

IMAGES

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Abstract

Biventricular noncompaction is a recently recognized rare form of cardiomyopathy. It is characterized by altered structure of myocardial wall as a result of intrauterine arrest of compaction of the myocardial fibers in absence of coexisting congenital lesion. Left ventricle is the most affected site for noncompaction, but right ventricular involvement has been reported in a few cases. Diagnosis is made with 2-dimensional echocardiography or cardiac magnetic resonance imaging. While major clinical manifestations are heart failure, arrhythmias and embolic events, pulmonary artery hypertension (PAH) has not been well elaborated in the literature. We present a 13-year old boy who had Biventricular noncompaction complicated by severe pulmonary hypertension. Pulmonary hypertension may be a consequence of increased pulmonary venous pressures caused by systolic and diastolic left ventricular dysfunction secondary to noncompaction. We review the literature particularly with reference to PAH in the context of our case.

MeSH

Echocardiography; Isolated Noncompaction of the Ventricular Myocardium

Noncompaction of the ventricular myocardium is characterized by a distinctive (“spongy”) morphological appearance of the 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 Right ventricular (RV) noncompaction on the other hand is less well described or characterized. This is because the right ventricular apex is normally heavily trabeculated, thus by echocardiography or magnetic resonance imaging (MRI), it may be difficult to distinguish it from normal variation. The diagnosis of ventricular noncompaction is usually made using echocardiography and cardiac (MRI).^{1,3} We present a patient with biventricular noncompaction and severe pulmonary hypertension, a very rare combination reported in the literature.

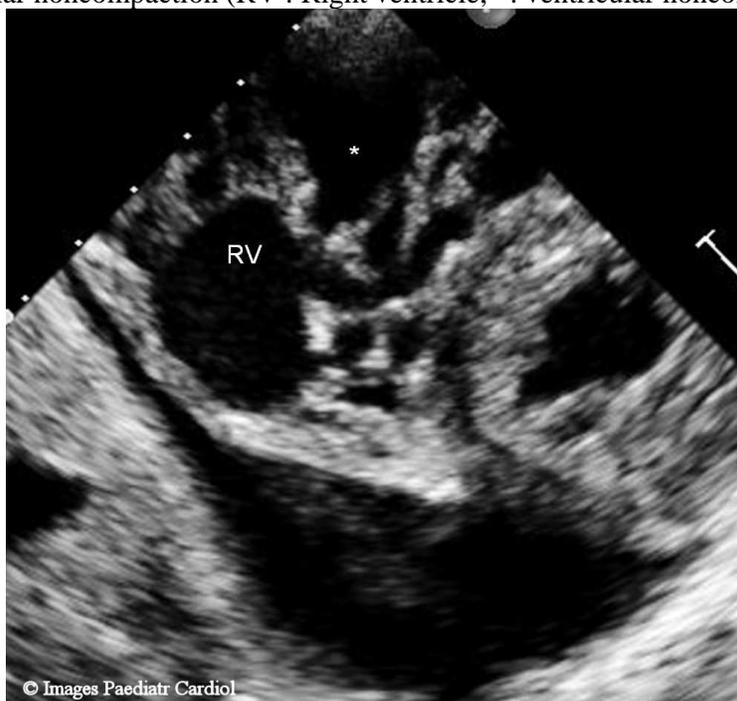
Case Report

A 13-year-old man with chronic heart failure was referred for diagnostic assessment. There was a history of progressively worsening effort intolerance and dyspnoea for the last year. There was no family history of heart disease or sudden death. On admission, his blood pressure was 100/64 mmHg and the pulse rate was 76 beats/min. The 12-lead ECG showed sinus rhythm. Chronic interstitial lung congestion and massive cardiomegaly was found on chest X-ray. Transthoracic echocardiography revealed a normally sized, hypokinetic left ventricle (ejection fraction 37%) accompanied by massive enlargement of the right and left atria. There was spontaneous echo contrast in both ventricles. The apical portions of both ventricles had a markedly trabeculated, spongy appearance, indicating biventricular noncompaction. The maximum end-systolic ratio of the noncompacted endocardial layer to the compacted myocardium was more than 2. Doppler examination showed color flow between the prominent trabeculations. Moreover, there was severe tricuspid regurgitation. The calculated peak systolic pulmonary artery pressure by tricuspid regurgitation (TR: gradient 75 mmHg) revealed severe pulmonary hypertension. The restrictive left ventricle inflow and decreased myocardial velocities suggested significantly elevated left ventricle end-diastolic pressure. The child was started on oral decongestants and anticoagulation.

Figure 1: 2D image echo image (modified apical 4C view) showing ventricular noncompaction. Note the dilated atrias secondary to high ventricular end diastolic pressures. (LV : Left ventricle, RV: Right ventricle, RA: right atrium, LA : left atrium, * : ventricular noncompaction)



Figure 2: 2D image echo image (modified parasternal long axis view) showing ventricular noncompaction (RV : Right ventricle, * : ventricular noncompaction)



Discussion

Noncompaction of the LV myocardium (the most common site of noncompaction) is a rather rare condition with a prevalence in adults of $<0.3\%$.² Rarely, only RV involvement has been reported, and biventricular noncompaction (BVN) forms a very rare subset with a few case reports.⁴ Although coexisting anomalies such as ventricular septal defect, aortic stenosis, coarctation and Ebstein's anomaly have been described, we shall be restricting our evaluation to those with isolated BVN, in the absence of other congenital heart diseases. Rare cases of fetal BVN regressing with age in the neonatal period have been described, while no such description is available for an adult with BVN.⁵

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. This characterization of non compaction is described only for the left ventricle. No such criteria exist for the right ventricle. Our diagnosis of right ventricle non compaction was based on a visual appearance on echocardiography. Some authors suggest that the same definition as for the left ventricle can be applied.

Predominant segmental location of the abnormality is almost always found in the apical and mid-ventricular areas of both the inferior and lateral walls.⁶ MRI is also used to detect ventricular noncompaction and has the advantage of good spatial resolution at the apex and lateral wall of the left ventricle.⁸ An important differential diagnostic consideration is the presence of prominent trabeculations in the RV, a

common variant of normal hearts, but these, however, most often course from the free wall to the ventricular septum. 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, conduction defects, syncopal episodes, systemic embolic events and arrhythmias.

A case report of biventricular noncompaction associated with severe pulmonary hypertension in a 20 year old who had presented with chronic heart failure and atrial fibrillation, claims to be the first case report of severe PAH in the literature. Their findings seem to be consistent with our case. Pulmonary hypertension may be a consequence of increased pulmonary venous pressures caused by systolic and diastolic heart dysfunction secondary to the noncompaction. We therefore reviewed the literature and found PAH to be a definitive feature present in cases although less so emphasized (table 1).

Table 1: Highlighting salient features of cases of isolated biventricular noncompaction from the literature search

Case	Age/Sex	Presentation	PAH	Diagnostic Modality	Journal	Year Of Publication
1 ⁹	17 y/M	Palpitation, syncope	Not known	echo	Dicle Tip Dergisi	2006 (author-1 st case report)
2 ⁸	26y/M	Mild effort dyspnea	Not Known	Echo	European J Echocardiography	2006
3 ¹⁰	58 y/M	Loss of appetite , leg edema	Not known	Echo, MRI	J Cardiol	2007
4 ¹¹	20yr/ M	heart failure, paroxysmal atrial fibrillation	Yes Severe PAH	Echo, MRI	Circulation J	2009 (claim as 1 st case report of severe PAH)
5 ¹²	23 y/F	edema	No	Echo, MRI, Cath	Chinese Medical Journal	2009
6 ⁷	29 yr/ M	palpations	Mild PAH	Echo, MRI, angio	Anadolu Kardiyol Derg	2010
7 ¹³	17y/M	Fever , dyspnea, cough , exercise intolerance	Yes Severe PAH	Echo, MRI	Chinese Medical Journal	2010
present	13 yr/ m	Heart failure	Yes Severe PAH	Echo		2010

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