Case Report

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Hypertrophic Obstructive Cardiomyopathy with Severe Aortic Valve Stenosis – A Case Report of a Commonly Overlooked Dual Pathology

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Abstract

We report acase of coexisting hypertrophic obstructive cardiomyopathy and aortic stenosis in a 69-year-old female who presented with exertional breathlessness and palpitations. Trans thoracic echocardiogram revealed severe valvular aortic stenosis and asymmetric septal hypertrophy with obstruction and mild to moderate mitral valve stenosis. The patient underwent concurrent aortic valve replacement with a tissue valve and extended septal myectomy. Post-surgery, the patient experienced significant symptom relief and improved functional status with no residual obstruction in the left ventricular outflow tract. This case highlights the necessity of assessing for sub-valvar obstruction while assessing valvular aortic stenosis.

Keywords: Aortic Valve Stenosis, Extended Septal Myectomy, Hypertrophic Obstructive Cardiomyopathy

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Introduction

Primary hypertrophic cardiomyopathy (HCM) co-existing with aortic valve stenosis (AVS) is rare.[1,3] Asymmetric septal hypertrophy is known to occur in patients with AVS, with some authors having reported the incidence to be up to 10%.[1–3] HCM and AVS cause left ventricular outflow obstruction with the prior causing dynamic while the latter causing fixed obstruction. Appropriate treatment of combined dual pathology significantly lowers mortality and improves long-term survival similar to regular age sex-matched individuals.[4] The management of HCM or AVS in isolation may be straightforward, however, when these entities occur together, it poses a challenge, more so in the era of increasing Transcutaneous Aortic Valve Implantation.[4,5] Herein, we present such a case and discuss its management.

Case report

A 69-year-old female presented with progressive exertional shortness of breath and palpitation for 6 months. She had been diagnosed with hypertension for 23 years, diabetes mellitus for 20 years, dyslipidemia for 15 years, and peripheral vascular disease with absent pulses in the right side below the popliteal arteries. Clinical evaluation revealed an audible ejection systolic murmur radiating bilaterally to the anterior neck. Her ECG showed sinus rhythm with right bundle branch block and features of left ventricular hypertrophy seen in HOCM, transthoracic echocardiography was significant for aortic valve stenosis with a peak pressure gradient of 75.7 mmHg and a mean gradient of 42.3 mmHg and an aortic valve area of 0.9 cm2, and a separate gradient across the left ventricle outflow tract (LVOT) of 90.7 mmHg (Figure 1), and a sigmoid-shaped interventricular

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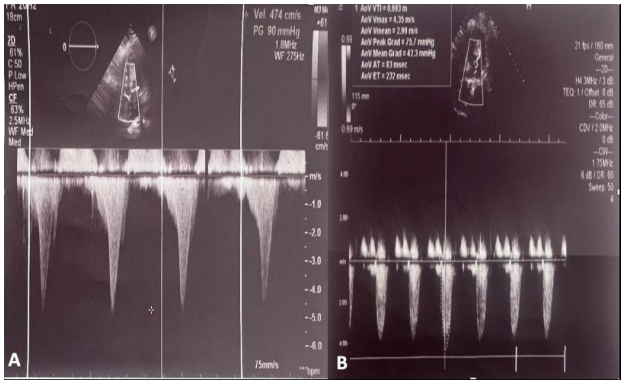

Figure 1. *2-Dimensional Transthoracic Echocardiogram showing gradients across (A) Left Ventricular Outflow Tract and (B) Aortic valve*

septum giving a shape of a dagger and asymmetric hypertrophy with a septal thickness of 1.64 cm in diastole and 1.82 cm in systole. (Figure 2) The mitral valve was found to be thickened with a mean gradient of 6 mm Hg and an area of 1.63 cm2 with a normal heart function. Pre-operative coronary angiography revealed a non-significant mid-LAD lesion which was documented to be 40%. She also had poor pulmonary reserve (FEV1 of 46%) and peripheral vascular disease with absent pulses on the right side below the popliteal arteries.

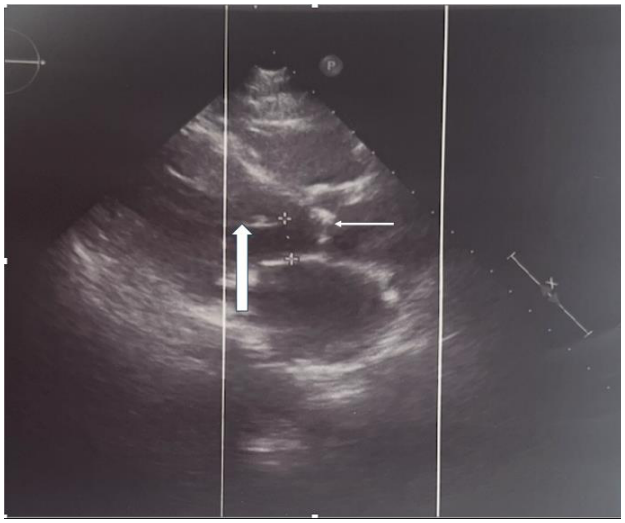

Figure 2. *Two-dimension Transthoracic Echocardiogram in parasternal long axis view showing hypertrophied IVS (thick arrow) causing obstruction to LVOT. Thickened and calcified aortic valve leaflets are also visualized. (thin arrow)*

She underwent aortic valve replacement with a 19-mm Edwards Inspiris Resilia bioprosthetic valve (Edwards Lifesciences, Irvine, CA, USA) and extended septal myectomy; with a total aortic cross-clamp time of 79 minutes and a total cardiopulmonary bypass time of 135 mins. Mitral valve leaflets were thickened but mobile with less than moderate obstruction on transesophageal echocardiogram and an intra-operative decision to not repair the valve was taken.

She was extubated on the day after surgery. She had complete atrioventricular dissociation post-operatively and was placed on epicardial pacing which was converted to a transvenous dual chamber pacemaker on the 7th postoperative day. She required a prolonged ICU stay of 10 days due to pulmonary issues and was discharged from the hospital on the 20th day.

She continues her regular follow-up and is in NYHA Class I status. Her follow-up transthoracic echocardiogram at 6 months showed a normal heart function with a normally functioning prosthetic aortic valve with a mean gradient of 8.2 mm Hg and no gradient across the LVOT.

Discussion

We present a case of combined HCM and AVS causing a fixed and dynamic obstruction to LVOT who underwent a concurrent procedure and improved clinically. Concentric left ventricular hypertrophy is a part of severe AVS, asymmetric hypertrophy of the ventricular septum can be an adaptation to chronically raised afterload or occur as a co-existent disorder of HCM.[6] Although symptomatic severe AVS is managed with surgical or transcatheter aortic valve replacement depending on the level of surgical risk[6], its occurrence with HCM in a patient poses diagnostic challenges. The detection of sub-valvular obstruction or abnormal anatomical variation in patients with AVS needs a high index of suspicion and a cautious echocardiography.[3]

HCM alone can be managed with surgical septal myectomy with excellent results,[7] however, when co-existent with severe AVS, valve replacement alone can lead to residual LV outflow obstruction and therefore needs to be combined with septal myectomy.[8] Correction of valvular stenosis without addressing septal hypertrophy may result in major postoperative morbidities and mortality.[3] To minimize or avoid residual obstruction, asymmetric septal hypertrophy with a sub valvular gradient above 30 mm Hg at rest or 50 mm Hg on exertion in a patient with symptomatic aortic stenosis, aortic valve replacement is recommended in addition to myectomy.[8] In the era of increasing transcutaneous aortic valve implantation (TAVI), the role of identifying the anatomy of sub-valvar structures and appropriate corrections is mandatory. There are case reports where patients have required emergency alcohol septal ablation following TAVR to minimize the effects of subvalvar obstruction.[9]

Preoperative right bundle branch block (RBBB) is a significant risk factor for the development of complete heart block (CHB), as LBBB is common after septal myectomy.[9] A large study by Cui and Schaff found that 34.8% of HOCM patients with preoperative RBBB who underwent transaortic septal myectomy developed CHB postoperatively. Our case had preoperative RBBB and developed CHB postoperatively necessitating insertion of a dual-chamber pacemaker.

Conclusion

This case highlights the importance of meticulously looking for sub-valvular obstruction in patients with aortic stenosis. It is important to address both valvular and septal pathology simultaneously to prevent or minimize post-operative morbidity and mortality from residual obstruction in patients with coexisting hypertrophic obstructive cardiomyopathy and aortic stenosis.

References

1. Nanda N, Gramiak R, Shah P, Stewart S, Deweese J. Echocardiography in the diagnosis of idiopathic hypertrophic subaortic stenosis co-existing with aortic valve disease. Circulation. 1974;50(4):752-757. doi:10.1161/01.cir.50.4.752.

2. Laskey WK, Kussmaul WG. Subvalvular gradients in patients with valvular aortic stenosis: prevalence, magnitude, and physiological importance. Circulation. 2001;104(9):1019-1022. doi:10.1161/hc3401.095041.

3. Ramamurthi A, Aker EM, Pandian NG. A case of aortic stenosis and hypertrophic cardiomyopathy. Echocardiography. 2012;29(10):1261-1263. doi:10.1111/j.1540-8175.2012.01812.x.

4. Desail MY, Alashi A, Popovic ZB, Wierup P, Griffin BP, et al. Outcome in patients with obstructive hypertrophic cardiomyopathy and concomitant aortic stenosis undergoing surgical myectomy and aortic valve replacement. J Am Heart Assoc. 2021;10(18):e018435. doi:10.1161/JAHA.120.018435.

5. Hess OM, Schneider J, Turina M, Carroll JD, Rothlin M, Krayenbuehl HP. Asymmetric septal hypertrophy in patients with aortic stenosis: an adaptive mechanism or a coexistence of hypertrophic cardiomyopathy. J Am Coll Cardiol. 1983;1(3):783-789. doi:10.1016/s0735-1097(83)80191-0.

6. Schaff HV, Juarez-Casso FM. Treatment strategies for hypertrophic cardiomyopathy: surgical. Am J Cardiol. 2024;212:S53-S63. doi:10.1016/j.amjcard.2023.10.053.

7. Shenouda J, Silber D, Subramaniam M, Alkhatib B, Schwartz RK, Goncalves JA, et al. Evaluation and management of concomitant hypertrophic obstructive cardiomyopathy and valvular aortic stenosis. Curr Treat Options Cardiovasc Med. 2016 Mar;18(3):17. doi:10.1007/s11936-016-0440-3.

8. Cui H, Schaff HV, Nishimura RA, Geske JB, Dearani JA, Lahr BD, et al. Conduction abnormalities and long-term mortality following septal myectomy in patients with obstructive hypertrophic cardiomyopathy. J Am Coll Cardiol. 2019 Aug 6;74(5):645-655. doi:10.1016/j.jacc.2019.05.053.

9. Quintar M, Villablanca P, Lee J, Wang DD, Frisoli T, O'Neill B, et al. Emergency alcohol septal ablation for shock after TAVR. JACC Case Rep. 2021;3(6):853-858. doi:10.1016/j.jaccas.2021.03.028.