Can left ventricular function recover following Bentall procedure in a patient with concomitant noncompacted left ventricular myocardium?

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Abstract

We report a rare case of successful left ventricular restoration of left ventricular hypertrabeculation/noncompaction (LVHT) after Bentall procedure in a patient with severe aortic regurgitation (AR) and aortic root aneurysm. At 1-year follow-up, he remained well with echocardiography showing the improved contractility of the noncompacted left ventricle. This case report emphasize that timely surgical correction of severe AR may also lead to improvement of systolic dysfunction along with concomitant LVHT.

Introduction

Left ventricular hypertrabeculation/noncompaction (LVHT) is a cardiac abnormality of unknown etiology, characterized by an increased number of trabeculations in the inner layer of the myocardium and a thinner than usual external compact myocardial layer. LVHT is found as a congenital abnormality but may be also acquired [1]. LVHT can present with normal ventricular function, as well as with both systolic and diastolic dysfunction [1]. There is no specific therapy for LVHT, while surgical treatment is focused on correction of any underlying congenital or adult abnormalities [1-3]. We report a rare case of successful left ventricular restoration of LVHT after Bentall procedure in a patient with severe aortic regurgitation (AR) and aortic root aneurysm.

Case report

A 54-years old Caucasian male patient with a body mass index 31.2 kg/m^2 with no history of congenital or adult cardiac illness presented with worsening exertional dyspnea. was referred to our institution because of diagnosed severe AR and aortic root aneurysm. He presented with worsening exertional dyspnea and had a history of chronic hepatitis C and unregulated arterial hypertension

Echocardiography demonstrated enlarged left ventricle with an end-diastolic diameter of 61 mm, left ventricle ejection fraction of 52%, and LVHT affecting the apex and lateral wall. Severe AR of a tricuspid aortic valve and an aortic root aneurysm were also detected. Computed tomography scan of aorta showed aortic root aneurysm with a diameter 6.81x6.20 cm (Figure 1). Coronary angiography detected a 40% stenosis of the left anterior descending artery.

The patient underwent a mediasternotomy. Cardiopulmonary bypass was initiated with cannulation of the ascending aorta and right atrium with mild hypothermia. Intermittent antegrade crystalloid cardioplegia (Custodiol) was delivered. Modified button Bentall procedure was performed using a mechanical valved conduit Carbomedics Carbo-Seal 25mm (LivaNova, London, United Kingdom). Intraoperatively, the trabeculations were confirmed by endoscopy (Figure 2) and as the ventricular function was preserved, it was decided to refrain from resection. The patient was weaned from CPB with mild inotropic support. Cross clamp time and CPB time were 88 and 128 minutes, respectively. The patient was transferred to the intensive care unit and extubated the same day. The patient was anticoagulated with warfarin on 4^{th} postoperative day following removal of chest drains. The postoperative course was uneventful and he was discharged after 11 days in a sinus rhythm.

At 1-year clinical follow-up, the patient was in New York Heart Association class I. Transthoracic echocardiography showed a well-seated mechanical aortic prosthesis, end-diastolic diameter of left ventricle of 43mm with ejection fraction of 66%.

Comment

In this rare case, we demonstrated that systolic dysfunction in LVHT can be reversible after concomitant severe AR is surgically corrected. The absence of a cardiomyopathy and good compliance with optimal medical treatment might have contributed to the recovery of systolic function. Recovery of left ventricular systolic function in LVHT following heart failure treatment has been previously described, although in different clinical scenarios [1, 4, 5].

It remains uncertain whether systolic dysfunction in the presented patient was related to LVHT, aortic regurgitation or longstanding unregulated hypertension. Most likely, all these factors together may have played a role. The mechanism of severe AR in our patient was distension of the aortic annulus due to ectasia of the ascending aorta while there was no structural valvular abnormality of the aortic valve [3]. Wilbring et al, have reported a patient suffering LVHT cardiomyopathy in combination with low-gradient aortic stenosis who underwent aortic valve replacement [6]. The authors emphasized that bad prognosis is primarily affected by impaired ventricular function, thromboembolic complications, and arrhythmias [6].

However, whether LVHT in our patient was congenital or acquired remains unclear, as he had no previous imaging to compare with. Despite management of LVHT has been described in a few case reports, further larger studies are needed to demonstrate whether LVHT can regress or not [4, 5]. Eurlings et al. have reported that LVHT may be either reversible or may have a dynamic course [4].

Surgical trabeculectomy has been reported rarely in LVHT and it was mostly performed in patients with isolated LVHT [7-9]. We refrained from trabeculectomy due to several reasons. Our patient did not have history of thromboembolic complications or arrhythmias. To our knowledge, there were only a few reports on surgical trabeculectomy, but none of these cases included a patient with severe AR. There was no evidence in the presented case that the trabeculations impeded diastolic filling of the ventricle, as observed in two out of three reported cases [8, 9].

In conclusion, timely surgical correction of severe AR may also lead to improvement of systolic dysfunction along with concomitant LVHT.

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Data Availability Statement

Data available on request due to privacy/ethical restrictions.

Patient Consent Statement

The patient agreed to the publication of this case report.

References

1. Finsterer J, Stöllberger C. Left Ventricular Noncompaction Syndrome: Genetic Insights and Therapeutic Perspectives. Curr Cardiol Rep 2020;22:84.
2. Barbukhatty KO, Boldyrev SY, Rossokha OA, et al. A rare case of coronary artery bypass grafting in a patient with left ventricular noncompaction. Ann Thorac Surg 2010;90:2047-2049.
3. Ohki S, Moriyama Y, Mohara J, et al. Aortic valve replacement for aortic regurgitation in a patient with left ventricular noncompaction. Ann Thorac Surg 2009;87:290-292.
4. Eurlings LWM, Pinto YM, Dennert RM, et al. Reversible isolated left ventricular non-compaction? Int J Cardiol 2009;136:e35–36.
5. Toyono M, Kondo C, Nakajima Y, et al. Effects of carvedilol on left ventricular function, mass and scintigraphic findings in isolated left ventricular non-compaction. Heart 2001;86:E4.
6. Wilbring M, Kappert U, Schön S, et al. Aortic valve replacement in noncompaction cardiomyopathy at two-year follow-up. J Card Surg 2009 Nov-Dec;24(6):684-6.
7. Shimamoto T, Marui A, Yamanaka K, et al. Left ventricular restoration surgery for isolated left ventricular noncompaction: report of the first successful case. J Thorac Cardiovasc Surg 2007;134:246-247.
8. Gan C, Hu J, Luo S, et al. Surgical restoration of left ventricular diastolic function: possible treatment for noncompaction cardiomyopathy. J Card Surg 2014;29:827-828.
9. Takamatsu M, Kamohara K, Sato M, et al. Effect of noncompacted myocardial resection on isolated left ventricular noncompaction. Ann Thorac Surg 2020;110:e387-e389.

Figure Legend

Figure 1

Computed tomography shows hypertrabeculation/noncompaction of the left ventricle and the dilated aorta (a). The aneurysmal ectasia of the ascending aorta increases in the supravalvular region (b).

Figure 2

Intraoperative videoendoscopy shows the noncompacted inner layer of trabecular meshwork with deep endocardial spaces of the lateral wall (a) and apex (b) of the left ventricle.