Case Report

Discrete Subaortic Stenosis in an Elderly Patient

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Abstract
Discrete subaortic stenosis is an uncommon condition in the elderly population. Echocardiographic examination can be useful for diagnosis with high accuracy. We present a case report of an elderly female patient who presented with accelerated hypertension and was subsequently diagnosed with discrete subaortic stenosis on transthoracic echocardiography. A significant, non-dynamic, left ventricular outflow tract pressure gradient in the presence of aortic regurgitation should raise the possibility of this condition in patients of geriatric age group.

Keywords
Subaortic stenosis, aortic regurgitation, asymmetrical septal hypertrophy, discrete subaortic membrane, left ventricular outflow tract

Abbreviations
AR: Aortic regurgitation
ASH: Asymmetrical septal hypertrophy
AVR: Aortic valve replacement
DSS: Discrete subaortic stenosis
HCM: Hypertrophic cardiomyopathy
IVSEDD: Interventricular septal end diastolic dimension
LVOT: Left ventricular outflow tract
PWEDD: Posterior wall end diastolic dimension
SAM: Systolic anterior motion
SAS: Subaortic stenosis
SM: Discrete subaortic membrane
TEE: Transesophageal echocardiography
TTE: Transthoracic echocardiography

Introduction
Subaortic stenosis (SAS) is a form of left ventricular outflow tract (LVOT) obstruction, presenting in both the pediatric and adult population. SAS may be associated with other cardiac lesions. Despite being easily diagnosed using non-invasive modalities, limited studies on the disease course in adult population are available. We report a case of discrete subaortic stenosis (DSS) in an elderly female patient, incidentally diagnosed during echocardiographic examination.

Case Report
A female patient in her early 70s presented to the emergency department with complaints of 2 episodes of epistaxis over the previous 4 days, which had subsided spontaneously before presentation to the hospital. She also complained of shortness of breath (New York Heart Association class II) since past few weeks. Patient had a 15-year history of essential hypertension, for which she was on irregular medical therapy. There was no significant family history of cardiac ailments. At presentation, she was afebrile with a blood pressure of 180/120 mmHg and a heart rate of 90/min. Respiratory rate was 18/min with a resting oxygen saturation of 95% on room air. Cardiovascular examination was significant for a grade III/VI ejection systolic murmur at the base. Chest auscultation revealed fine crepitations at both the lung bases. Rest of the systemic examination was unremarkable.

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Electrocardiogram was suggestive of normal sinus rhythm and left ventricular hypertrophy with strain pattern (Figure 1). Troponin I level was raised: 27 ng/L (99th percentile of upper limit of normal: 14 ng/L), as was NT-proBNP: 4,009 pg/mL (normal <125 pg/mL). Transthoracic echocardiography (TTE) showed asymmetrical septal hypertrophy (ASH) — interventricular septal end diastolic dimension (IVSEDD) = 26 mm, posterior wall end diastolic dimension (PWEDD) = 15 mm, IVSEDD/PWEDD ratio of 1.7 (Figure 2A). There was no systolic anterior motion (SAM) of the mitral leaflets. A discrete subaortic membrane (SM) was visualized, arising from the interventricular septum (Figure 2B), 11 mm inferior to the annulus of the right coronary cusp of the aortic valve (Figure 3A). Peak and mean pressure gradient of 70 mmHg and 41 mmHg respectively was recorded across the LVOT, at the level of SM, with a peak velocity of 419 cm/s on continuous wave Doppler (Figure 3B). Significant flow turbulence was also noted. Aortic valve cusps appeared sclerosed with mild aortic regurgitation (AR) (Figures 4A and 4B). Mitral valve showed annular calcification with mild mitral regurgitation. The left ventricular ejection fraction was 60%, calculated by the Simpson method. There were no regional wall motion abnormalities. Grade I left ventricular diastolic abnormality was seen with an E/e´ratio <8. Right ventricular structure and function was found to be normal. Pulmonary artery systolic pressure was normal at 20 mmHg.

In view of the clinical presentation and imaging findings, patient was medically managed. She was referred to the cardiac surgery team for definitive therapy, which the patient wanted to get done at a later date.

Discussion

DSS has been reported in 8% to 10% of all pediatric LVOT obstruction patients, and 6.5% of adult congenital heart disease patients.1,2 Associated cardiac lesions include ventricular septal defect, coarctation of aorta, atrioventricular septal defects, patent ductus arteriosus, bicuspid aortic valve, and double outlet right ventricle. However, it is a rarely diagnosed entity in the elderly.3 Available evidence points toward an acquired structural defect at the level of LVOT, in a backdrop of genetic susceptibility.3 The morphological characteristics of the obstruction ranges from a slender fibrous/fibromuscular membrane to a thick tunnel stenosis at the LVOT.4 In the elderly, progression of DSS is slow, with frequent development of AR. In the largest study among 134 adult patients of DSS, AR was reported in 81% patients; however, significant AR (moderate to severe) was found in only 19%.2 Plausible mechanisms for development of AR include a direct injury to valve cusps by the turbulent jet flow and a direct involvement by the fibrous membrane adjacent to aortic valve.

Our patient had presented with accelerated hypertension and during echocardiographic examination, a significant LVOT gradient was found. Especially because of the presence of ASH, a preliminary diagnosis of hypertrophic cardiomyopathy (HCM) was considered. However, the continuous wave doppler tracing revealed an earlier peaking wave with no features of dynamic LVOT obstruction (absent SAM). A closer inspection of the TTE images revealed a discrete SM leading to flow turbulence and LVOT gradient.

![Figure 1. Electrocardiogram at Presentation Showing Left Ventricular Hypertrophy With Strain Pattern.](image-url)
The TTE findings coupled with no significant family history suggestive of HCM confirmed the diagnosis of DSS. TTE and transesophageal echocardiography (TEE) have high accuracy in diagnosing DSS. TEE is required in patients with limited TTE acoustic window and during pre/intraoperative assessment.

Surgical resection is the preferred therapy of DSS in adults. The timing of surgery has not been clearly established in view of the rarity of this condition and its slow progressive nature. Current guidelines recommend surgical resection in symptomatic patients with a mean pressure gradient across LVOT of ≥40 mmHg or severe AR. Surgery should also be considered in asymptomatic DSS patients with LV systolic dysfunction, marked left ventricular hypertrophy, and in patients with hypotensive response during exercise testing.

Concomitant aortic valve replacement (AVR) may be required depending on the severity of AR.

Early surgery has been suggested in view of progressive AR developing in DSS. However, in a study involving 149 adult patients of DSS, followed up for a median of 6.3 years, revealed that progression to moderate-to-severe AR was rare in these patients. Hence, the inclination towards early surgery for prevention of AR progression must be curtailed pending further long-term studies.

Recurrence rates following surgery has ranged from 16% in patients with membranous DSS to 46% in patients with tunnel stenosis. Increased rates of concomitant AVR has been reported in reoperation patients due to progression of AR. Our patient was referred for surgical resection because of her symptomatic status and characteristic echocardiographic findings.
Conclusion

We herein report a rare case of DSS, diagnosed in an elderly patient. Careful attention to the clinical presentation and echocardiographic examination, particularly the site and nature of LVOT gradient, is of paramount importance in coming to a final diagnosis. Due to the slow progressive nature of the condition, timing of surgery must consider the symptomatic status of the patient, severity of obstruction, left ventricular function, and associated aortic valve disease.

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Author Contributions

Dr Jaideep Dey, Dr Nadeem Akhtar, and Dr Vivek Shama were involved in concept/design and drafting of this manuscript. Dr Arif Mustaqueem and Dr Sameer Shrivastava provided critical revision and approval of the final manuscript for submission.

Declaration of Conflicting Interests

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