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Case Report

A case of syncope after exertion due to sigmoid septum evaluated by exercise stress echocardiography

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ABSTRACT

A sigmoid septum is associated with sharp angulation and aging of the aortic root; however, it does not affect the pressure gradient in the left ventricular outflow tract and is generally asymptomatic. This report describes a 73-year-old woman who presented with syncope after exertion. Echocardiography revealed that the cause was left ventricular outflow tract stenosis associated with a sigmoid septum; her symptoms improved with beta-blocker therapy. Exercise stress echocardiography was performed to determine treatment efficacy. Sigmoid septum causes syncope on exertion; however, drug therapy is effective. Exercise stress echocardiography is effective in determining treatment efficacy. If syncope is present, a sigmoid septum should be considered as a cause.

Learning objectives:

1. A sigmoid septum is part of or resembles hypertrophic cardiomyopathy, resulting in left ventricular outflow tract (LVOT) stenosis that is exacerbated by exertion and may cause syncope.
2. A sigmoid septum is a differential diagnosis for the cause of syncope and is diagnosed using cardiac echocardiography.
3. LVOT stenosis due to a sigmoid septum can be improved with drug therapy such as beta-blockers.
4. The effects of beta-blocker therapy can be determined by exercise stress echocardiography.

Introduction

A sigmoid septum is a condition in which the left ventricular outflow tract shows asymmetric focal protrusion of the myocardium resulting from an acute angulation of the aortic root due to aging; it does not cause an increased pressure gradient in the left ventricular outflow tract but is often asymptomatic and does not affect exercise capacity [1–3]. Reports of symptomatic sigmoid septum are rare, and there is no consensus on treatment for the condition [4]. A patient with a sigmoid septum fainted after exertion, and exercise stress echocardiography demonstrated that treatment with a beta-blocker was effective.

Case report

The patient was a 73-year-old woman who visited our hospital because of new-onset syncope after exertion. Chest auscultation revealed a Levine III/IV systolic murmur at the second right sternal border, leading to the suspicion of left ventricular outflow tract stenosis; an echocardiogram was performed. Echocardiography revealed hypertrophy of 18 mm confined to the septal base. The thickness of the anterior wall of the middle-left ventricle was 8 mm, the maximum flow velocity of the left ventricular outflow tract was 4.3 m/s, and the maximum pressure gradient was 74.2 mm Hg; moderate mitral regurgitation due to systolic anterior movement (SAM) of the mitral valve was observed (Fig. 1A, B, and C).

The patient was diagnosed with syncope due to left ventricular outflow tract stenosis caused by a sigmoid septum. Her symptoms dissipated following the administration of bisoprolol (1.25 mg/day). Subsequently, echocardiography was performed after drug treatment; the maximum flow velocity and pressure gradient of the left ventricular outflow tract were 1.5 m/s and 7.4 mm Hg, respectively, which were all

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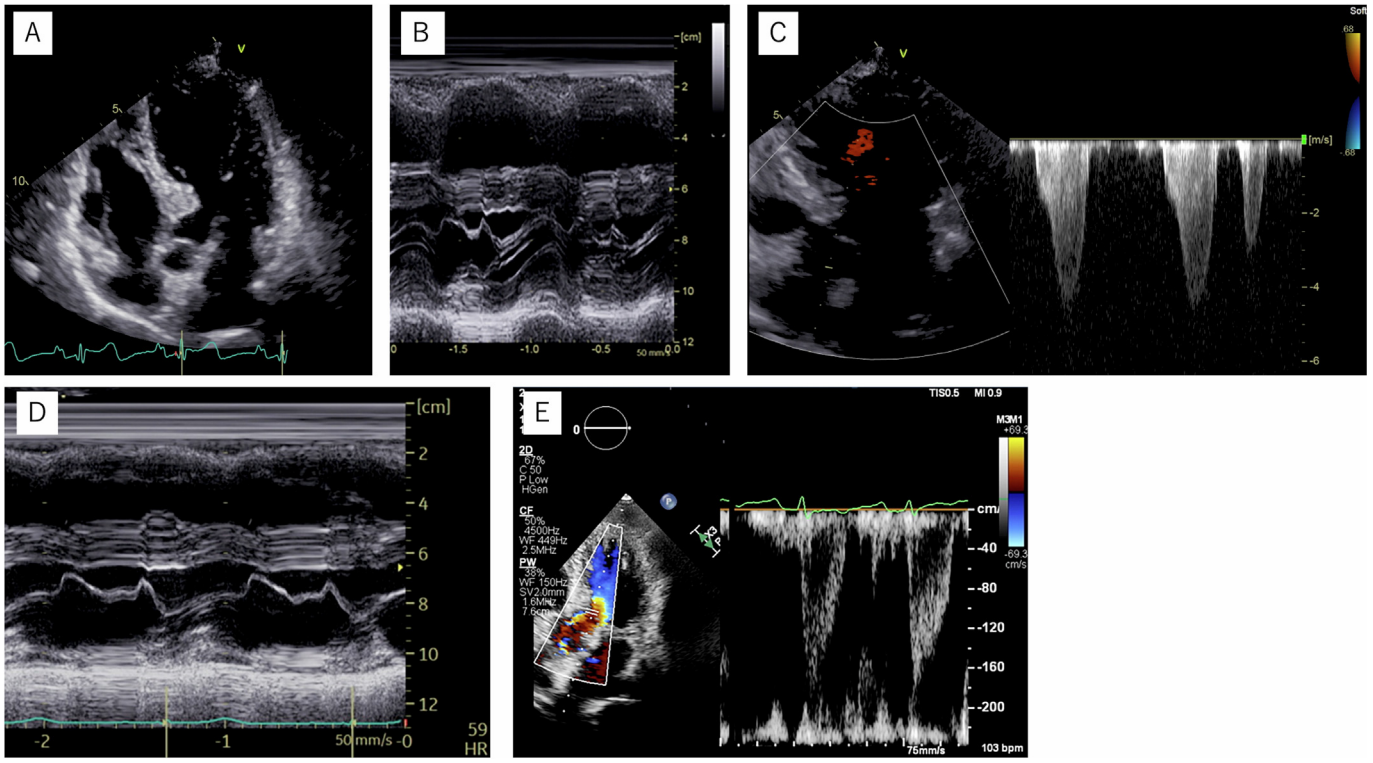

Fig. 1. Transthoracic echocardiographic images. (A) Echocardiography (B-mode) at admission. Echocardiography shows that the thickness of the anterior wall of the middle-left ventricle was 8 mm and the base of the septum was thickened to 18 mm. (B) Echocardiography (M-mode) at admission. Echocardiography shows the systolic anterior movement (SAM) of the mitral valve. (C) Echocardiography (continuous wave Doppler) at admission. Echocardiography shows an increase in the maximum left ventricular outflow tract flow velocity. (D) Echocardiography (M-mode) post-treatment. Echocardiography shows disappearance of the SAM of the mitral valve. (E) Exercise stress echocardiography (continuous wave Doppler) post-treatment. Exercise stress echocardiography shows no increase in the maximum left ventricular outflow tract flow velocity.

within normal limits. SAM was observed to have disappeared (Fig. 1D). Exercise stress echocardiography was performed to evaluate the effects of the drug during exertion. Exercise loading was performed using a treadmill [Bruce protocol Stage 3 (39 s); the maximal load was 10.2 METs]. No syncope attacks, abnormal blood pressure fluctuations, or electrocardiographic abnormalities were observed during stress. Echocardiography revealed no increased maximum left ventricular outflow tract flow velocity and maximum pressure gradient after maximal stress, with values of 1.8 m/s and 12.0 mm Hg, respectively. In addition, there was no exacerbation in the rest phase (Fig. 1E). After six months of follow-up, the patient did not experience post-exertional syncope.

Discussion

This case demonstrates two key points: 1) a sigmoid septum increases the pressure gradient in the left ventricular outflow tract, causing syncope after exertion, and 2) exercise echocardiography is useful for assessing drug effects.

A sigmoid septum can cause syncope after exertion. A sigmoid septum was previously defined as a proximal focal area (within the first third of total septal length) of localized septal hypertrophy with a dune-like structure protruding in the left ventricular outflow tract, a thickness ≥13 mm in men and ≥12 mm in women, and >50 % greater than the septal thickness at its mid-distal point [2]. In addition, a sigmoid septum is associated with sharper angulation of the aortic root and aging; the prevalence is from 1.5 to 7 %, and patients generally have a good prognosis [2,5]. Furthermore, a sigmoid septum does not increase the left ventricular outflow tract velocity [2,3]; however, cardiac loading on the sigmoid septum may be symptomatic. Previous studies have described dyspnea on exertion and syncope after drinking alcohol or nitrates [1,4,6]. In the present case, echocardiography at rest showed a thickening of the base of the left ventricular septum, an increased pressure gradient in the left ventricular outflow tract, and SAM of the mitral valve, morphologically suggesting hypertrophic obstructive cardiomyopathy as the differential diagnosis. However, a contrast-enhanced cardiac magnetic resonance imaging scan was performed and showed no evidence of heterogeneous myocardial hypertrophy or late gadolinium enhancement (LGE) findings. A sigmoid septum is recognized as a subtype of hypertrophic cardiomyopathy; however, there is a suggested possibility of it being an acquired, non-hereditary pathology [2,7]. Moreover, previous studies have indicated variations in the background characteristics of these patients (Table 1). Hypertrophic cardiomyopathy has been attributed to mutations in the sarcomere gene; the absence of LGE, as in this case, suggests that the sarcomere gene may not be mutated [8]. In addition, patients with sigmoid septum are older than those with hypertrophic cardiomyopathy [2]. Considering the absence of LGE and the advanced age, we have diagnosed this case as sigmoid septum rather than hypertrophic cardiomyopathy. When symptoms due to left ventricular outflow tract stenosis, such as unexplained shortness of breath, chest pain, and syncope, are suspected, echocardiography should be performed when considering the possibility that the symptoms are due to a sigmoid septum.

Patients with a sigmoid septum may present with symptoms such as syncope and dyspnea due to an increased left ventricular outflow pressure gradient caused by increased cardiac contraction with exertion, similar to obstructive hypertrophic cardiomyopathy. The surgical treatment of patients with subjective symptoms of a sigmoid septum has been investigated but has not been considered effective [9]. Percutaneous septal myocardial ablation improves shortness of breath during exertion [10]. However, some reports of improvements with beta-

Table 1
Characteristics of a sigmoid septum in previous studies.

Author	Study population	Prevalence rate of sigmoidal shape	Age (years)	Male	Hypertension
Canepa et al. [1]	700 healthy adults Male 350 (50 %) Age 64 ± 15 years	49 (7 %)	76 ± 11	SiS: 50 % Others: 57 % NS	SiS: 55 % Others: 37 % $p = 0.01$
Binder et al. [7]	382 patients with HCM Male 210 (55 %) Age 41.6 ± 19.0 years	181 (47 %)	48.5 ± 17.0	SHCM: 102 (56 %) Others: 108 (54 %) NS	SHCM: 33 (18 %) Others: 19 (9 %) $p = 0.02$
Neubauer et al. [8]	2755 patients with HCM. Male 1853 (71 %). Age 49 ± 11 years	1197 (46 %)	51.6 ± 10.3	SHCM: 839 (70 %) Others: 1032 (72 %) NS	SHCM: 504 (42.2 %) Others: 451 (31 %) $p < 0.0001$

All studies were diagnosed by echocardiographic or magnetic resonance imaging morphologic evaluation. Patients with a sigmoid septum were relatively elderly compared with sigmoidal hypertrophic cardiomyopathy patients. There was no significant difference in sex by sigmoidal shape in either literature in the healthy adults; there was no association between sigmoid septum and hypertension. However, among the patients with hypertrophic cardiomyopathy, there was a significant increase in the prevalence of hypertension among those exhibiting a sigmoidal shape.
HCM, hypertrophic cardiomyopathy; SiS, sigmoid septum; NS, not significant; SHCM, sigmoidal hypertrophic cardiomyopathy.

blockers and/or cibenzoline have been published. Considering the degree of invasiveness of the procedure, drug therapy should be considered first [1,4,6]. In this case, syncope did not recur after the administration of beta-blockers.

Exercise stress echocardiography could be useful for evaluating the efficacy of medications in patients with a sigmoid septum. The treatment of symptomatic patients with a sigmoid septum includes treating the underlying disease, percutaneous septal muscle ablation, beta-blockers, and/or cibenzoline. Although subjective symptoms improved with all these procedures, there has been no clear evaluation of the treatment efficacy, and no method has been established to determine this. Furthermore, the effects of pharmacological treatments following cardiac stress remain unknown. In this study, exercise stress echocardiography, which can apply physiological cardiac stress, showed no increase in the left ventricular outflow tract pressure gradient or systolic forward motion of the mitral valve under cardiac stress and no syncope symptoms, indicating the effectiveness of the drug therapy. In cases such as this, where there is a possibility of a serious outcome such as syncope, it is important to determine the efficacy of drug therapy using exercise stress echocardiography.

Conclusions

A sigmoid septum can cause syncope after exertion, and the administration of beta-blockers improves subjective symptoms such as syncope. Resting echocardiographic findings and exercise stress echocardiograms are useful for assessing drug effects.

Consent statement

Informed consent was obtained from the patient for the publication of the case and accompanying images.

Declaration of competing interest

The authors declare that there is no conflict of interest.

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