CASE REPORT

Significant left ventricular outflow tract obstruction secondary to systolic anterior motion in a patient without hypertrophic cardiomyopathy: An echocardiographic study

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Dynamic left ventricular outflow tract obstruction (LVOTO) can be hemodynamically significant and can adversely affect the heart and quality of life. It is caused by systolic anterior motion (SAM) of the anterior mitral valve into the LVOT. The mechanism underlying SAM has been an area of special interest. However, SAM occurrence in the absence of septal hypertrophy is exceedingly uncommon. Here we present a case of a young male patient who sought medical care with a complaint of exertional dyspnea, New York Heart Association functional Class 2–3, and was found to have SAM and severe LVOTO at rest without hypertrophic cardiomyopathy. Continuous wave Doppler signal showed a peak velocity of 4.96 m/s along the LVOT, with a pressure gradient at rest of 98.44 mmHg, calculated using the modified Bernoulli equation. The patient is not known to have any medical conditions, nor had a family history of cardiac condition or sudden death. Trans-thoracic echocardiography showed concentric remodeling of the LV without hypertrophy. Trans-esophageal echocardiography was performed for further assessment of the anatomy. The anterior mitral leaflet (AML) and posterior mitral leaflet (PML) lengths were 3.7 cm and 1.3 cm, respectively (normal AML < 3 cm; normal PML < 1.5 cm). In our patient, the LVOTO is significant enough to result in a decreased cardiac output, which explains the symptoms experienced, due to which he developed concentric remodeling. The only finding in this patient explaining SAM is an elongated AML.

Keywords: Echocardiography, Left ventricular outflow obstruction, Systolic anterior motion

1. Introduction

Dynamic left ventricular outflow tract obstruction (LVOTO) can be hemodynamically significant and can adversely affect the heart and quality of life. It is caused by systolic anterior motion (SAM) of the anterior mitral valve into the LVOT. The mechanism underlying SAM has been an area of special interest. Hypertrophic cardiomyopathy (HCM) is the most prevalent cause of SAM of the mitral valve and LVOTO [1]. While

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several theories have been proposed to explain the occurrence of SAM and LVOTO, the 'drag effect' theory is widely accepted [2]. Occurrence of SAM in the absence of septal hypertrophy is exceedingly uncommon. In this report, we present a case of a young male patient who sought medical care with a complaint of exertional dyspnea and was found to have SAM and severe LVOTO at rest without HCM.

2. Case report

A 38-year-old male patient was referred to the cardiology department at our institute with a complaint of exertional shortness of breath, New York Heart Association functional Class 2–3, with no orthopnea or paroxysmal nocturnal dyspnea. The patient is not known to have any medical conditions; besides, there was no family history of cardiac condition nor sudden death. Various investigations were performed, including electrocardiogram and trans-thoracic echocardiography (TTE), which showed concentric remodeling of the left ventricle without hypertrophy and the following measurements: interventricular septal thickness, 1.2 cm; posterior wall thickness, 1.2 cm; left ventricular internal diameter end diastole, 4.5 cm; relative wall thickness, 52%; left ventricle mass, 197.99 g; left ventricle mass index, 95.65 g/m^2 (Fig. 1A). SAM of the anterior mitral leaflet (AML) was noted (Fig. 1B) with prominent flow acceleration using color Doppler. Severe LVOTO at rest was also noted, with a continuous wave Doppler signal showing a peak velocity of 4.96 m/s along the LVOT and pressure gradient at rest of 98.44 mmHg, which was calculated using the modified Bernoulli equation (Fig. 1C). The patient had a normal left ventricular ejection fraction of 70% with no regional wall motion abnormalities. Mild posteriorly directed jets of mitral regurgitation were also identified. Mid-left ventricular parasternal short-axis view confirmed the normal position of the papillary muscles (PMs) and a PM diameter of 1.1 cm. Trans-esophageal echocardiography (TEE) was performed for further assessment of the anatomy. The AML and posterior mitral leaflet (PML) lengths were 3.7 cm and 1.3 cm, respectively (normal AML < 3 cm; normal PML < 1.5 cm). The AML-to-PML ratio was 2.8 (Fig. 2A) and the LVOT diameter was 2.2 cm. The distance from the mitral valve coaptation point to the septum was 1.5 cm, protrusion height (calculated from the mitral annular plane to the most protruding mitral leaflet tip) was 2 cm (Fig. 2B), residual length (calculated as the uncoapted portion that protrudes distally beyond the coaptation point) was 0.6 cm, and mitral aortic angle was 110°. The patient was started on a β-blocker, which improved his symptoms.

3. Discussion

SAM of the mitral valve can be classified into valvular and chordal. Chordal SAM is hemodynamically insignificant [1]. Hypertrophic obstructive cardiomyopathy (HOCM) is the commonest cause of SAM [1]. Several mechanisms have been suggested to explain the occurrence of SAM. Classically, in HOCM, it is assumed that as the velocity of the blood accelerates through the LVOT, pressure decreases, a phenomenon termed the 'Venturi effect'. The AML will then be sucked into the lower-pressure LVOT, which causes the obstruction, but this presents only a small part of the story. The septal hypertrophy seen in HOCM will affect the ejection flow stream of the left

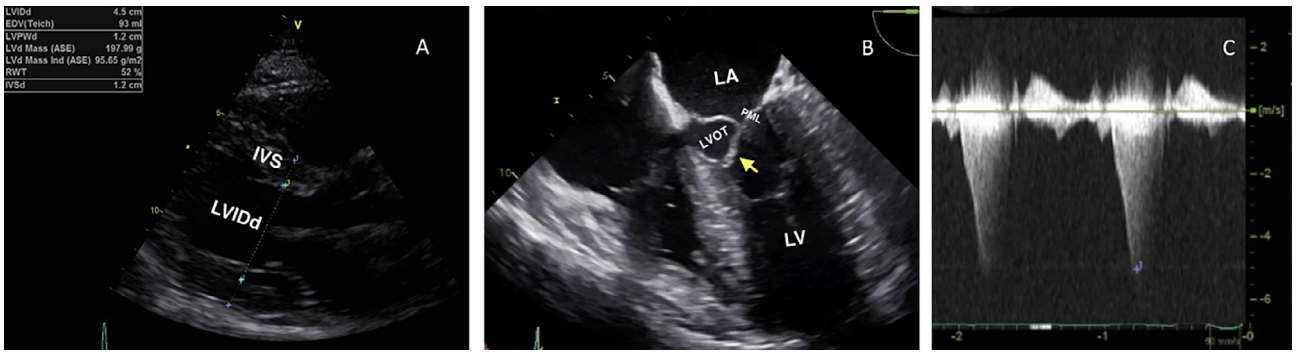

Figure 1. Echocardiography showing SAM. (A) Trans-thoracic echocardiographic imaging in the parasternal long-axis view showing concentric remodeling of the left ventricle (LV). (B) Transesophageal echocardiography. Mid-esophageal four-chamber view showing a systolic anterior motion of the anterior mitral leaflet (yellow arrow) into the left ventricular outflow tract (LVOT) causing severe LVOT obstruction at rest with gradient of 98 mmHg by continuous wave Doppler (C). IVS = interventricular septum; LA = left atrium; LVIDd = left ventricular internal diameter end diastole; PML = posterior mitral leaflet.

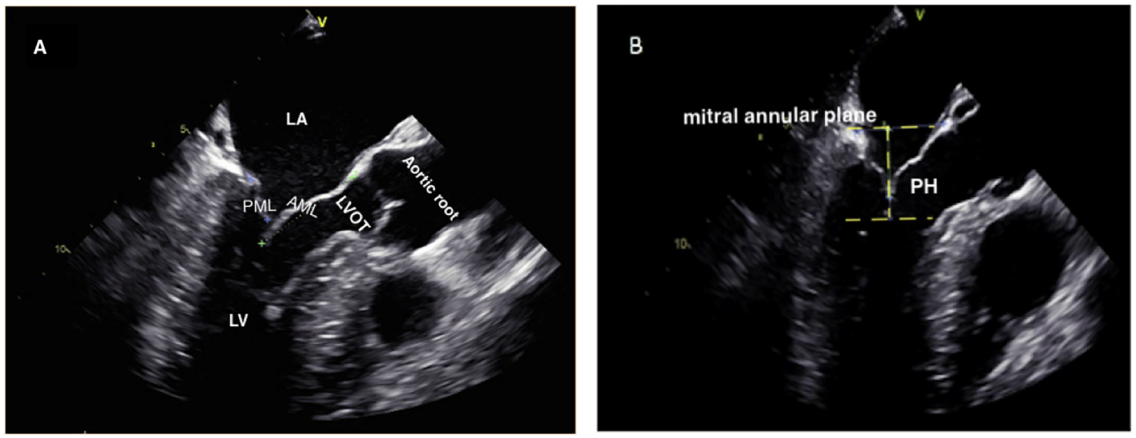

Figure 2. Measurements of the mitral annulus. (A) Trans-esophageal echocardiography (TEE) mid-esophageal long-axis view at an angle of 141° showing an elongated anterior mitral leaflet (AML; green dotted line) of 3.7 cm and posterior mitral leaflet (PML; blue dotted line) of 1.3 cm. (B) TEE at an angle of 141° at the beginning of coaptation in systole showing the protrusion height (PH). LA = left atrium; LV = left ventricle; LVOT = left ventricular outflow tract.

ventricle and redirects it. Consequently, the flow stream comes from a more posterior direction and hits the AML, thereby pushing it into the LVOT [3,4]. Addressing this by myomectomy alone did not relieve SAM in all patients because there are other characteristic anatomic features found in HOCM, such as mitral leaflet elongation and anterior displacement of the PM [3]. Some features have been suggested to be associated with SAM in the absence of septal hypertrophy, such as PM hypertrophy, an anterior fusion of the PM, the presence of accessory PM, and mitral annular calcification [2]. Although our patient did not have any of these features, he had an elongated AML (defined as >3 cm), which can protrude along the direction of the ejection flow stream of the left ventricle causing the obstruction [2]. In our case, the LVOTO is significant enough to result in a decreased cardiac output, thereby explaining the symptoms experienced, due to which the patient developed concentric remodeling. The only finding in this patient explaining SAM is an elongated AML. Uematsu et al. [1] compared HCM associated with SAM to non-HCM SAM, and concluded that the latter group was older, had a high prevalence of sigmoid septum, and their LVOT pressure gradients were lower. However, in our case, the patient was young, with no sigmoid septum, and a significant LVOT pressure gradient of 98.44 mmHg. In another study, Halpern et al. [3] analyzed the echocardiographic imaging before and after the resect–plicate–relea se operation on patients with HOCM, and on those with a septal thickness equal to or less than 1.8 cm, in whom the obstructive mechanisms depended more on the mitral apparatus abnormalities, rather than on septal thickness [3]. Thus, surgically addressing anomalies of the mitral apparatus is particularly crucial for those group of patients because they are involved in the pathophysiology of obstruction more than the modest septal thickening [4]. Leaflet plication was performed on patients with AML length more than 3 cm, which shortened and stiffened the leaflet, and decreased the protrusion height, thus alleviating the symptoms [3]. Similarly, we believe that plication of the AML in our case is a valid treatment option in reliving LVOT obstruction.

Table 1. Risk factors for systolic anterior motion.

• Anterior mitral leaflet >3 cm
• Anterior displacement of the papillary muscle (PM)
• PM hypertrophy
• Presence of accessory PM
• Mitral annular calcification
• Sigmoid septum

4. Conclusion

SAM pathophysiology is better understood now than before. It is a multifactorial mechanism with recognized risk factors (Table 1). Therefore, comprehensive assessment of these factors with TTE, TEE, or cardiac magnetic resonance imaging is essential. In our case, elongated AML alone produced SAM with significant obstruction.

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