A RARE CASE OF NON COMPACTION OF RIGHT VENTRICLE

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Abstract:
Isolated ventricular non compaction is a rare type of cardiomyopathy with uncertainty. This results from arrest of ventricular compaction of myocardium during embryogenesis. This disorder is diagnosed mainly by transthoracic echocardiography by presence of excessive prominent trabeculations with deep intertrabecular recesses. Magnetic resonance imaging is needed in some cases for confirming the diagnosis. Clinical manifestations include heart failure, atrial and ventricular arrhythmias, embolic events. Usually left ventricle is involved in majority of patients, isolated right ventricle involvement has been reported rarely. Here we report a case of isolated right ventricular noncompaction in a 19 year old male who presented with severe right heart failure and atrial fibrillation.

Keyword: Ventricular noncompaction, Trabeculations, Cardiomyopathy, Right heart failure.

Nineteen year old male presented to our department with history of dyspnoea NYHA class IV, pedal edema, facial puffiness, abdominal distension, since 10 years of age. He was treated as with recurrent admissions to hospital in his home town and was referred to our department for expert opinion. On reviewing his past details he was diagnosed to have heart failure and was treated with diuretics and digoxin for past 9 years. His symptoms worsened intermittently over that period and for past 6 months symptoms were static. No other significant past history. No family history of similar illness. On examination he was conscious, dyspnoeic, tachypnoeic, pale, mildly cyanosed with pedal edema, no clubbing, pulse rate 110/min, irregularly irregular, blood pressure – 100/60 in right upper limb. Jugular venous pressure was elevated with prominent V waves. Cardiovascular exam showed cardiomegaly with hyperdynamic apical
impulse in left 6th space 2 cm lateral to mid-clavicular line, prominent epigastric pulsation, grade 2 parasternal heave. Auscultation showed S1, S2 normal in intensity, right ventricular S3 present, grade 3/6 pansystolic murmur in apex and grade 4/6 pansystolic murmur in left sternal border. Lung fields showed bi basal crepts. Abdominal examination showed hepatomegaly with free fluid. Investigations showed blood haemoglobin of 9 gm%, peripheral smear showed microcytic hypochromic anaemia, PCV 32%, polymorphs 62%, lymphocytes 31%, monocytes 5%, eosinophils 2%. Urea 28 mg, creatinine 0.9 mg, liver function tests showed mild elevation of SGOT and SGPT, thyroid function tests were normal. Ultrasound abdomen revealed mild ascites with congested liver. ECG showed atrial fibrillation with ventricular rate of 78/min, normal axis, RBBB, Chest X ray PA view showed massive cardiomegaly, with RA and RV enlargement, dilated pulmonary arteries. Echocardiography showed dilated right atrium and right ventricle along with right ventricle apex and free wall showing numerous excessive prominent trabeculations and deep intertrabecular recesses, Colour flow imaging showed the intertrabecular recesses are filled with blood from the ventricular cavity. He also had severe low pressure tricuspid regurgitation with severe right ventricular dysfunction. He was diagnosed to have isolated non compaction of right ventricle and treated with decongestive measures, oral anticoagulants, rate control for atrial fibrillation.

DISCUSSION Isolated non compaction of right ventricle is a rare form of cardiomyopathy classified by WHO classification of cardiomyopathies as unclassified cardiomyopathy. It was first reported by Chin et al in 1990 with 8 cases. Isolated non compaction of RV is a rare disorder with uncertain etiology. Reported prevalence is between 0.014 to 1.3% of general population. During development heart is like a sponge like mesh with interwoven myocardial fibres. This spongy like heart undergoes compaction and converted into solid structure, the arrest of compaction of ventricles leads to noncompaction which is common in left ventricle, biventricular involvement and rarely isolated right ventricle can be involved. Noncompaction can sporadic or familial. Mutations in different genes has identified include G4.5 found in Tafazzins, Gene for cytoskeletal protein CYPHER/ZASP, Chromosome 11p15 etc. Clinical manifestations vary ranging from asymptomatic status to heart failure, arrhythmias, thromboembolism. Diagnostic criteria for isolated non compaction of ventricles by Echocardiography are as follows: Absence of any coexisting cardiac anomalies, Presence of numerous excessive prominent trabeculations with deep intertrabecular recesses, Intertrabecular recesses are filled by direct blood flow from ventricular cavity as visualised by colour flow imaging, Ratio of noncompacted layer to that of compacted layer of >2. Finding of this two layered structure of ventricular wall is predominant in apical, mid and superior regions of right ventricle free wall. In our patient echocardiography showed prominent trabeculations with deep intertrabecular recesses predominantly involving the apical and entire free wall of right ventricle with colour Doppler showing blood flow from ventricular cavity into trabeculations and recesses. Also there is right atrial dilatation and low pressure severe tricuspid regurgitation. Left ventricular structure and function were normal. There are no associated cardiac anomalies. Transthoracic echocardiogram is a good tool for diagnosis of
noncompaction. Additional investigations like TEE, ventriculography, computed tomography scan, magnetic resonance imaging are also used in diagnosis of this condition. Though echocardiography remains the cornerstone in the diagnosis of ventricular noncompaction sometimes the apical portion of ventricle cannot be imaged properly by echocardiography. This leads to underestimation of degree of noncompaction so MRI is the method of choice in those patients. The ratio of noncompacted layer to that of compacted myocardium of more than 2.3 which has sensitivity of 86% and specificity of 99% in diagnosis by MRI.\(^7\)\(^8\)\(^9\)

We have done magnetic resonance imaging in our patient which showed prominent trabeculations with intertrabecular recesses in apical and mid superior regions of right ventricular free wall with ratio of noncompacted to that of compacted layer of more than 2.4. Thus the diagnosis was confirmed both by transthoracic echocardiography and magnetic resonance imaging in our patient. Jenni et al in follow up of 34 patients of isolated ventricular noncompaction seen between 1986 to 1999 largest reported in literature showed the clinical manifestations were heart failure [53%], sudden cardiac death [35%], embolic events [24%], syncope [18%] and atrial fibrillation [26%].\(^5\) Our patient presented predominantly with right heart failure and atrial fibrillation. Patients with isolated ventricular noncompaction have variable clinical course ranging from asymptomatic state to severe cardiac disability and death. Prognosis is worst in patients who have NYHA class III–IV symptoms, dilated ventricles, chronic atrial fibrillation, left bundle branch block.\(^4\) Our patient is in worse prognosis category. Our patient was treated with loop diuretics, ACE inhibitors, aldosterone antagonists, digoxin, beta blockers, warfarin. He is currently awaiting heart transplantation.

REFERENCES:


