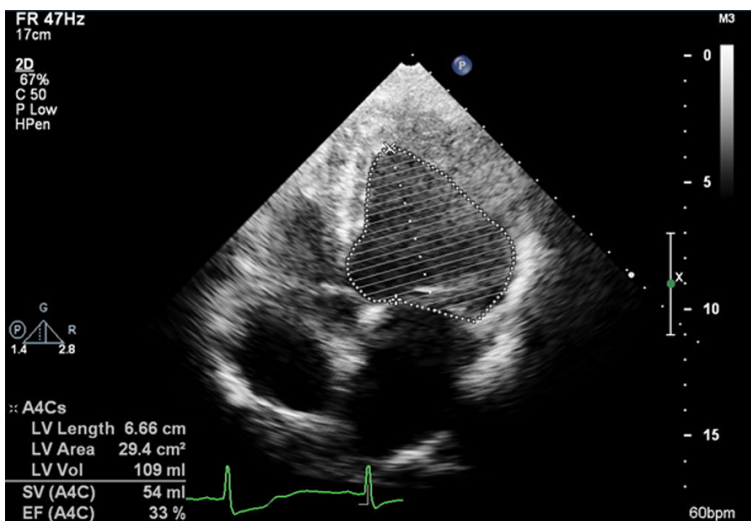


Figure 4. The result of left ventricular ejection fraction measured by a bi-plane method indicated that the systolic function was impaired.

netic. (4) The flow between the intertrabecular recesses can be identified by using the color Doppler method.

In patients with LVNC, treatment options vary on an individual basis, ranging from medical management in mild cases to heart transplantation in patients with refractory symptoms [7]. Besides treatment for congestive heart failure and antiarrhythmic therapy, if indicated, anticoagulation therapy is recommended because of the risk of systemic embolism [2]. Some guidelines, including the Brazilian one [13], recommend anticoagulation to patients with de-

creased systolic function with ejection fraction below 40%, history of thromboembolism or atrial fibrillation [14]. Monitoring asymptomatic patients is encouraged, whether diagnosis is incidental or the consequence of familial screening, because of possible future complication in those individuals [6, 15]. The prognosis of patients with LVNC is determined by the degree and progression of heart failure, presence of thromboembolic events and arrhythmias [9]. Recently, Murphy et al. [14] noted improvement in prognosis compared to what was reported in previous studies. For a mean period of 46 months, one death was found in a cohort of 45 patients with ventricular noncompaction, who were clinically monitored at regular intervals every six months in a specialized center. Our patient's mild symptoms responded well to medical therapy, and her short-term prognosis was excellent.

Although LVNC was a relatively rare cause of heart failure in patients, our case suggested that it should be included in the differential diagnosis in patients who presented with first-time symptoms. In recent years, LVNC is a disease that

has been increasingly diagnosed in clinical practice mainly due to improvements in echocardiographic techniques and use of cardiac resonance, suggesting that hypertrabeculation also occurs in other comorbidities and that it may be falsely diagnosed as LVNC. Therefore, we suggested that the diagnosis of suspected myocardial noncompaction should be carefully evaluated by imaging methods to avoid inappropriate and exaggerated diagnoses.

Disclosure of conflict of interest

None.

Left ventricular noncompaction cardiomyopathy

Address correspondence to: Dr. Shu-Yan Chen, Department of Geriatrics, Xinhua Hospital Affiliated to Shanghai Jiao Tong University School of Medicine, 1665 Kongjiang Road, Shanghai 200092, China. Tel: +862125078999; E-mail: shuyanchencn@hotmail.com

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