Biventricular Noncompaction Associated With Left Ventricular Systolic and Diastolic Dysfunction and Severe Pulmonary Hypertension in a Young Man

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Noncompaction of the ventricular myocardium is a recently recognized genetic cardiomyopathy. The left ventricle is the most affected site, but right ventricular involvement has been reported in some cases. Diagnosis is made with 2-dimensional echocardiography or cardiac magnetic resonance imaging. The major clinical manifestations are heart failure, arrhythmias and embolic events. A 20-year old man had left and right ventricular noncompaction complicated by severe pulmonary hypertension, which is one of the first cases of biventricular noncompaction associated with severe pulmonary hypertension. Pulmonary hypertension may be a consequence of increased pulmonary venous pressures caused by systolic and diastolic heart dysfunction secondary to noncompaction. (Circ J 2009; 73: 2163–2165)

Key Words: Echocardiography; Magnetic resonance imaging; Ventricular noncompaction

Noncompaction of the ventricular myocardium is a recently recognized genetic cardiomyopathy characterized by a distinctive (“spongy”) morphological appearance of the left ventricle.1 Prominent trabeculations are a normal feature of the developing myocardium in utero and left ventricular (LV) noncompaction is thought to result from a failure of the trabecular regression that occurs during normal embryonic development.2 The diagnosis of ventricular noncompaction is usually made using echocardiography and cardiac magnetic resonance imaging (MRI).1,3 We present a patient with myocardial noncompaction of both ventricles and severe pulmonary hypertension.

Case Report
A 20-year-old man with chronic heart failure and paroxysmal atrial fibrillation was referred for diagnostic assessment and therapy. Atrial fibrillation and progressively worsening effort dyspnea had started 3 years ago. There was no family history of heart disease or sudden death. On admission, his blood pressure was 100/65 mmHg and the pulse rate was 76 beats/min. The 12-lead ECG showed sinus rhythm, pathological right-axis deviation, P-mitrale and pulmonale, and LV and right ventricular (RV) hypertrophy. Chronic interstitial lung congestion was found on chest X-ray. Transthoracic echocardiography revealed a normally sized, hypokinetic LV (ejection fraction 35%) accompanied by massive enlargement of the right and left atria. The apical portions of both ventricles had a markedly trabeculated, spongy appearance, indicating biventricular noncompaction (Figures 1A–C). The maximum end-systolic ratio of the noncompacted endocardial layer to the compacted myocardium for the left and right ventricles was >2. Doppler examination showed color flow between the prominent trabeculations (Figure 1D). Moreover, there was severe tricuspid regurgitation. The calculated peak systolic pulmonary artery pressure (140 mmHg) revealed severe pulmonary hypertension. The restrictive left ventricle inflow and decreased myocardial velocities (Figures 2A, B) suggested significantly elevated left ventricular end-diastolic pressure. MRI showed a typical pattern of an apical inner zone of noncompacted myocardium distinguished from the thin outer zone of compacted myocardium (Figures 3A, B). Right heart catheterization showed elevated pulmonary artery pressures (99/43/52 mmHg), and elevated mean pulmonary capillary wedge and right atrial pressures (24 and 16 mmHg, respectively), and a cardiac index of 1.51 L·min⁻¹·m⁻². Coronary angiography was performed and revealed normal coronary arteries. Left ventriculography showed a honeycomb-like appearance in the anterior, apical and inferior wall segments, and global hypokinesia. The LV ejection fraction was 25% and the LV and diastolic pressure was 27 mmHg. During hospitalization, venous thromboembolic disease, lung diseases, congenital heart diseases and other rare conditions as a potential cause...
Figure 1. Transthoracic echocardiography. (A) Apical 4-chamber view showing huge bi-atrial dilatation, normally sized left ventricle, moderately enlarged right ventricle and noncompaction of the left ventricle. (B) Modified apical 4-chamber view showing noncompaction of the left ventricle. (C) Modified apical 4-chamber view showing noncompaction of the right ventricle. (D) Apical 4-chamber view with color Doppler flow mapping between the intertrabecular spaces. LA, left atrial; LV, left ventricular; RA, right atrial; RV, right ventricular.

Figure 2. (A) Transmitral echo Doppler profile shows E/A = 2.9, short deceleration time = 140 ms and short left ventricular isovolumetric relaxation time = 60 ms, suggesting restrictive physiology. (B) Pulsed tissue Doppler of lateral mitral annulus-E/E' = 18.

Figure 3. Cardiac magnetic resonance imaging was performed using a 32-channel MR system (Siemens Avanto SQ-Class Tim [76×32] with 32-channel cardiac coil (InVivo)). True FISP sequence was used for imaging. Double-layered appearance, marked trabeculations and deep intratrabecular recesses of the LV and RV myocardium on the 4-chamber (A) and short-axis (B) views (arrows). LA, left atrial; LV, left ventricular; PE, pericardial effusion; RA, right atrial; RV, right ventricular.
of pulmonary hypertension were excluded. The patient was considered for heart and lung transplantation, but he refused.

Discussion

Noncompaction of the LV myocardium is a rather rare condition with a prevalence in adults of <0.3%, but recently many cases of noncompaction have been reported.2,4,5 The left ventricle is the most affected site of noncompaction but RV involvement only has been reported in some cases.6,7 The echocardiographic characteristics of ventricular noncompaction include, in the absence of any coexisting lesions, segmental thickening of the LV myocardial wall consisting of 2 layers: a thin, compacted epicardial layer and an extremely thick layer with prominent trabeculations and deep recesses. A maximum end-systolic ratio of the noncompacted endocardial layer to the compacted myocardium of >2 is characteristic. Color Doppler echocardiography usually reveals deeply perfused intertrabecular recesses. Predominant segmental location of the abnormality is almost always found in the apical and mid-ventricular areas of both the inferior and lateral walls.8,9 MRI is also being used to detect ventricular noncompaction and has the advantage of good spatial resolution at the apex and lateral wall of the left ventricle.10 In the current literature we found no special definition of RV noncompaction, but some authors suggest that the definition for the left ventricle can be applied.10 In the present patient, the diagnosis of left ventricle noncompaction is undoubtedly; however, visualization of the right ventricle is more difficult because of its irregular shape. Therefore, diagnosis of RV noncompaction seems to be more difficult. An important differential diagnostic consideration is the presence of prominent trabeculations as a common variant of normal hearts, but these, however, most often course from the free wall to the ventricular septum.9 In the patient reported here, the ventricular septum was almost normal and the noncompaction affected the free wall and the apex of the right ventricle. The major clinical manifestations of ventricular noncompaction are heart failure, arrhythmias and embolic events.11 To the best of our knowledge, this is one of the first reports of biventricular noncompaction associated with severe pulmonary hypertension. Pulmonary hypertension may be a consequence of increased pulmonary venous pressures caused by systolic and diastolic heart dysfunction secondary to the noncompaction.

References