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Significant left ventricular outflow tract obstruction secondary to systolic anterior motion in patient without hypertrophic cardiomyopathy, an echocardiographic study

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Introduction:

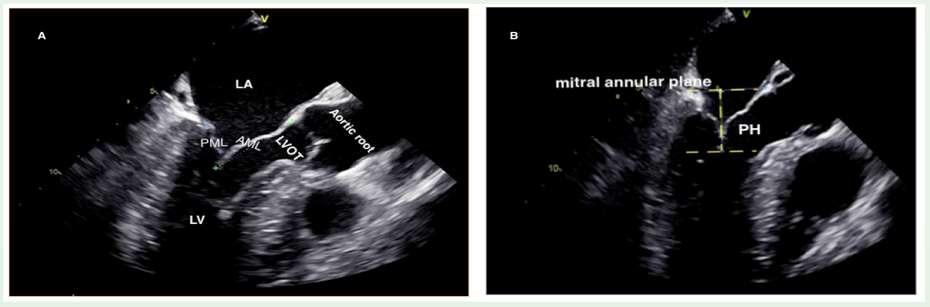
Dynamic left ventricular outflow tract obstruction(LVOTO) can be hemodynamically significant and affect adversely the heart and the quality of life. Systolic anterior motion (SAM) of the anterior mitral valve into the LVOT is the mechanism. However, mechanism of the SAM has been an area of special interest. Hypertrophic cardiomyopathy(HCM) is the most prevalent cause of systolic anterior motion (SAM) of the mitral valve, and left ventricular outflow tract obstruction (LVOTO)[1] . Several theories have been suggested to explain the occurrence of SAM and LVOTO, drag effect is a more accepted one.[2] SAM occurrence in the absence of septal hypertrophy is exceedingly uncommon. Herein, 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.

Case presentation:

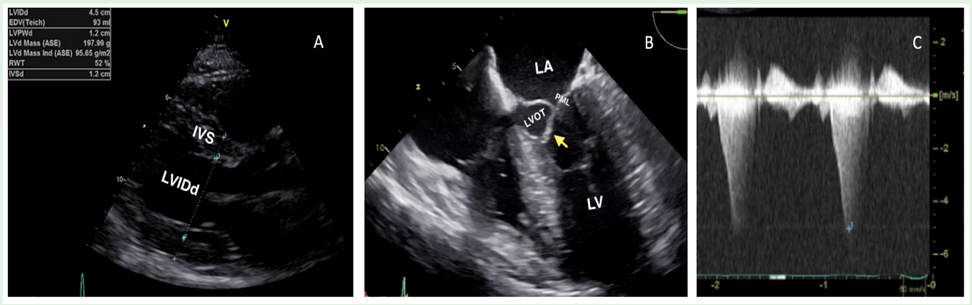
A 38-year-old male patient was referred to the cardiology department at our institute with a complaint of exertional shortness of breath, New York Heart Association (NYHA) functional class 2-3, with no orthopnea or paroxysmal nocturnal dyspnea. The patient is not known to have any medical conditions,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

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 was thought the as the velocity of the blood accelerates through the LVOT, pressure decreases, a phenomenon known as the Venturi effect, then the anterior mitral leaflet will be sucked into the lower pressure LVOT and causes obstruction, but this tells only a small part of the story. The septal hypertrophy seen in HOCM will affect the ejection flow stream of the left ventricle, redirecting it, so it comes from a more posterior direction, resulting in hitting the AML and pushing it into the LVOT. [3,4] Addressing this by myomectomy alone did not relieve SAM in all patients since there are other characteristic anatomic features found in HOCM such as mitral leaflet elongation, and anterior displacement of the papillary muscle. [3] Others have suggested some features to be associated with SAM in the absence of septal hypertrophy, such as papillary muscle(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 more than 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, and as a consequence the patient developed concentric remodeling. The only finding in this patient explaining SAM is an elongated anterior mitral leaflet . In a study done by Shoko Uematsu, et al. comparing between HCM associated with SAM and non-HCM SAM, concluded that the latter group was older, had a high prevalence of sigmoid septum, and their LVOT pressure gradients were lower. [1] However, in our case, the patient was young, with no sigmoid septum, and a significant LVOT pressure gradient of 98.44 mmHg. Another study conducted by Dan G. Halpern, et al. analyzed the echocardiographic imaging before and after the resect-plicate-release operation done for patients with HOCM, and on patients 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 since 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 plication of the AML in our case is a valid treatment option in reliving LVOT obstruction.



(Figure 2) A- TEE mid-esophageal long axis view at an angle of 141° showing an elongated anterior mitral leaflet (green dotted line) of 3.7 cm and posterior mitral leaflets (blue dotted line) of 1.3 cm. **B-** TEE at an angle of 141° at the beginning of coaptation in systole showing a protrusion height (PH).



(Figure 1) A- Trans-thoracic echocardiographic (TTE) imaging in the parasternal long axis view showing concentric remodeling of the left ventricle (A). Transesophageal Echocardiography (TEE) Mid-esophageal four-chamber view showing a systolic anterior motion of the anterior mitral leaflet (AML) (yellow arrow) into the left ventricular outflow tract (LVOT) causing severe left ventricular outflow tract obstruction (LVOTO) at rest with gradient of 98 mmHg by continuous wave Doppler (C).

Table 1: Risk factors for Systolic Anterior Motion:

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

Conclusion:

SAM pathophysiology is more understood now than before. It is a multifactorial mechanism with recognized risk factors (table 1). Comprehensive assessment of these factors with TTE, TEE, or cardiac magnetic resonance imaging is essential. In our case, elongated anterior mitral leaflet alone produced SAM with significant obstruction.

References:

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