NONCOMPACTION OF LEFT VENTRICLE WITH VENTRICULAR SEPTAL DEFECT
-A CASE REPORT

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Abstract:
Noncompaction of left ventricle in association with congenital heart disease is a rare entity. We encountered a case of noncompaction of left ventricle with large muscular ventricular septal defect with severe pulmonary hypertension. Echocardiography was used for confirmation. We report this case because of its rarity.

Keyword: Noncompaction of left ventricle, trabeculae, pulmonary hypertension

Introduction
Isolated non compaction of ventricular myocardium (IVNC) is a rare congenital disorder. Noncompaction of left ventricular myocardium in association with other congenital heart defects is quite rare. We report a case of noncompaction of left ventricle with large ventricular septal defect with bidirectional shunt due to severe pulmonary hypertension.

Case report
33 yr old male presented to us with history of breathlessness of class 2 severity and fever of one month duration. He gave history of heart disease since childhood but lost all his records. On examination he was febrile and had mild central cyanosis, jaundice, mild pedal edema, elevated JVP and Grade I clubbing. SpO2 at room air was 90%. His pulse was regular at 120 bpm and blood pressure was 110/80 mm Hg. Cardiac examination revealed normally placed apical impulse with parasternal heave, palpable and single second sound with no added sounds or murmur. ECG revealed sinus rhythm, right axis deviation, biventricular hypertrophy with monophasic R in V1 and normal QRS duration.(Fig1).
Figure-1 ECG shows sinus rhythm, right axis deviation, biventricular hypertrophy

Chest X ray showed cardiomegaly, right atrial enlargement and features of pulmonary arterial and venous hypertension. (Fig.2).

Figure-2 Chest X-ray shows dilated pulmonary artery, cardiomegaly and pulmonary artery hypertension

Echocardiography revealed dilated ventricles with global hypokinesia and severe biventricular dysfunction with an LV ejection fraction of 25%. Mid and apical segments of left ventricle appeared highly trabeculated with prominent intertrabecular recesses with color flow moving in and out of the recesses.(Fig3,4).

Further examination also revealed a 13 mm basal muscular ventricular septal defect with bidirectional shunt, mild tricuspid regurgitation jet with a peak gradient of 85 mm Hg suggesting severe pulmonary arterial hypertension and no evidence of vegetation.(Fig.5,6)
Figure-5 Echocardiogram: Modified four chamber view showing muscular ventricular septal defect and noncompaction of left ventricular apex.

Figure-6 Echocardiogram – The color map showing the shunt across the defect and mild tricuspid regurgitation.

Blood investigations showed polymorphonuclear leucocytosis. Rest of the workup for fever was negative. Liver enzymes were elevated. He improved with a course of antibiotics and decongestive measures including digoxin, diuretics, sildenafil and other supportive measures. Subsequently his liver enzyme levels normalized. His left ventricular dysfunction persisted.

Discussion
In 2001 Jenni et al(1) proposed IVNC should be classified as distinct cardiomyopathy and pathoanatomical and echocardiographic criteria were described. Left ventricular noncompaction (LVNC) is characterized by the presence of extensive trabeculated myocardial wall within the luminal aspect of the compact myocardium of the ventricle. The trabeculae are excessive in number and more prominent than normal. Noncompaction may occur in isolation, usually with clinical features of dilated cardiomyopathy or it may be associated with congenital or acquired heart disease.

Echocardiography is the reference standard for diagnosis. Noncompaction is usually considered to result from persistence of highly trabeculated myocardium found in early cardiogenesis of the human embryo. LVNC is characterized by segmental thickening of left ventricular wall consisting of 2 layers of thick compacted epicardial and thickened endocardial layer with marked trabeculations and deep intraventricular recesses. Thick trabeculated layer extends from mid portion of left ventricle (LV) to apex. Between the network of trabeculae are deep recesses in continuity with LV cavity but not with epicardial coronary system. LVNC most commonly affects mid lateral, mid inferior and apical segments of LV. Interventricular septum may be infrequently involved and base of left ventricle is never involved. Noncompaction may affect both ventricles. The normal architecture of Right ventricle (RV) is dominated by a trabecular pattern; this has made the condition less apparent in the RV and diagnosis of RV noncompaction is currently qualitative. Non compaction is not thought to affect atria. In our case LVNC is associated with muscular ventricular septal defect (VSD). Literature review reveals VSD is one of the most common congenital heart disease noted in association with LVNC appearing in 53% of survey of reports concerning LVNC with congenital heart disease. Muscular VSD’s may comprise 90% of total VSD (3) noted to occur in association with LVNC. The association may be
1 coincidental because VSDs are the commonest congenital heart disease accounting for approximately 20% of all cardiac pathologies

2 due to developmental association of VSD and LVNC. In this regard it is of interest to note that the formation of interventricular septum is the result of coalescence of trabecular sheets, thus development of muscular interventricular septum and compaction of myocardium are closely linked processes. Small VSD have been proposed to result from incomplete or abnormal coalescence of embryonic trabecular sheet(4).

Management of this condition includes decongestive measures, antiarrhythmic drugs if indicated and anticoagulation if patient is at risk of systemic embolism. Very sick patients with recurrent and refractory congestive cardiac failure may require cardiac transplantation. Complications (5) include congestive cardiac failure (53%), ventricular arrhythmia (41%), systemic embolism (24%) and sudden cardiac death

Conclusion
Our patient had noncompaction of left ventricle, large muscular ventricular septal defect with severe pulmonary hypertension (Eisenmenger complex). This combination has not been reported so far in literature as of our knowledge.

REферENCES:


