AN INTERESTING CASE OF BIVENTRICULAR NONCOMPACATION

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Abstract:
Isolated noncompaction of ventricular myocardium is a form of cardiomyopathy characterized by a pattern of prominent trabecular meshwork and deep intertrabecular recesses. It is thought to be caused by arrest of normal endomyocardial morphogenesis. Previous reports show in biventricular noncompaction, left ventricle is involved predominantly more than right ventricle. Here we present a case of biventricular noncompaction where right ventricle is involved more than left ventricle associated with skeletal deformity and developmental delay. The 23 year old man presented with complaints suggestive of congestive heart failure of 9 month duration. Examination revealed raised JVP with normal auscultatory findings. Echo showed prominent trabeculations involving both ventricles with right ventricle being involved more. Colour Doppler showed entry of blood into these recesses in continuity with ventricular cavity. Contrast Echo showed free entry into and out of these recesses. MRI showed prominent trabeculations involving both ventricles. It also showed an end-systolic ratio of non-compacted to compacted myocardium ratio of more than 2.0.

Keyword: Biventricular noncompaction, Trabeculations, intertrabecular recesses, contrast Echo, MRI

BIVENTRICULAR NONCOMPACATION- A RARE CASE REPORT

INTRODUCTION:
Isolated noncompaction of the ventricular myocardium (INVM) is a rare congenital heart disease that results from an abnormal arrest in endomyocardial embryogenesis (1). It is characterized by the presence of prominent ventricular myocardial trabeculations and deep intertrabecular recesses in the absence of other structural heart defects. Recent genetic analysis has presented evidence that mutations in the G4.5 gene on the Xq28 chromosomal region are responsible for the pathogenesis of INVM (7). Previous reports show in biventricular noncompaction left ventricle is involved predominantly more than right ventricle. Previous reports also show INVM...
is also associated with facial dysmorphism and developmental retardation. Here we present a case of biventricular noncompaction where right ventricle is involved in a predominant manner associated with abnormal facies and developmental delay.

**CASE REPORT:** A 23 year old man presented with complaints of shortness of breath of 9 months duration, abdominal distension followed by swelling of legs for 1 month duration. He had no previous history of heart disease. He was born of second degree consanguineous marriage and his siblings are normal. He was a known patient of seizure disorder. Examination revealed low set ears, prominent forehead, high arched palate and micrognathia and bilateral hydrocele. Systemic examination revealed raised JVP with prominent v and y descent with normal auscultatory findings. Chest X ray showed enlarged right chambers with normal pulmonary vasculature. Echo showed prominent trabeculations and intertrabecular recesses in both ventricles predominantly in the right with evidence of biventricular dysfunction with restrictive physiology.

**Fig.1 Showing facial dysmorphism**
**Fig.2 Showing low set ears**
**Fig.3.ECHO Modified short axis view showing noncompaction involving left ventricle**
DISCUSSION:

Fig.4. Colour Doppler showing blood flow into the inter trabecular recesses of right ventricle

Fig.5. Colour Doppler showing flow of blood into the recesses of LV
Doppler echo also showed flow of blood into the intertrabecular recesses. Contrast echo showed entry of saline contrast into the intertrabecular recesses. ECG showed first degree AV block. 24 Hour Holter study showed asymptomatic ventricular ectopics. Cardiac MRI showed hypertrabeculation and hypokinesia of both ventricles. MRI also showed ratio of non-compacted to compacted myocardial ratio of 3.7 and 2.3 in left and right ventricles respectively. The patient was treated with antifailure measures and was referred for cardiac transplantation.

Fig.6. Contrast echo showing entry of bubbles into the recesses
Fig.7. MRI showing noncompaction involving the ventricle

Fig.8. MRI showing noncompaction involving right ventricle

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Although the etiology of INVM is not fully elucidated, the disease is thought to be a morphogenetic abnormality that involves an arrest of compaction of the loose myocardial meshwork during fetal development. Normally between the foetal 5th week and 8th week, inter-trabecular spaces are obliterated and ventricular compaction occurs from the base towards the apex and from epicardium to endocardium, and an arrest in the progression of ventricular compaction results in non-compaction. (8) The left ventricle is uniformly affected, but biventricular noncompaction has been reported with right ventricular noncompaction described in less than one-half of patients (2). Because of difficulty in distinguishing normal variants in the highly trabeculated right ventricle from the pathological noncompacted ventricle, several authors dispute the existence of right ventricular noncompaction (1,3). Moreover as the echocardiographic quality is operator dependent, the prevalence of RV noncompaction is underestimated in the past. MRI, however, may accurately depict morphological abnormalities such as prominent trabeculations with deep intratrabecular recesses and motion abnormalities of RV as the present case and strengthen the diagnosis. Patients who are symptomatic at presentation and who follow a rapidly progressive clinical course may show hemodynamic properties similar to DCM, whereas asymptomatic patients may follow a slowly progressive course of restrictive hemodynamic physiology, our case study has demonstrated. The complex anatomy of the abundant trabecular network may limit distensibility of the left ventricle and cause restrictive hemodynamics. (6) Furthermore, progressive subendocardial ischemia and subendocardial fibrosis, presumably related to isometric contraction of the penetrating intratrabecular recesses, might also contribute to the development of restrictive hemodynamics later in childhood. (2,6) The following echocardiographic criteria for IVNC have been reported by Jenni et al (1): (1) The absence of any coexisting cardiac anomalies. (2) The characteristic appearance of numerous, excessively prominent trabeculations and deep intertrabecular recesses. (3) Intertrabecular spaces filled by direct blood flow from the ventricular cavity as visualized from colour flow Doppler. (4) An end systolic ratio of noncompacted to compacted layers greater than 2. Noncompaction cardiomyopathy presents in three forms-Heart failure, Arrhythmias and embolic events with heart failure being the most common mode of presentation (53%) (3). Management is aimed at the symptomatic treatment of heart failure, arrhythmias and prevention of embolic events. Cardiac transplantation is indicated in cases of refractory congestive heart failure. Prognosis is worse in patients with heart failure NYHA classes III-IV, LV end diastolic diameter 60 mm, LBBB and chronic atrial fibrillation. (3)

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Clinician updates


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