SURGICAL REPAIR OF SEVERE MITRAL AND TRICUSPID VALVE REGURGITATION IN PATIENT WITH LEFT VENTRICLE NONCOMPACTION - A CASE REPORT

Aayushi Donariya[1], Herin Patel[1], Fagun Gajjar[1], Shobhit Mathur[1], Archit Patel[1], Kartik Patel[1]*

[1] Department of Cardiovascular & Thoracic Surgery, U. N. Mehta Institute of Cardiology and Research Center, Civil Hospital Campus, Asarwa, Ahmedabad, Gujarat, India

* **Corresponding author:** drkartikpgi@yahoo.com

Abstract

Left ventricular non-compaction (LVNC) cardiomyopathy is a rare congenital cardiomyopathy which is characterized by prominent and extensive trabeculation and deep intertrabecular recesses communicating with left ventricular cavity. Here we present a rare case report of patient with severe secondary Mitral regurgitation (MR) and severe tricuspid regurgitation (TR) in LVNC cardiomyopathy who underwent MV Repair using ring annuloplasty with #28 CG future ring and kay's suture annuloplasty for severe TR and its postoperative management. Patient recovered well after surgery and was discharged with improved hemodynamics. The valve surgery in patients with such cardiomyopathy is very rare. This case will add to the small growing literature of surgery in severe secondary Mitral regurgitation in LVNC and the problems we faced while coming off bypass and its postoperative management.

Keywords: *Cardiomyopathy, Left ventricular non-compaction, Mitral regurgitation*

INTRODUCTION

Left ventricular non-compaction (LVNC) cardiomyopathy is a rare congenital anomaly characterized by thin compacted epicardium and deep non compacted extensive endothelium-lined trabeculations and intertrabecular recesses which is continuous with the left ventricular cavity.[1] It is primary congenital cardiomyopathy that affect both ventricles but mainly left ventricle and characterized by intrauterine arrest of compaction of myocardium which result as presence of prominent multiple trabeculation and intertrabecular recesses which communicate with ventricular cavity and with normal epicardial coronaries.[2] Many patients remain asymptomatic and other present with congestive heart failure, arrhythmias and thromboembolism. The diagnosis is made by echocardiography or cardiac MRI by evaluating morphological criteria for LVNC. The major treatment modality is symptomatic management. Here we report a case of a patient with severe secondary mitral valve regurgitation and severe tricuspid valve regurgitation in LVNC cardiomyopathy, its preoperative, intraoperative and postoperative course and difficulties and how we overcome with it.

CASE REPORT

A 35-year-old female presented with sudden onset of breathlessness and palpitation and decreased urine output for couple of days. She was admitted and evaluated in emergency department. She was diagnosed to have exacerbated acute left heart failure with acute kidney injury (AKI) and treated with aggressive decongestive therapy.

She was suffering from dilated cardiomyopathy (DCM) for last two years and was on anti-heart failure treatment. Her transthoracic echocardiography was suggestive of severe mitral regurgitation (MR) due to myxomatous anterior mitral valve leaflet with moderate left ventricular dysfunction and severe TR. She was discharged on anti-heart failure medications and asked to follow up in cardiology and CTVS OPD. On follow up visits her AKI resolved but she was still symptomatic despite aggressive medical management. Hence she was referred to us for surgery; though her ejection fraction has improved significantly. In preoperative work up we found her indirect bilirubin to be on higher side (total bilirubin 2.1 mg/dl and indirect bilirubin 1.6mg/dl) and other investigations were unremarkable. Several investigations for diagnosis of its increased bilirubin has been done which include peripheral blood smear; retic count; LDH; haptoglobin and vitamin B12 levels; USG whole abdomen; indirect and direct coomb's test; electrophoresis: Negative for typical beta thalassemia traits but diagnosis remains unclear. After stabilization of patient, consent for surgery taken and posted as an elective case for MV repair or MVR and TV repair.

Intra operative Trans oesophageal echocardiography (TEE) revealed non compacted Left ventricle with numerous thick trabeculae and intertrabecular deep recesses. (Figure 1) It also revealed severe mitral and tricuspid valve regurgitation. Median sternotomy was performed and cardiopulmonary bypass (CPB) was instituted with aorto-bicaval cannulation. RA opened and through superior septal approach opened LA. We found small non compacted left ventricle cavity and prominent and extensive deep trabeculations and intertrabecular recesses in it and diagnosed this rare finding intraoperatively. (Figure 2)

Mitral valve was assessed and found to have dilated annulus with prolapsed anterior mitral leaflet (AML). Posterior mitral valve leaflet (PML) was tethered because of LVNC. There was commissural cleft at AML which was repaired along with annuloplasty done using 28 CG future ring. Secondary chordae of posterior leaflets were released and thinning of posterior leaflet is done by peeling. Finding the morphology suitable for repair of mitral valve we opted MV repair over MVR as it obviate lifetime anticoagulation and associated complications with it. Tricuspid valve was found to have severe regurgitation due to anterior tricuspid leaflet cleft which was repaired. Posteroseptal annuloplasty done for dilated annulus. While taking off bypass, we found trouble weaning patient off CPB due to unstable hemodynamic. Hence, intraaortic balloon pump (IABP) was placed and patient was weaned off CPB gradually.

In the post-operative Intensive Care Unit (ICU), patient was extubated after 72 hours of mechanical ventilation and weaned off inotropic support and IABP gradually then after. During this period, she went into atrial fibrillation; anti-arrhythmic drugs were added and optimized. Post-operative 2D echo with IABP showed mild mitral valve regurgitation with mild tricuspid valve regurgitation. She had improved left ventricular function and mild right ventricular dysfunction. (Figure 3) But after removal of IABP 2D echo was suggestive of mild MR with moderate LV dysfunction. Anti-heart failure medications started (Valsartan, Sacubtril) and optimized. Patient recovered well gradually then after and discharged with stable hemodynamics on day 12. She had been on follow up since then and doing well clinically.

DISCUSSION

Heart and myocardium develop through condensation and compaction of deep intertrabecular recesses and formation of capillaries and it is believed that LVNC result as failure of development of this phase of left ventricular cavity.[3] The benign subtype of LVNC has preserved left ventricular systolic and diastolic function. The arrythmogenic subtype with ventricular arrthymias has worst prognosis. Another subtypes mimic hypertrophic cardiomyopathy, dilated cardiomyopathy and restrictive cardiomyopathy. In congenital subtype, LVNC associated with other congenital heart disease including septal defect, pulmonary stenosis and with single ventricles.

The prevalence of LVNC is very rare and ranges in adult population from 0.05% to 0.24% per year in echocardiographic databases. Male are more commonly diagnosed than females.[4] The Jenni criteria or Zurich criteria, the most accepted echocardiographic criteria which states that the ratio between uncompressed myocardium to compacted myocardium ratio>2.0 at the end of systole in parasternal short axis view.[5] Peterson et al. proposed cardiac magnetic resonance imaging (CMRI) criteria which state that ratio of noncompacted-to-compacted myocardial ratio greater than 2.3 as measured in end-diastole.

Patients may be asymptomatic or may present with acute heart failure in nearly 50% of cases, arrhythmias including atrial fibrillation in 25% of cases and thromboembolic complication in 20% cases. The most common reason for hospital admission is heart failure as with our case and it is caused by systolic as well as diastolic dysfunction.

The management includes heart failure treatment, prevention of thromboembolic complication by prophylactic use of oral anticoagulation therapy and use of implantable cardiac defibillator which helps in prevention of sudden cardiac death due to ventricular tachycardia. There is still no guideline to direct us how to deal with secondary MR whose cause is not ischemic heart disease. According to ACC/AHA guideline; chronic secondary MR mitral valve surgery can be done in ischemic heart disease under class 2b indication.

However, no corrective operative therapy exists for this anomaly. There are very few anecdotal case reports of surgical repair on LVNC exist in literature till now. Takeshi Shimamoto et al performed MV repair and TV repair along with left ventricular restoration (LVR) surgery in 57-year patient with LVNC and valvular regurgitation.[6] Peri-membranous ventricular septal defect (VSD) closure done

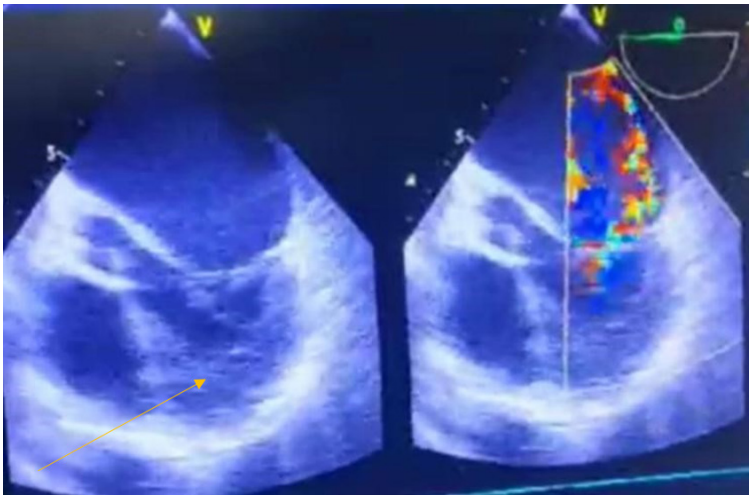

Figure 1 *Left ventricle non compaction with abnormal numerous trabeculation and intertrabecular recesses.*

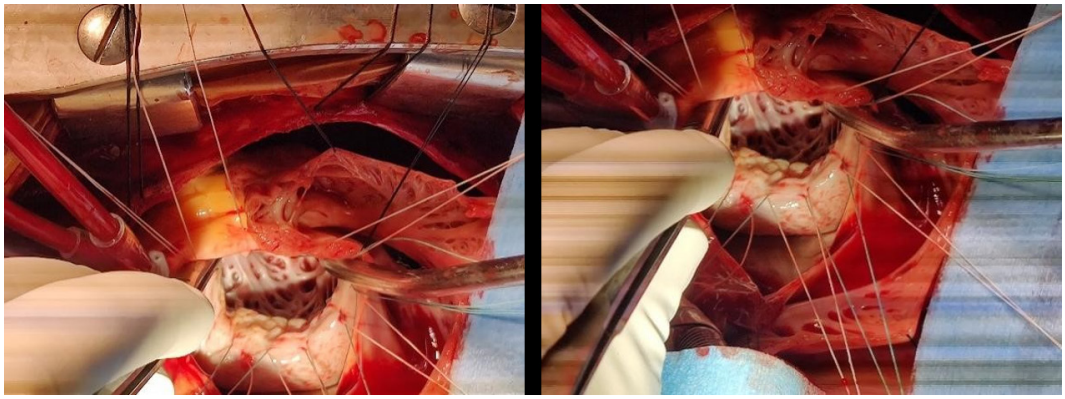

Figure 2 *Intra operative images of Left ventricle noncompaction with prominent numerous trabeculae and deep intertrabecular recesses.*

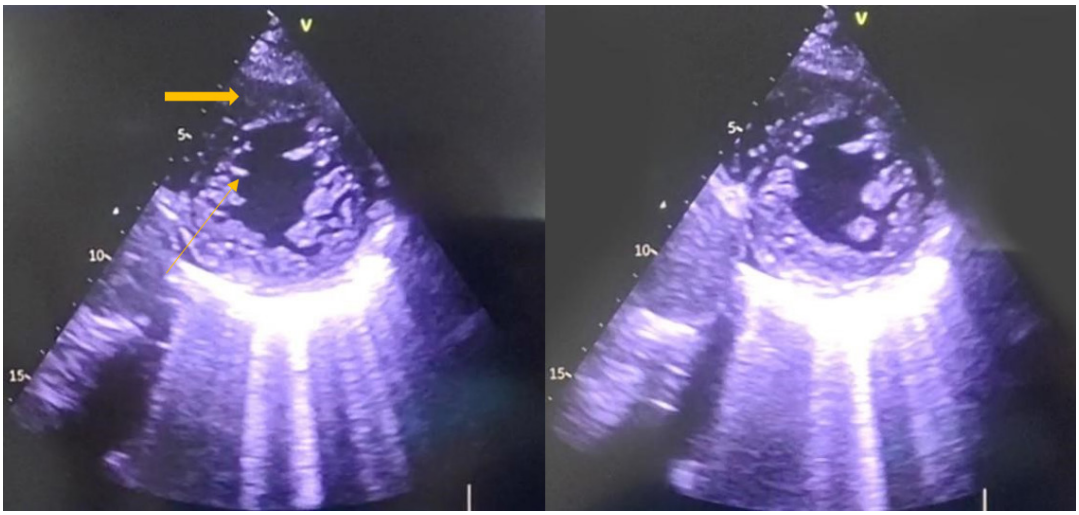

Figure 3 *Non compaction left ventricle with numerous trabeculae (arrow) and intertrabecular recesses (thick arrow).*

by Takamichi Uchiyama et al.[7] Another case of non-ischemic left ventricle aneurysm in LVNC was repaired by by Michael A Catalano et al. A case of coronary artery bypass graft (CABG) with MV repair in 73-year-old male with myocardial infarction and mitral valve regurgitation in LVNC by Mehrnoush toufan et al has been reported.[8] A few aortic valve replacement surgeries in aortic regurgitation lesion in patients with LVNC have been reported so far. All the above mentioned studies have not reported their difficulty in intraoperative and postoperative period. Our case added to the growing knowledge that these chronic secondary MR in LVNC can be operated with due risk if symptoms persist even after optimal medical therapy; intraoperatively we can overcome difficulty with the help of IABP. There are very limited number surgeries done on LVNC as a cause of severe secondary MR. Hence we don't now the troubles in surgeries and their long term outcome. Similary in postopertive period we can optimise the ionotropes and restart the anti-heart failure treatment followed by removal of iabp and managing arrhythmia meanwhile.

CONCLUSION

The LVNC is a relatively new entity of cardiomyopathy associated with left ventricular dysfunction, structural disease and arrhythmias. The imaging techniques play key role in diagnosis. The present case showed cardiac surgery done for valvular lesion in patient with LVNC improved the clinical condition and recovered cardiac function. Hence it adds the growing literature of surgery in LVNC.However there is limited evidence that mitral valve surgery improves survival in symptomatic patients with secondary MR. Surgery may improve symptoms and quality of life in these patients whose symptoms persist despite goal directed medical therapy while durability of the repair is dependent primarily on regression or progression of ventricular dilation.

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