Biventricular Noncompaction and Bilateral Outflow Obstruction

Echocardiographic and Computed Tomography Imaging Features

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A 21-year-old male patient was admitted to our hospital with acute dyspnea and worsening of his general condition. His medical history was remarkable for a known noncompaction of the left (LV) and right ventricle (RV) that was diagnosed at the age of 6.

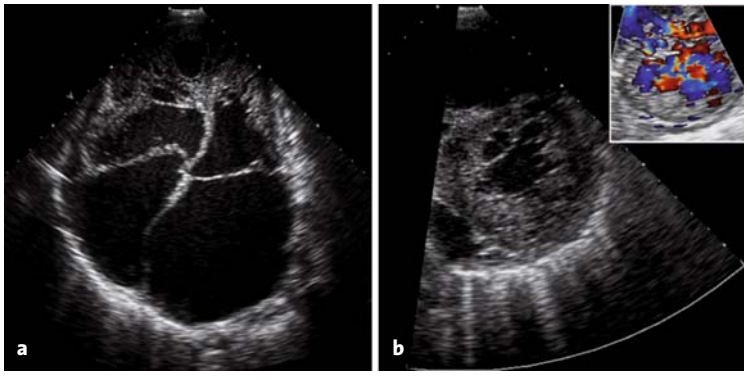
Transthoracic echocardiography (TTE) was performed (Acuson, Sequoia C512, Siemens) to assess LV function and to confirm biventricular noncompaction. TTE showed an enlargement of both atria, as well as the RV. The LV was normal in size but showed hypokinesia of the septum and posterior wall. Furthermore, hypoplasia of the aortic root with a normal tricuspid aortic valve was seen. The LV ejection fraction was 48% according to Simpson's biplane method. The structure of the ventricular walls revealed a compact epicardial layer and an endocardial layer consisting of a trabecular meshwork and deep intertrabecular spaces with a ratio of end-systolic noncompact/compact layer > 2 for both the LV and RV (Figures 1a and 1b). Color Doppler revealed flow between the ventricular cavities and the intertrabecular spaces (see Figure 1b). In addition, the RV systolic pressure was markedly raised (estimated by continuous-wave Doppler at 98 mmHg, while the patient's blood pressure was recorded at 100/75 mmHg).

Because of the increased pulmonary artery pressure, a contrast-enhanced chest computed tomography (CT) was performed to rule out pulmonary embolism. In addition, the CT examination was intended to search for coexisting congenital cardiac anomalies. Therefore, CT was performed using a triple rule-out protocol with prospective electrocardiography (ECG) gating (dual-source CT, Somatom Definition, Siemens). Acute pulmonary embolism was ruled out, and no signs of chronic pulmonary embolism were seen, while enlarged pulmonary arteries indicated the presence of pulmonary arterial hypertension. CT similarly confirmed the ventricular noncompaction predominating in the mid- and apical portions of both ventricles (Figures 2a and 2b). Associated congenital cardiac anomalies other than a hypoplastic thoracic aorta (diameter 1.2 cm) could also be ruled out (Figure 2c).

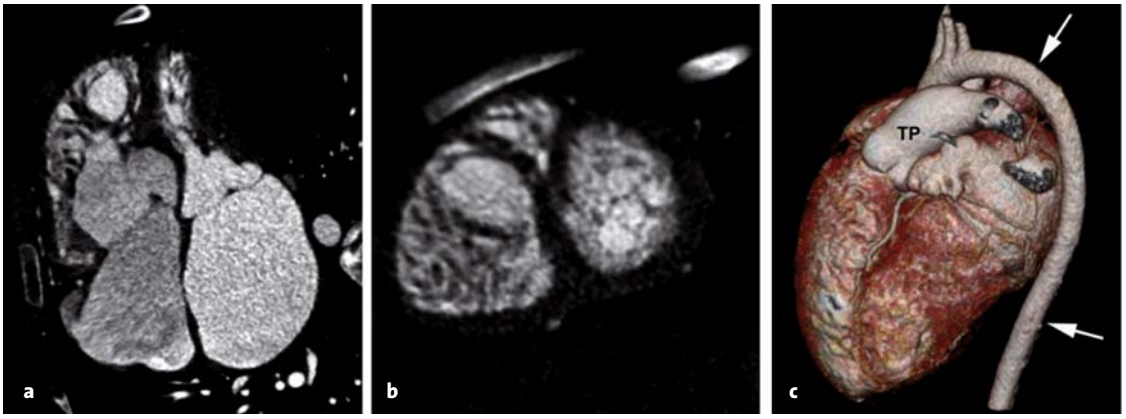
Due to progressive worsening of his clinical condition, along with a rapid reduction of ejection fraction to 34%, the patient was placed on the heart transplantation list where he is still awaiting a donor heart.

Isolated ventricular noncompaction is a rare form of cardiomyopathy caused by arrest of intrauterine endomyocardial morphogenesis in the early phase of embryogenesis with lack of compaction of the loose meshwork of ventricular trabeculae [1]. The arrested development leads to diffuse prominent deep trabeculations in hypertrophied and hypokinetic myocardial LV segments.

A comparable, though different entity is ventricular noncompaction associated with congenital heart disease. This form of noncompaction is typically associated with obstruction of the LV and/or RV outflow tract [2]. In these patients, regression of the embryogenic sinusoids is impaired during ontogenesis by ventricular pressure overload resulting in deep recesses that may communicate with both ventricular cavities and even with the coronary artery system [3]. This type of noncompaction often involves both ventricles.



Figures 1a and 1b. TTE. a) Apical four-chamber view demonstrating the enlargement of the atria and the trabecular structure of the ventricular walls. b) Parasternal short-axis view showing the LV apical segments with a compact epicardial layer and an endocardial layer consisting of a prominent trabecular meshwork and deep intertrabecular recesses. Note the color Doppler flow between the ventricular cavity and intertrabecular spaces (inset).



Figures 2a to 2c. a) CT reformation similar to the apical four-chamber view of TTE demonstrating the enlarged atria and the biventricular noncompacted myocardium. b) Apical short-axis CT reformation (similar to TTE) demonstrating the thin, compacted myocardium on the epicardial side and the thicker noncompacted myocardium on the endocardial side. c) Volume-rendered CT reformation demonstrating the enlarged pulmonary trunk (TP) and the hypoplastic thoracic aorta (arrows).

The diagnosis of noncompaction is commonly made with echocardiography using established diagnostic criteria [4]. The imaging diagnostic criteria for CT, however, have not yet been defined. It appears most reasonable to adhere to the criteria published for cardiac magnetic resonance imaging [5], measuring the thickness of the noncompacted and compacted myocardium perpendicular to the compacted myocardium on diastolic long-axis reformations, while choosing that plane with the most pronounced trabeculations. A ratio of > 2.3 then accurately identifies ventricular noncompaction.

Using current CT technology, the diagnosis of ventricular noncompaction can easily be made, including the depiction or exclusion of coexisting congenital heart disease and – such as in our patient when using a triple rule-out protocol – the exclusion of pulmonary embolism.

References

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