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RESEARCH ARTICLE

BIVENTRICULAR NON-COMPACTION CARDIOMYOPATHY WITH LEFT VENTRICULAR DYSFUNCTION

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Abstract

Non-compaction of the ventricular myocardium involving both ventricles is a rare condition and characterized by deeply increased trabeculations in one or more segments of the ventricle. Non-compaction is thought to be due to intrauterine arrest of compaction of the loosely interwoven meshwork present in fetal myocardial primordium. More recent studies suggest that non-compaction of the ventricular myocardium can be acquired later on in life sporadically.The apical segment of the left ventricle is most commonly affected, but biventricular involvement is also described. Herein, we describe an unusual case of biventricular non-compaction cardiomyopathy with left ventricular systolic and diastolic dysfunction. A 54-year-old man presented with clinical heart failure as well as arrhythmia. Transthoracic echocardiography revealed left ventricular non-compaction with depressed ejection fraction. Cardiac magnetic resonance imaging (MRI) further revealed right ventricular noncompaction.

Introduction:-

Noncompaction of the ventricular myocardium is a recently recognized genetic cardiomyopathy characterized by the spongy appearance of the left ventricle[1]. It can be a genetic or sporadic condition.Left ventricular non-compaction (LVNC) has been found in 0.014%–1.3% of patients undergoing echocardiography and biventricular non-compaction is even rarer. Prominent trabeculations are a normal feature of the developing myocardium in utero and left ventricular (LV) non-compaction is thought to result from a failure of the trabecular regression that occurs during normal embryonic development[2]. Alternatively, the pronounced hypertrabeculation may be due to altered regulation in cell proliferation, differentiation and maturation during ventricular wall formation[3,4].The diagnosis of ventricular noncompaction is usually made using transthoracic echocardiography and cardiac MRI.The patient may have variable Clinical manifestations of heart failure, thromboembolic events, chest pain,arrhythmias and risk of sudden cardiac arrest.

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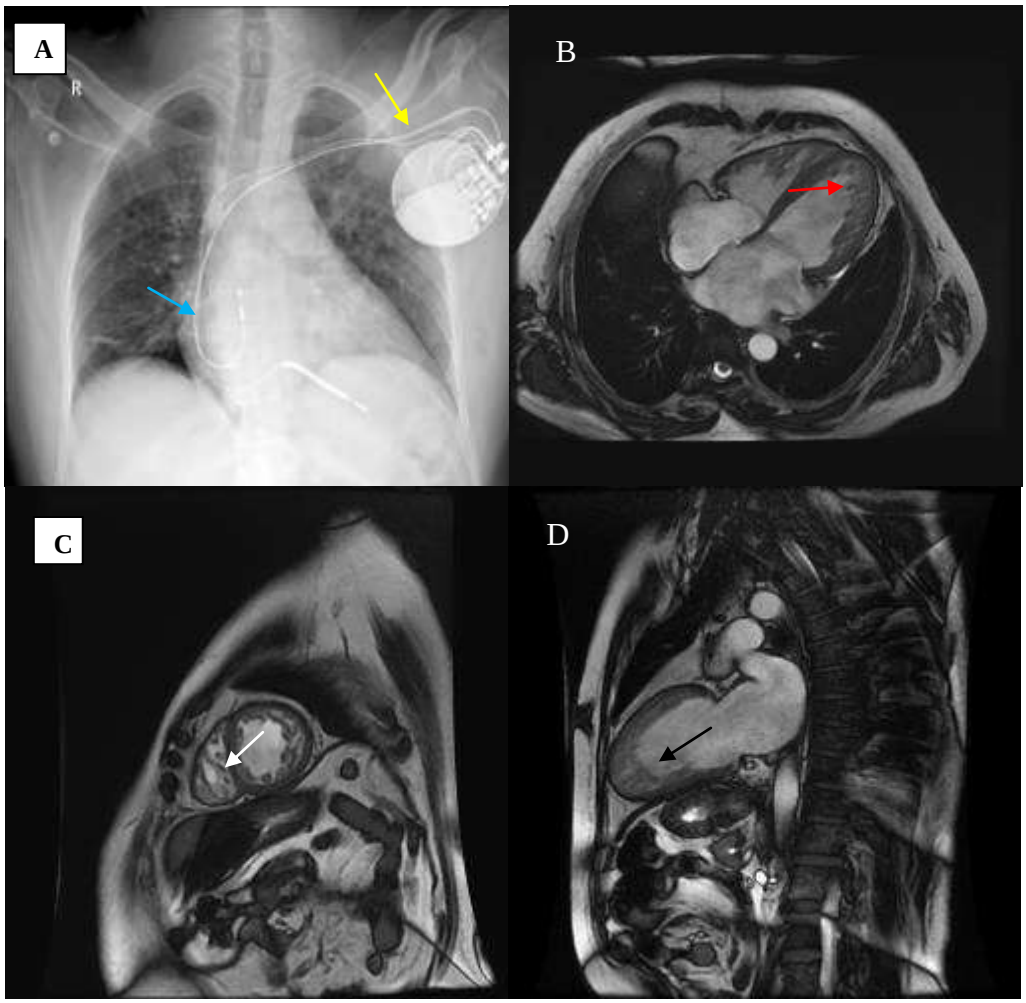
Case Description

A 54-year-old male presented with complaints of dyspnoeaand syncope. On admission, his blood pressure was 100/65mmHg and his pulse rate was 100beats/min. ECG showed supraventricular tachycardia with a right axis deviation and evidence of left ventricular hypertrophy. Echocardiography showed global hypokinesia with a thickened trabecular meshwork of the left ventricle, and an ejection fraction of 39%. Chest X-Ray (Panel A) showed Cardiomegaly (blue arrow) and pacemaker (yellow arrow). Cardiac MRI four-chamber view (Panel B) showed increased ventricular myocardial thickness with an increase in subendocardial trabeculations(Red arrow)and an increased ratio (>2.5) of non-compacted thickness to compacted myocardial thickness.An increase in subendocardial trabeculations (white arrow) of the right ventricle was noted in the short-axis view (Panel C).The vertical long-axis view (Panel D) showed thickened trabecular meshwork of the left ventricle (Black arrow).Ventricular non-compaction can be sporadic or familial. Biventricular non-compaction is very rare and associated with heart failure and arrhythmias. Our patient, who did not have RV trabeculations on initial echocardiography, showed RV trabeculations on cardiac MRI. Cardiac MRI scores over echocardiography in evaluation of the RV noncompaction. Thus, the prevalence of RV involvement may have been underestimated in the past. CMR aids in the diagnosis of biventricular involvement.



Discussion:-

Left ventricular noncompaction is rare cardiomyopathy, that is thought to arise during embryogenesis secondary to arrested myocardial development. This results in a thickened myocardium comprised of non-compacted myocardium, characterized by prominent deep intertrabecular recesses and a thin compacted layer of the myocardium[5]. The American Heart Association defines LVNC as genetic cardiomyopathy while the European Society of Cardiology classifies it as unclassified cardiomyopathy[6]. Mutations in the G4.5 gene on the Xq28

chromosome result in a wide range of X-linked cardiomyopathies in the paediatric population. However, this mutation was not found in the adult population, where the autosomal dominant mode of transmission was more common.

Echocardiography criteria for diagnosis of left ventricular non-compaction[7]:
(1) A thickened left ventricular wall composed of two layers: a thin compacted epicardial layer and a non-compacted endocardial layer with numerous prominent trabeculations and deep recesses: a maximum ratio of non-compacted to compacted myocardium being >2:1 at end-systole in the parasternal short-axis view.
(2) colour Doppler evidence of flow within the deep intertrabecular recesses.
(3) prominent trabecular meshwork in the left ventricular (LV) apex or midventricular segments of the inferior and lateral wall.

Visualization of the right ventricle is more difficult because of its irregular shape. Therefore, diagnosis of RV noncompaction seems to be more difficult. An important differential diagnostic consideration is the presence of prominent trabeculations as a common variant of the normal heart.

Cardiac MRI criteria for Left ventricular non-compaction:
(1) A maximum end-diastolic non-compacted to compacted myocardial thickness ratio of >2.3 was the best criterion for LVNC and yielded a sensitivity of 86% and specificity of 99% for LVNC.
(2) A trabeculated LV mass > 20% of global LV mass was identified as a criterion for LVNC with a sensitivity of 94% and a specificity of 94%.
(3) Fractal dimension as a quantitative measure of trabeculation is high in LVNC and may be very accurate; however, furtherfollow-up is needed to better identify its role.
(4) An end-systolic non-compacted to compacted (thickness) ratio ≥ 2.0 was found to be more strongly associated with heart failure and with adverse events (heart failure, death, heart failure readmission, embolic events and ventricular arrhythmias) than other criteria for LVNC.

Conclusion:-

There are no specific guidelines or treatments for the management of non-compaction cardiomyopathy. Patients are managed according to their specific clinical presentation and corresponding guideline. The benefit of ICD placement in this population is unknown as well. The deep trabeculations of the heart may predispose patients to complications, such as ventricular perforation in the setting of device placement. In conclusion, non-compaction cardiomyopathy has a genetic origin that can be found in isolated form or in association with other cardiomyopathies. It can also be acquired later on in life in response to mechanical stress, in patients who are genetically predisposed. A better understanding of its genetics may further delineate the natural course of this disease. This will help in early diagnosis as well as its management. More studies are necessary to determine its proper management.Genetic counselling should be advised as it can be familial.

Abbreviations
LVNC - Left ventricular non-compaction
CMR- Cardiac magnetic resonance imaging
MRI - Magnetic resonance imaging
RV – Right ventricle
LV – Left ventricle
ICD - Implantable cardiac defibrillator

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Conflict Of Interest
No Conflict of interest.

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