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Myocardial bridging associated with biventricular hypertrophic obstructive cardiomyopathy in the young adult patient: A case report

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ABSTRACT

Hypertrophic obstructive cardiomyopathy is a rare cardiac disease occurring in approximately 0.2% of the population and is usually left ventricular hypertrophy associated with or without mitral regurgitation due to SAM syndrome. Medical treatment, first septal ablation with absolute alcohol or septal left ventricular myectomy is the treatment methods for this disease. We report here the surgical results of a case of obstructive bilateral ventricular hypertrophy associated with a myocardial bridging of the left anterior descending artery. Surgical unroofing on the left anterior descending artery and septal myectomy were performed.

Keywords: Hypertrophic obstructive cardiomyopathy, SAM, myocardial bridging

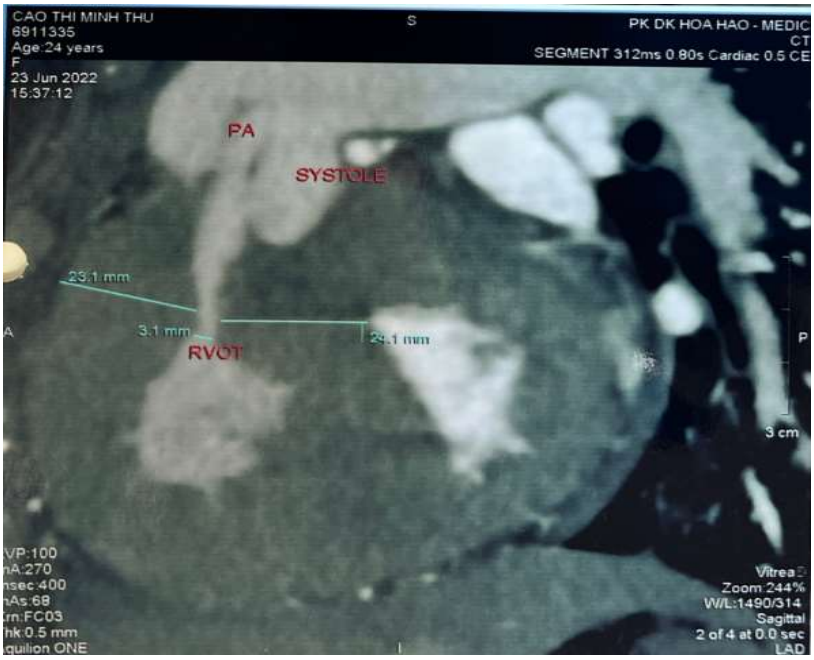
1. INTRODUCTION

Hypertrophic obstructive cardiomyopathy (HOCM) is an uncommon inherited heart disease that is characterized by cardiac hypertrophy, unexplained by abnormal loading conditions. Inherited HOCM is one of the leading causes of sudden cardiovascular death in patients under 35 years of age. The prevalence of the disease is estimated at about 1/500 of the population (Maron, 2002). There are at least 11 pathogenic mutations identified in more than 2000 positions, of which mutations on 4 genes MYH7, MYBCP3, TNNT2 and TNNI3 account for 80-90% of cases. Clinical manifestations are very variable, ranging from no symptoms or only dyspnea on exertion and chest pain to heart failure and sudden death. The guidelines in 2020 recommend surgical septal myectomy as the standard treatment in symptomatic HOCM for adult patients (Ommen et al., 2020). The disease mainly causes left ventricular hypertrophy with obstruction and often has mitral regurgitation due to systolic anterior motion (SAM) syndrome. Rarely, the disease has severe stenosis of both ventricular outflow tracts. We report a case of surgery in a patient with biventricular hypertrophy causing bilateral

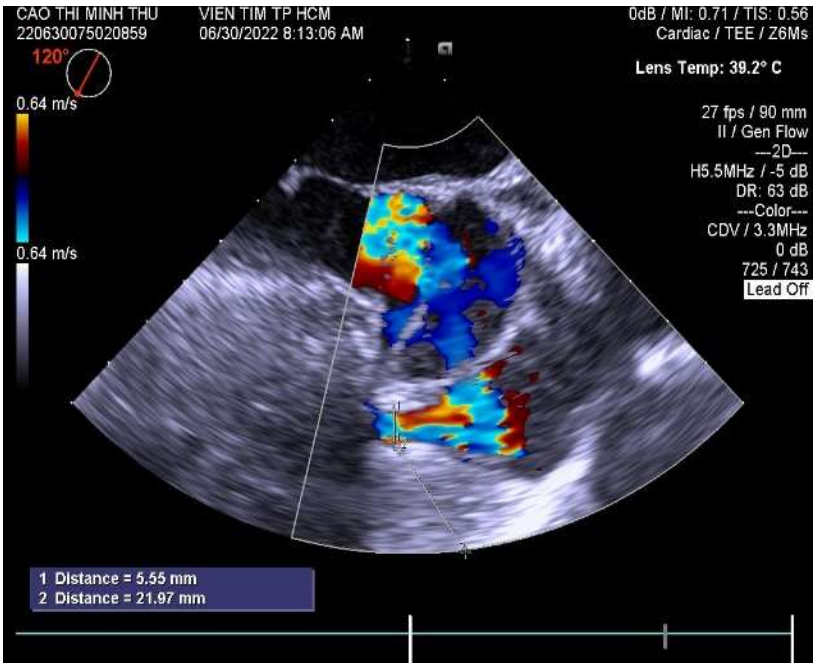
outflow tract obstruction associated with a muscular bridging of the left anterior descending artery.

2. CASE PRESENTATION

A 24-year-old girl, residing in the province, was hospitalized because of shortness of breath when lying down and chest pain (post-sternal pain lasting 30 minutes of attack), 2-3 attacks a day. In 2017, she was hospitalized with a diagnosis of pulmonary valve stenosis, subaortic stenosis and ventricular septal defect but refused treatment. The results of the cardiac ultrasound in June 2022 showed asymmetric left ventricular hypertrophy with a ventricular septal thickness of 28mm and a gradient max of 99 mmHg. Severe right ventricular hypertrophy also with V max= 5.4m/s. There has a diaphragm under the aortic valve with a diameter is 13mm, 6.6mm apart from the valve. Mild aortic and mitral regurgitation Transesophageal echocardiography (TEE) at surgery showed severe narrowing of both ventricular outflow tracts (Figures 1, 2). MSCT showed a clear image of muscular bridging 33.8mm long and 8.8mm deep of the left anterior descending artery (Figure 3). Holter ECG reported a sinus rhythm.



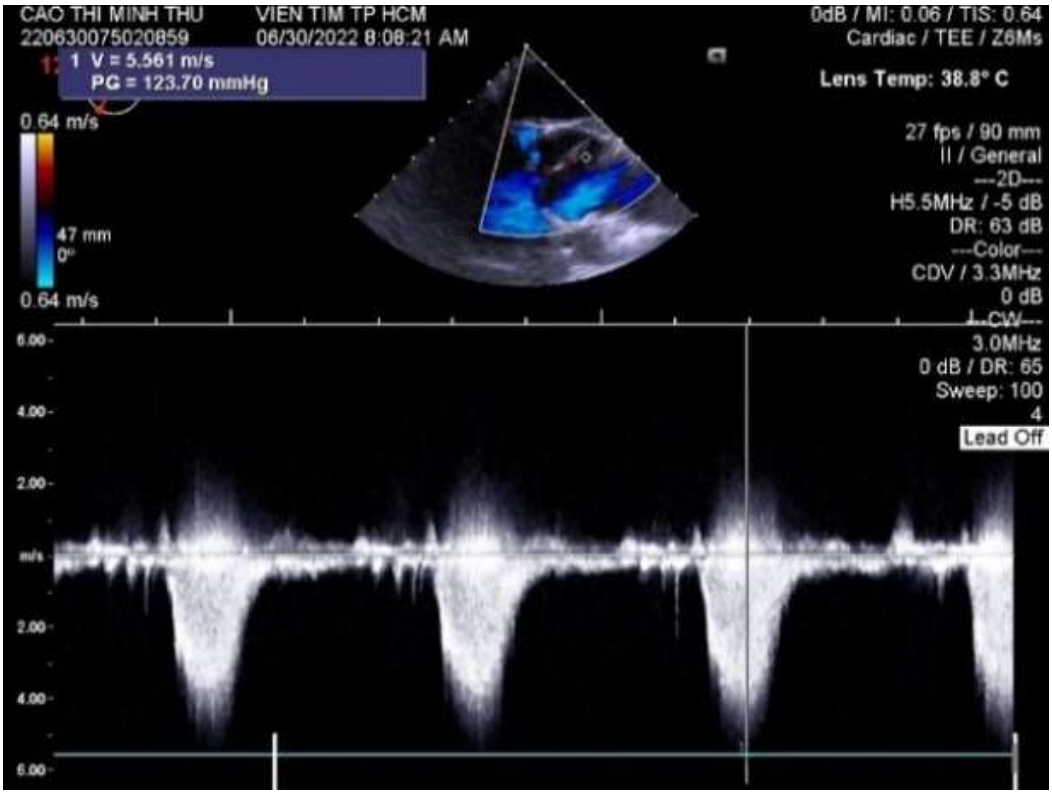
A



B

Figure 1 Severe stenosis of RVOT; 1A shown on MSCT and 1B shown on TEE (black arrow)

A



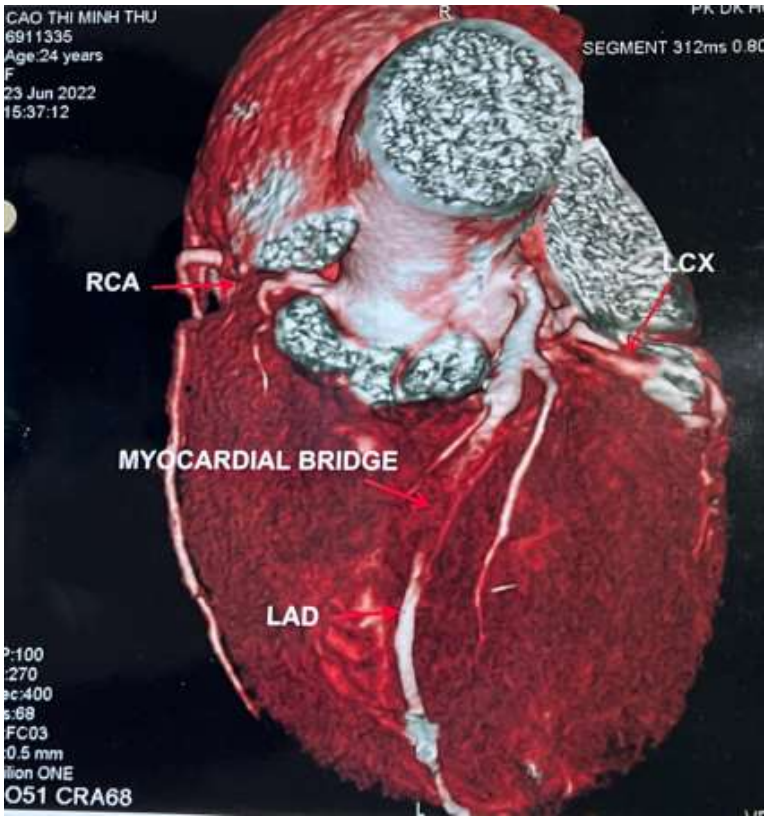
B

Figures 2A and 2B Severe stenosis of LVOT (shown on TEE)

She underwent unroofing myotomy (Figure 4) and resects the crest below the aortic valve (Figure 5). The modified Morrow procedure also was performed and wide excision of the trabeculae causes narrowing through the pulmonary artery for the right

ventricle (both cutting under guided by 3D TEE). After surgery, the maximum pressure gradient across both ventricles' outflow tract was significantly reduced (left ventricle: From 123 down to 15mmHg; right ventricle: From 90 down to 16mmHg). The patient was discharged after 12 days without any atrioventricular block.

A



B

Figures 3A and 3B An image of LAD myocardial bridging in MSCT

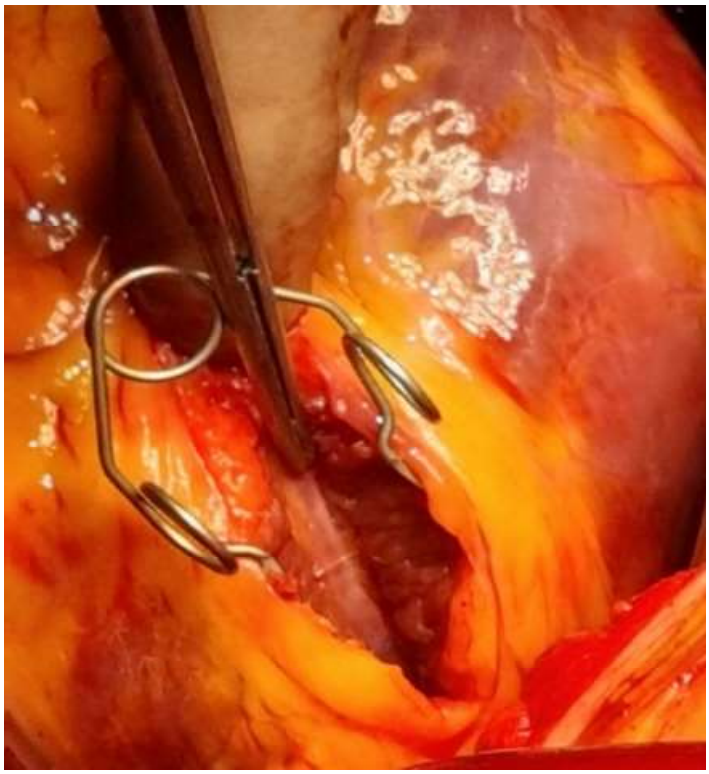


Figure 4 Unroofing myotomy LAD

Figure 5 Crest infra-aortic valve was resected

3. DISCUSSION

Bi-ventricular HOCM is rare, with only a few cases reported in the literature (Maron, 2018; Mozaffarian and Caldwell, 2001). If associated with the infravalvular crest and myocardial bridging it is even rarer. Our case is the most complicated and rare form due to the combination of all 3 lesions. In terms of diagnosis, cardiovascular MRI gives clear and beautiful diagnostic images but the

cost is still high (Ommen et al., 2020; Celenk and Celenk, 2022; Nguyen et al., 2022). Meanwhile, in our opinion, 2D and 3D TEE are very good means of both diagnosing and guiding intra-operative myomectomy and checking the effectiveness of surgery before weaning from extracorporeal circulation.

In terms of treatment, surgery gives better results than absolute alcohol first septal ablation by catheterization when compare with mortality, complications as well as long-term effectiveness (Wang et al., 2022; Seo et al., 2020; Olivotto et al., 2009; Hemmati et al., 2019). However, it is necessary to thoroughly resect and carefully evaluate the postoperative mitral regurgitation to ensure long-term results. When performing the modified Morrow's procedure, we favor doing a myectomy through both the aortic and anterior transverse mitral opening, depending on the thickness and length of the muscle to be resected. According to us, if only cut through the aortotomy it will be very difficult to cut all the muscle mass and sometimes cut into the mitral sub-valvular apparatus due to limited vision.

Regarding current medical treatment, in addition to beta-blockers, verapamil and antiarrhythmics, recently the FDA has approved the use of Mavacampten (Camzyos®) for patients with symptomatic obstructive hypertrophic cardiomyopathy with normal left ventricular function but not indicated for surgery.

4. CONCLUSION

The surgical unroofing myomectomy and biventricular septal resection are safe options for patients with HOCM with isolated myocardial bridging. However, it is necessary to follow closely the patient and warn them of the possibility of recurrent chest pain. They also need to continue with postoperative medications despite the reduction of coronary compression.

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Informed Consent

Informed Consent was obtained from the patient.

Author's contribution

All the authors contributed equally to the case report.

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Conflict of interest

The authors declare that there is no conflict of interests.

Data and materials availability

All data sets collected during this study are available upon reasonable request from the corresponding author.

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