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RESEARCH ARTICLE

LEFT VENTRICULAR NON COMPACTION CARDIOMYOPATHY ASSOCIATED WITH MITRAL VALVE PROLAPSE

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Abstract

Left ventricular non compaction (LVNC) is a rare condition characterized by prominent ventricular trabeculations and intertrabecular recesses,Mitral regurgitationwhen present is usually a result of annular dilation, mitral valve prolapse due to chordae tendinae rupture is quite rare.We present the case of a 48 year old male with no medical history who presented with worsening exertional dyspnea classified as NYHA class III. The physical examination found a systolic murmur heard at the apex. Transthoracic echocardiography and cardiac magnetic resonance confirmed the diagnosis of Left ventricular non compaction associated with posterior mitral valve prolapse with severe mitral regurgitation and preserved ventricular function who was successfully operated and found asymptomatic during the follow up.

Introduction:-

Left ventricular non-compaction (LVNC) is a condition characterized by excessive ventricular trabeculations and intertrabecular recesses, the left ventricular muscle is therefore thick and sponge-like. It's quite rare with reported incidence rates in adults ranging from 0.014% to 0.24% (1, 2). In the case of LVNC the presence of mitral regurgitation is often due to annular dilatation, mitral valve prolapse due to the rupture of a chordae tendinae in in this context is a rare occurrence.

Casereport

We present the case of a 45-year-old male patient with no significant past medical history, who was referred to our department due to worsening exertional dyspnea classified as NYHA class III. The physical examination found a systolic murmur heard at the apex described as blowing murmur graded at 4/6, with no signs of congestive heart failure. The electrocardiogram showed a regular sinus rhythm with left atrial enlargement and left ventricular hypertrophy. Transthoracic echocardiographyshowed moderate dilation of the left heart chambers with a hypertrabeculated myocardium and posterior (P1 and P2) mitral valve prolapse due to chordae tendinae rupture with severe eccentric mitral regurgitation and moderate systolic dysfunction. Cardiac magnetic resonance CMR confirmedthe diagnosis of LVNC.

The patient underwent cardiac surgery for mitral valve replacement employing mechanical prosthesis, with uncomplicated postoperative course.

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The three monthsfollow-up showed significant clinical improvement and transthoracic echocardiography indicated a well-functioning prosthesis with preserved left ventricular function.

Figure 1:- (a) Apical 3 chamber view showing the P2 prolapse with prominent ventriculartrabeculations, (b) Apical 4 chamber view showing the deep intertrabecular recesses.

Figure 2:- Apical 4 chamber view showing the P1 prolapse with a severe eccentric mitral regurgitation.

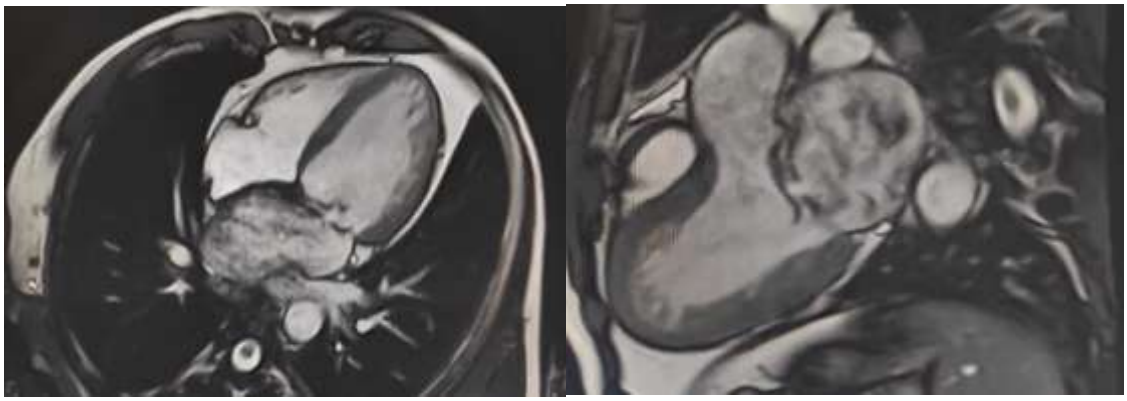

Figure 3:- Longitudinal cine cardiac MRI sequences showing the posterior mitral valve rolapsed and the prominent myocardial trabeculations.

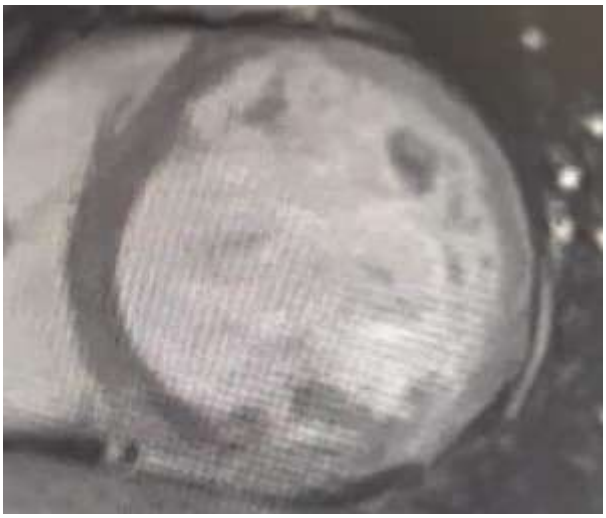

Figure 4:- Short axis cine cardiac MRI sequence showing a double layered myocardium, the inner non compacted and outer compacted with a non compacted to compacted myocardium ratio greater than 2.3 rendering the diagnosis of LVNC.

Discussion:-

Left Ventricular Non-Compaction (LVNC) is a heterogeneous myocardial disorder characterized by prominent trabeculations, intertrabecular recesses, and a left ventricular myocardium with two distinct layers: a compacted and a non-compacted layer. (3) Imaging studies and pathological assessments further demonstrate that LVNC presents as a spongy left ventricular myocardium, with abnormal trabeculations typically most pronounced at the left ventricular apex. Although Left Ventricular Non-Compaction (LVNC) primarily involves the left ventricle, there are documented cases of isolated right ventricular non-compaction as well as biventricular non-compaction.

Left Ventricular Non-Compaction (LVNC) results from hypoplasia of the compacted layer of the myocardium, attributable to the premature arrest of compaction of endocardial and myocardial fibers during embryonic development. (4), new data suggest that the excessive trabeculations are a proliferation defect (5).

The clinical presentation of Left Ventricular Non-Compaction (LVNC) is contingent upon the degree of non-compaction, the presence of associated lesions, left ventricular ejection fraction (LVEF), and the age at which symptoms first manifest. In many patients with LVNC, the initial symptoms frequently include congestive heart failure (CHF), ventricular arrhythmias, and thromboembolic events.

Manygenetic mutations in the genes that encode for the sarcomeric, cytoskeletal and nuclear membrane proteins have been associated with LVNC, similarly other gene mutations have been associated with mitral valve prolapse, one report found this association in genetically related patients suggesting a common origin of both conditions (6).

Mitral regurgitation is often caused by mitral annular dilatation in LVNC, mitral leaflets are not affected although malformation of papillary muscles and abnormal chordal attachment to valve leaflets are observed in severe cases of LVNC (7). Mitral valve prolapse (MVP) due to a rupture of the chordae tendineae is rare; To our knowledge, only 4 cases of LVNC associated with mitral valve prolapse have been reported in the literature.

Valve repair techniques aimed at the leaflet and annulus, such as resection suture techniques and ring annuloplasty, are suitable for treating mitral regurgitation associated with Left Ventricular Non-Compaction (LVNC). However, the potential for papillary muscle hypoplasia must be considered, as it may complicate surgical intervention and render the durability of artificial chordae interventions unpredictable. Left Ventricular Non-Compaction (LVNC) is associated with a significant risk of thromboembolic complications, necessitating chronic anticoagulation therapy. Consequently, mitral valve replacement with a mechanical valve may be warranted in cases where repair of mitral lesions is challenging without the use of artificial chordae techniques (8). In our case, valve replacement with a mechanical prosthesis was the surgical technique employed by the surgical team.

Conclusion:-

The association between Left Ventricular Non compaction and Mitral Valve Prolapse represents an intriguing intersection of two complex cardiac conditions. Understanding this relationship enhances our ability to diagnose, manage, and counsel patients effectively. As research continues to elucidate the genetic and developmental pathways linking these conditions, clinicians can look forward to more precise and personalized approaches to cardiovascular care.

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