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Dynamic Left Ventricular Outflow Tract Obstruction–when to Intervene?

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Abstract

Left ventricular outflow tract obstruction (LVOTO) in hypertrophic cardiomyopathy (HCM), most commonly is provoked by the contact between the hypertrophied basal interventricular septum (IVS) and the systolic anterior motion (SAM) of the anterior leaflet of mitral valve, during systole, thus narrowing the left ventricular outflow tract (LVOT). Several theories have been proposed to explain the occurrence of SAM and LVOTO, the "drag effect" theory is widely accepted.

Despite SAM, one of the others morphological features that can contribute to LVOTO is an insertion of an accessory muscle bundle extending from the apex to the basal anterior septum of free wall of the left ventricle.

In this case report we present a case of 71-year-old man with dyspnea and syncope, exercise induced, as a result of severe dynamic LVOTO. The LVOTO was a result of HCM, mostly affecting the basal IVS, with concomitant insertion of an accessory muscle bundle at the basal segment of IVS, that was additionally thickening the IVS, and SAM of the anterior mitral lealflet (AML), that were narrowing the LVOT and causing high LVOT gradients (86,3 mm Hg) at rest. The patient was symptomatic, he had dyspnea and syncope, exercise induced. The patient underwent a septal myectomy and mitral valve repair, which successfully reduced the gradients and relieved the patient of the symptoms.

Keywords: accessory muscle bundle, hypertrophic cardiomyopathy; LVOT obstruction, syncope; systolic anterior motion;

Introduction

Left ventricular outflow tract obstruction (LVOTO) is one of the reasons for syncope in patients with hypertrophic cardiomyopathy (HCM). Left ventricular outflow tract (LVOT) obstruction in HCM is provoked by the contact between the hypertrophied basal interventricular septum (IVS)

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and the systolic anterior motion (SAM) of the anterior leaflet of mitral valve, during systole, thus narrowing the LVOT. There are different theories that are explaining the occurrence of SAM and LVOTO, and the "drag effect" theory is widely accepted. A resting or provocable peak gradient of 50 mmHg or higher is considered the threshold for septal reduction procedures in symptomatic patients. (1)

Despite SAM, one of the others morphological features that can contribute to LVOTO is an insertion of an accessory muscle bundle stretching from the apex to the basal anterior septum of free wall of the left ventricle (2).

Syncope and pre-syncope occur in 15-25 % of the patients with HCM (3). Of exceptional importance is to detect the reason for syncope, and if treatable, to resolve it. Surgery reduces the symptoms of syncope and pre-syncope and is effective in more than 80% of patients. (4)

In this report we present a case of severe dynamic LVOTO as a result of HCM, affecting mostly the basal IVS, with concomitant insertion of an accessory muscle bundle at the basal segment of IVS, additionally thickening the IVS, and SAM of the anterior mitral leaflet (AML), narrowing the LVOT and causing high LVOT gradients at rest. The patient underwent a septal myectomy and mitral valve repair that successfully reduced the gradients and relieved the symptoms.

Case presentation

A 71-year-old man presented in an emergency room complaining of dyspnea and syncope, exercise induced. Dyspnea induced by physical activity was recurring over the past two years, worsening over the time and in the last 4 months was more intense, after COVID 19 infection. The syncope recurred during physical activity twice. From past medical history the patient has only hypertension grade 1, which was well regulated with medication. On physical examination the patient had systolic murmur at the left sternal edge that radiates to the right upper sternal edge and apex. ECG revealed signs of left ventricular hypertrophy.

The two-dimensional transthoracic echocardiography (TTE) revealed asymmetric hypertrophy of the IVS, with sigmoid septum with thickness of 19 mm at the beginning of the basal anterior septum that bulges into the left ventricular (LV) cavity. At the basal segment of IVS an insertion of an accessory muscle bundle was also revealed, that additionally was thickening the basal IVS (Figure 1 and 2). The remaining segments of IVS were slightly thickened, 15 mm in diastole, the posterior wall (PW) thickness was 13 mm and the septal/ PW thickness ratio was 1.4, in addition to HCM. Resting SAM of the AML was present, that resulted with LVOTO (Figure 3).

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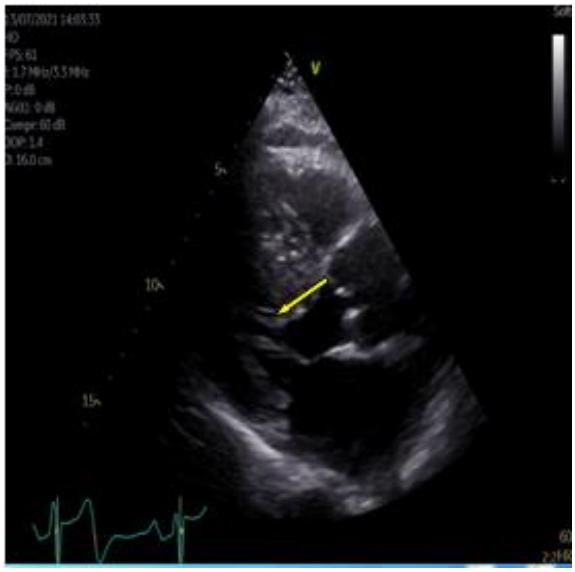

Figure 1

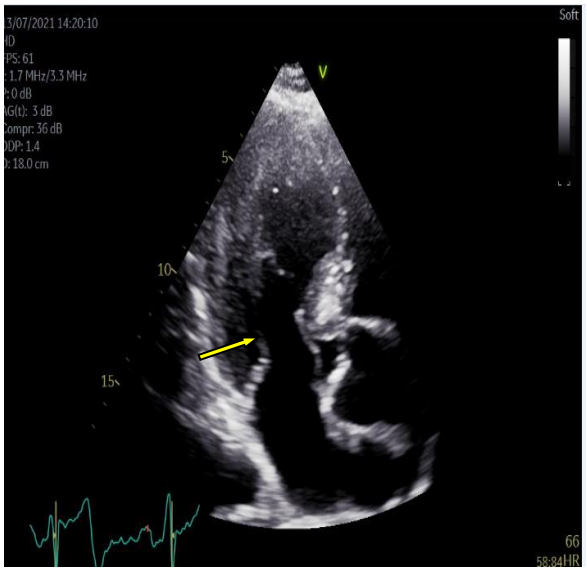

Figure 2

Figure 1. Two-dimensional PLAX view of sigmoid, hypertrophied septum and insertion of an accessory muscle bundle (yellow arrow).

Figure 2. Hypertrophy of the basal IVS on 3C view in which position LVOTO is better seen (yellow arrow)

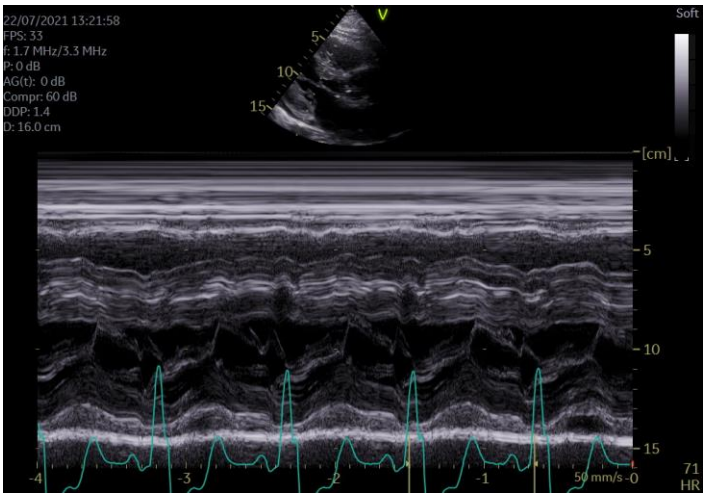

Figure 3. M-mode echocardiography showing SAM of the AML

By continuous wave (CW) Doppler, LVOT maximum velocity was 4,1 m/s and peak pressure gradient of 86,3 mmHg was measured, with "dagger-shaped" envelope (Figure 4). These velocities were measured at rest. There was also a SAM-related, functional, eccentric, moderate mitral regurgitation due to "coanda effect", with maximal flow velocity at the late systole (Figure

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5). Elevated left ventricular filling pressure was also measured, with grade I diastolic dysfunction. Dilatation of the ascending aorta up to 51 mm and mild aortic regurgitation was also recorded (Figure 6)

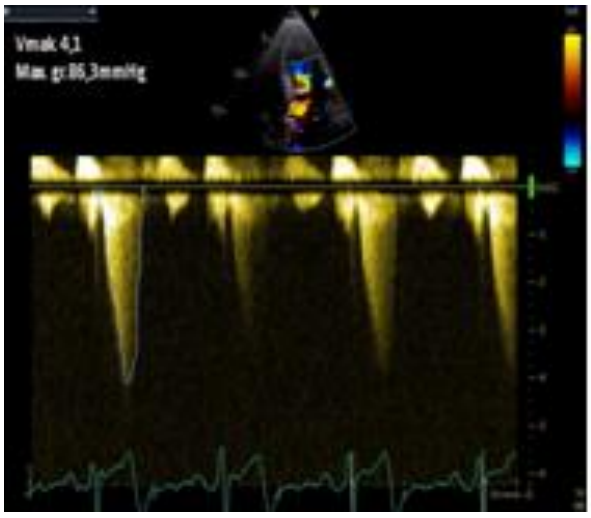

Figure 4. CW Doppler with sample volume at LVOT which shows LVOT maximum velocity 4,1 m/s with "dagger-shaped" envelope and peak pressure gradient of 86,3 mmHg.

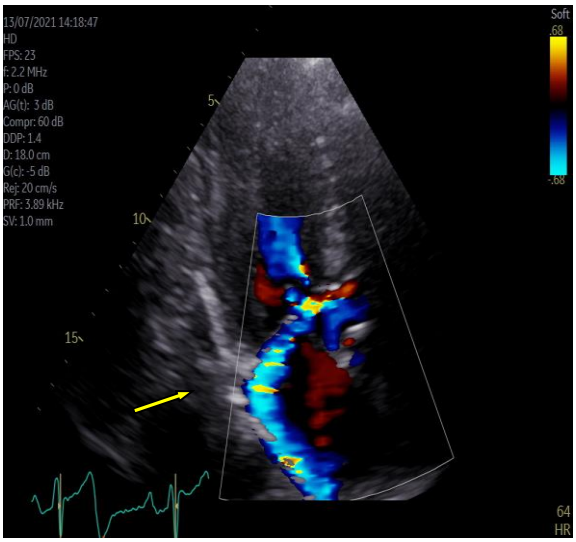

Figure 5

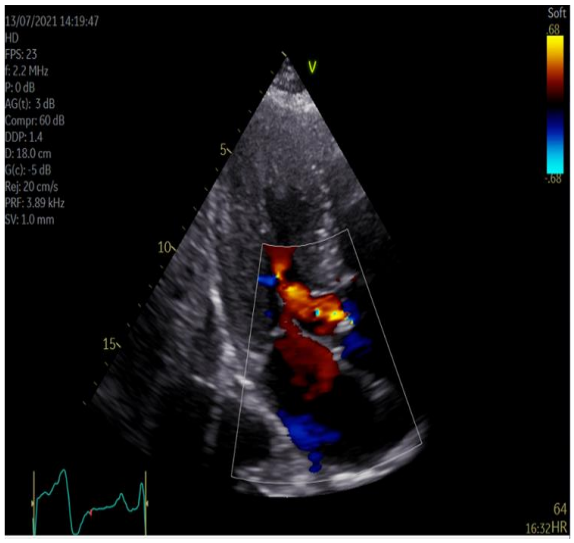

Figure 6

Figure 5. Color Doppler echocardiography showing eccentric moderate mitral regurgitation with postero-laterally oriented jet, due to "coanda effect" (yellow arrow)

Figure 6. Color Doppler echocardiography showing mild aortic regurgitation due to dilation of the ascending aorta.

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Computed tomography angiography (CTA) of the aorta was performed and it showed aneurysmatic dilatation of the ascending aorta at the level of the pulmonary trunk with diameter of 51mm, without intramural thickening or dissecting flap (Figure 7). The ascending part of the aortic arch was also dilated, with diameter of 41 mm with loop along the transversal segment. Descending part of the arch was with diameter of 30 mm, and the diameter of the descending aorta was 25 mm.

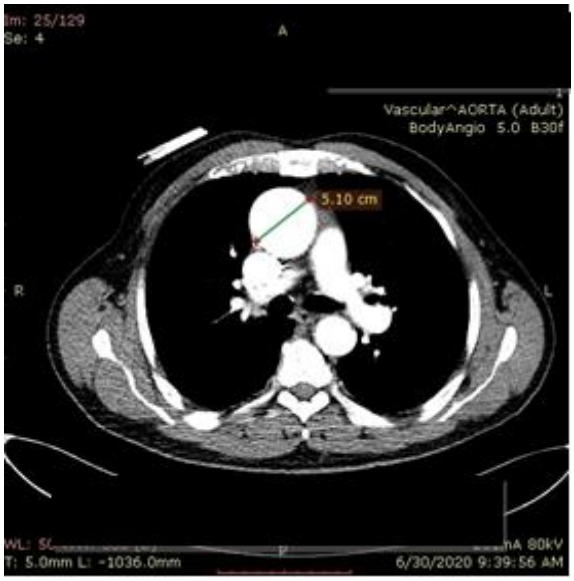

Figure 7- CTA of the aorta- aneurysmatic dilatation of the ascending aorta, with diameter of 51 mm

Because of the hemodynamically significant LVOTO and symptoms, an indication for surgical treatment was established and the patient was referred to a cardiac surgeon. The performed procedure included septal myectomy, mitral valve repair and interposition of tubular graft in the ascending aorta. The operative and postoperative period was without complications and the patient was relieved of symptoms.

Postoperative echocardiography revealed normal dimensions of the left ventricle with normal global systolic function. IVS was with thickness of 14 mm (Figure 8). The continuous wave Doppler velocities through LVOT showed no LVOT obstruction with maximum velocity of 1,3 m/sec and peak gradient of 7 mmHg at rest (Figure 9). Aorta at the level of sinus of Valsalva was 41 mm , while the ascending aorta with the inserted graft, was with normal dimensions.

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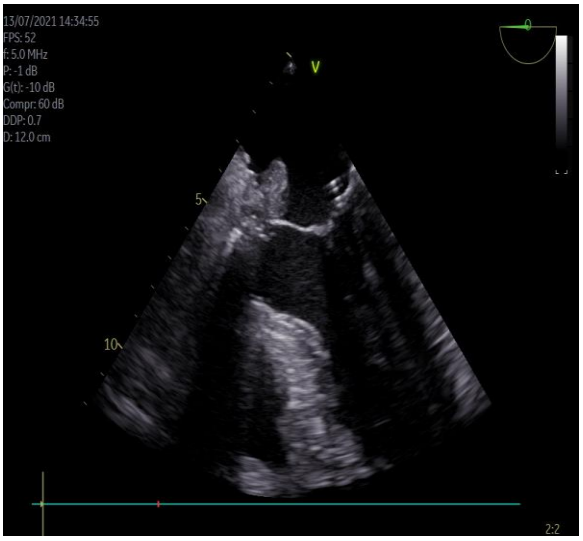

Figure 8. Transoesophageal echocardiography performed postoperatively, showing reduced thickness of the basal IVS.

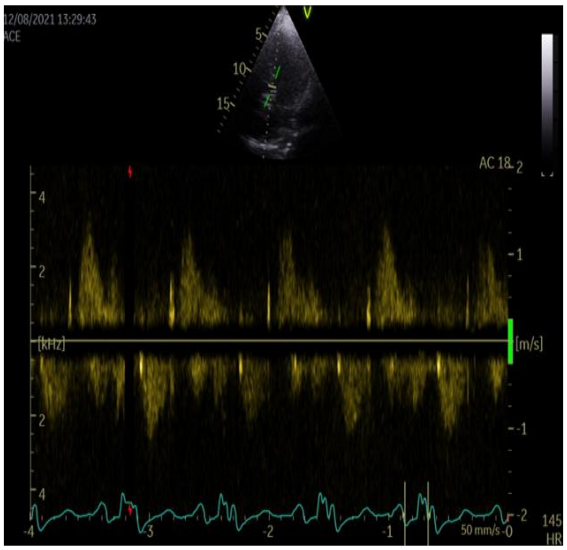

Figure 9. CW Doppler on LVOT showed reduced velocities postoperatively (Vmax 1,3 m/sec and peak gradient of 7 mmHg)

Discussion

The case of this patient demonstrates an uncommon presentation of severe LVOTO due to HCM, which is provoked by the basal septal hypertrophy, SAM of the AML and the uncommon insertion of an accessory muscle bundle, that caused additional increasing of the diameter of the basal IVS and made the gradients higher through the LVOT. The severe LVOTO was manifested with dyspnea and syncope.

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LVOTO is defined as peak Doppler LVOT pressure gradient ≥30 mm Hg at rest or during physiological provocation such as Valsalva manoeuvre, standing and exercise. A gradient of ≥50 mm Hg is usually considered to be the threshold at which LVOTO becomes haemodynamically important (5).

About one-third of patients with HCM have LVOT obstruction at rest. In another one-third of patients, without outflow obstruction at rest, a gradient can be provoked by physiologic and pharmacologic interventions that diminish LV end-diastolic volume or increase LV contractility such as the Valsalva manoeuver. (6).

LVOTO causes acute reductions in cardiac output, elevated LV filling pressures, and myocardial ischemia, which can result in symptoms of chest pain, exertional dyspnea, presyncope, and syncope (6). Cardiac output reduction is considered for dominant mechanism of abnormal blood pressure response, during exercise, causing syncope in patients with dynamic LVOTO (7).

The obstruction in LVOT in patients with HCM can occur due to SAM, but also can occur due to papillary muscle abnormalities (hypertrophy of the muscle, anterior and internal displacement, direct insertion into the anterior mitral valve leaflet) and mitral leaflet abnormalities such as elongation or accessory tissue(5). Other morphological feature that can contribute to LVOTO is an insertion of an accessory muscle bundle, stretching from the apex to the basal anterior septum of the free wall of left ventricle. Insertion of an accessory muscle bundle is a unique congenital morphological malformation contributing to LVOTO (2).

It was performed a study with 230 patients with HCM, where cardiac magnetic resonance (CMR) was used to evaluate the presence of an accessory muscle bundle stretching from the apex to the basal IVS or free LV anterior wall in these patients. An apical-basal accessory muscle bundle was present in the majority of evaluated patients, with percentage of 63 % (145 pts) (2). It is considered that the accessory muscle bundle is congenital malformation typical for patients with HCM. But more data are needed for this theory, because the study included only patients with HCM in the evaluation of an accessory muscle bundle presence, and the presence in the general population was not evaluated (2).

In presence of SAM, coaptation of mitral leaflets is also impaired, leading to mitral insufficiency. Mitral regurgitation (MR) due to SAM has eccentric, postero-laterally oriented jet, occurring in the late systole. This jet is typical for functional, SAM realted MR. If the jet is central or anteriorly oriented, pansystolic, it should be done further assessment for intrinsic mitral valve abnormality. The severity of SAM-related MR is in direct proportion with the severity of LVOTO. (5)

Patients with HCM often have impaired untwisting motion of the LV, due to the hypertrophy, so they have diastolic dysfunction. Accessing and grading of the diastolic dysfunction is helpful for the evaluation of symptoms. (5)

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In symptomatic patients with obstructive HCM who have associated cardiac disease requiring surgical treatment, such as in our case, mitral regurgitation and dilated ascending aorta, surgical myectomy is the procedure of choice. (8)

Surgical myectomy is effective in more than 80% of performed procedures and in experienced centres, pre-operative mortality rates were estimated that have been below 2% . Surgery has also been effective in reducing the symptoms of presyncope and syncope. In a subsequent review of 65 patients who had undergone septal myectomy between 1986 and 1992, significant improvement was observed in 86% of patients with pre-syncope preprocedure and in 100% of patients with syncope (9).

Echocardiography has important role of detection, grading the severity of LVOTO, deciding for treatment options, especially in symptomatic patients, and follow up of the patients either for disease staging in those on medical treatment or after surgical treatment.

In our patient, the septal bulge due to the HCM, the insertion of an accessory muscle bundle at the basal IVS, that was thickening the IVS additionally, and the SAM of the AML, were the contributing factors that provoked severe dynamic LVOTO.

Even difficult to detect with TTE, the presence of an accessory muscle bundle was recorded with TTE and the same was confirmed by the surgeon intraoperatively.

LVOT peak gradient in this patient, at rest, was higher of the threshold at which LVOTO becomes haemodynamically important (86,3 mmHg), while during exercise it is supposed that was even higher, so because of that the patient had exercise induced syncope.

The dagger-shaped Doppler waveform derived with pulsed wave Doppler is typical morphology for peak gradient at dynamic LVOTO (1,5).

Turbulent flow and high pressure in LVOT damaged the aortic valve and it was also reason for dilatation of the ascending aorta and mild aortic regurgitation.

Because of the hemodynamically significant LVOTO and symptoms, an indication for surgical treatment was established and the patient was referred to a cardiac surgeon. At the patient was performed septal myectomy, reconstruction of anterior mitral leaflet, and also resection of the ascending aorta with interposition of tubular graft with diameter of 30 mm.

The operative and postoperative periods were without complications and with complete recovery and symptoms resolution. Postoperative echocardiography revealed normal velocity and gradient in the LVOT, with mild mitral regurgitation, slightly dilated aorta at the level of sinus of Valsalva and normal dimensions of the ascending aorta at the level of the graft. Postoperative assessment of the patient showed normal hemodynamic parameters and gas exchange. The patient was discharged from hospital at the 13th day postoperatively in good general condition and normal post-surgical wound healing.

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Conclusion

Symptomatic patients with basal septal hypertrophy that contributes to LVOT obstruction, with hemodinamically significant gradients, had an indication for surgical or interventional treatment. According to the guidelines, symptomatic patients with obstructive HCM who have associated cardiac disease are requiring surgical treatment, such as in our case with mitral regurgitation and dilated ascending aorta, surgical myectomy is the procedure of choice.

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