A Case Report of Asymmetry of Systolic Anterior Motion of the Mitral Valve, a Not Widely Well-Known but Practically Important Echocardiographic Presentation in Hypertrophic Cardiomyopathy

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INTRODUCTION

Systolic anterior motion (SAM) of the mitral valve is well known as a typical feature of hypertrophic cardiomyopathy with left ventricular outflow tract (LVOT) obstruction.[1,2] A variety of patterns of SAM have been shown in some previous reports.[3-8] However, there have been very few reports that focused on the asymmetry or laterality of SAM.[9,10] Here we report a patient with hypertrophic obstructive cardiomyopathy (HOCM) who showed SAM unevenly on the medial side of the anterior mitral leaflet. The existence of asymmetry of SAM should be carefully examined because this phenomenon may cause underestimation of the LVOT pressure gradient (PG) obtained from standard planes in an apical five-chamber or long-axis view. To measure LVOT PG more precisely, anatomic localization of SAM can provide important information because it is necessary to scan the optimal plane in which SAM exists predominantly.

CASE PRESENTATION

A 70-year-old man previously diagnosed with HOCM presented to our hospital with worsening of exertional dyspnea of New York Heart Association functional class II. On physical examination, the fourth sound and a mid to late systolic murmur (Levine grade III/IV) were heard. Electrocardiography showed a left ventricular (LV) strain pattern with high R waves in precordial leads and ST-segment depression and T-wave inversion in leads I, aVL, and V_4 to V_6 (Figure 1), suggestive of LV hypertrophy. All procedures followed were in accordance with the ethical standards of the responsible committees on human experimentation (institutional and national) and with the Declaration of Helsinki of 1964 and later versions.

Transthoracic echocardiography demonstrated a normal LV internal dimension, an LV ejection fraction of 64%, asymmetric septal hypertrophy, SAM of the anterior mitral leaflet, and midsystolic semiclosure of the aortic valve in a parasternal long-axis view on two-dimensional imaging (Video 1). M-mode echocardiography at the aortic valve level showed midsystolic semiclosure of the aortic valve objectively (Figure 2). These findings might indicate the presence of LVOT obstruction. Aliased flow at the LVOT on color-flow Doppler imaging in apical five-chamber and long-axis views during systole also suggested LVOT obstruction (Figure 3). However, continuous-wave Doppler echocardiography showed an LVOT PG of 10 mm Hg at rest in an apical five-chamber view. During the Valsalva maneuver, the peak LVOT PG rose to 19 mm Hg. As there was a discrepancy between some findings that indicated the existence of LVOT obstruction (auscultatory findings, SAM of the mitral valve and midsystolic semiclosure of the aortic valve, and aliased flow at the LVOT) and the obtained LVOT PG, we performed detailed observation of the mitral valve again to search for possible explanations.

On reevaluation in the parasternal long-axis view, SAM of the mitral valve was clearly observed in the medial plane by a tilting maneuver of the transducer toward the inferior-medial side from a standard parasternal long-axis view (Figure 4A, Video 2). Motion of the anterior mitral leaflet showed complete systolic septal contact, with an acute angle between the anterior mitral leaflet base and its tip position. Conversely, in the lateral plane by a tilting maneuver toward the anterior-lateral side (Video 3), although "pseudo"-incomplete SAM of the chordae tendineae without systolic septal contact was observed (Figure 4B, *yellow arrowhead*), SAM of the mitral valve leaflet became unclear (Figure 4B, *yellow arrow*). Zoomed evaluation of the mitral valve in the parasternal short-axis view revealed that the portion of SAM was located on the medial to central side of the mitral valve. Notably, systolic forward movement of the anterior mitral leaflet was detected mainly on the medial side (Video 4). Further close observation on M-mode imaging at the mitral valve level in the parasternal short-axis view demonstrated the presence of SAM on the medial to central side of the anterior mitral leaflet with complete systolic septal contact (Figure 5A) and the absence of SAM on the lateral side (Figure 5B). Following these observations, we tried to angle the transducer with combination of slight counterclockwise rotation and tilting motion across the LVOT toward the medial side from the standard apical five-chamber view, and high pulse repetition frequency Doppler echocardiography showed an LVOT PG of 54 mm Hg (Figure 6A), and continuous-wave Doppler echocardiography showed a peak LVOT PG of 70 mm Hg (Figure 6B) at rest in the medial plane. During the Valsalva maneuver, the peak LVOT PG rose to 98 mm Hg. We then manipulated the transducer with an angling maneuver across the LVOT toward the lateral side, and the significant LVOT PG disappeared (Figure 6C) in the lateral plane. Furthermore, moderate mitral regurgitation with a posterior-laterally directed jet due to SAM was observed in the medial plane in the

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VIDEO HIGHLIGHTS

Video 1: Two-dimensional imaging in the parasternal long-axis view showing asymmetric septal hypertrophy with a normal ejection fraction and SAM of the mitral valve.

Video 2: Medial plane in the parasternal long-axis view. SAM of the anterior leaflet with complete septal contact was observed.

Video 3: Lateral plane in the parasternal long-axis view. Although "pseudo"-SAM of the chordae tendineae was observed, SAM of the mitral valve was unclear.

Video 4: Zoomed mitral valve in the parasternal short-axis view. SAM was located on the medial to central side of the mitral valve with marked detection of systolic forward movement of the anterior mitral leaflet mainly on the medial side.

Video 5: Color Doppler imaging in the medial plane in the parasternal long-axis view showing moderate mitral regurgitation due to SAM of the mitral valve

Video 6: Color Doppler imaging in the lateral plane in parasternal long-axis view showing no significant mitral regurgitation.

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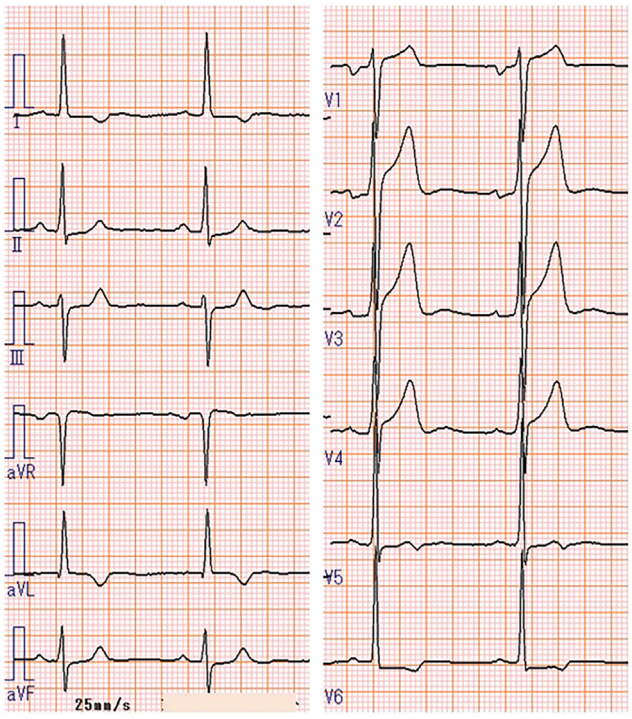

Figure 1 Electrocardiogram at the time of the visit. Sinus rhythm with high voltage in the precordial leads and ST-segment depression and T-wave inversion in leads I, aVL, and V_4 to V_6, suggestive of LV hypertrophy.

parasternal long-axis view (Figure 7A, Video 5). Conversely, significant mitral regurgitation disappeared in the lateral plane (Figure 7B, Video 6).

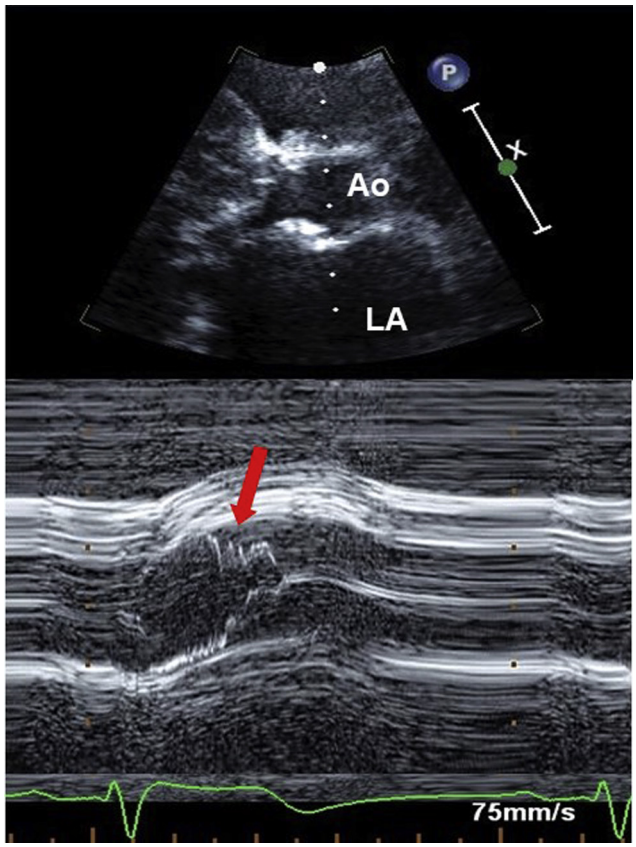

Figure 2 Two-dimensional M-mode image showing systolic semiclosure of the aortic valve *(red arrow)*. *Ao*, Aorta; *LA*, left atrium.

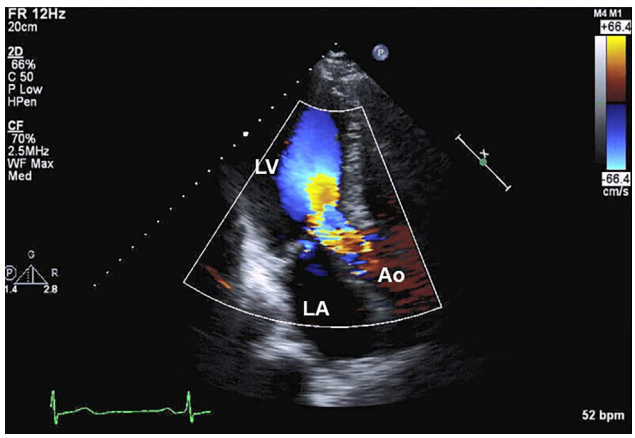

Figure 3 Color Doppler method showing aliased flow at the LVOT during systole in the apical long-axis view. *Ao*, aorta; *LA*, left atrium; *LV*, left ventricle.

DISCUSSION

The presented case emphasizes the importance of an understanding of the existence of asymmetry of SAM in patients with HOCM. In this case, SAM existed predominantly on the medial side of the mitral valve, resulting in a narrow laterally located LVOT and an increased

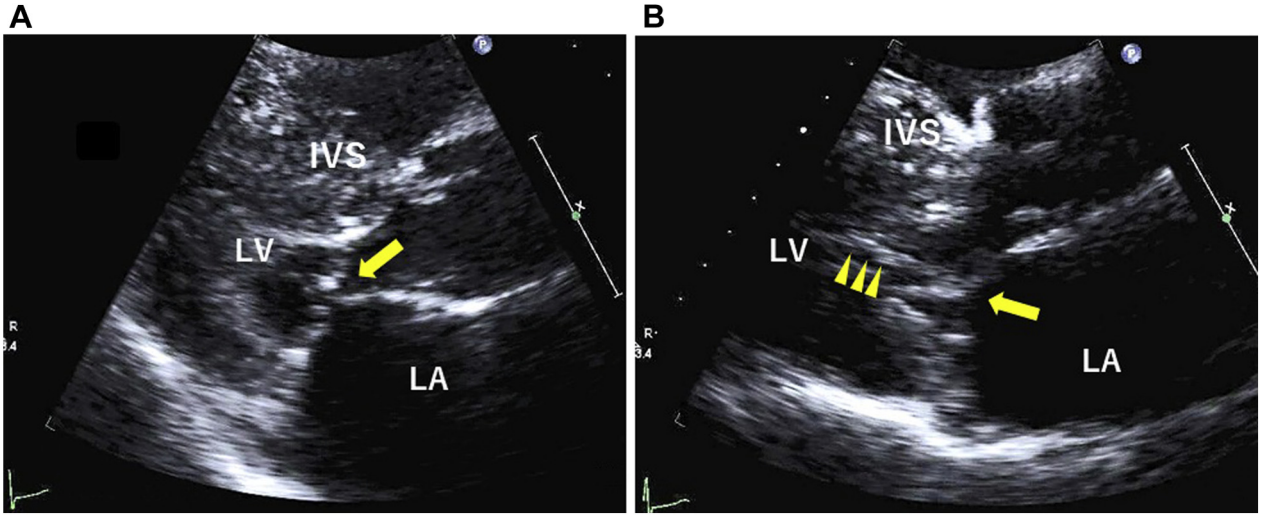

Figure 4 Parasternal long-axis view. **(A)** In the medial plane, SAM of the mitral valve was observed *(yellow arrow)*. **(B)** In the lateral plane, although SAM of the chordae tendineae moving toward the LVOT was observed *(yellow arrowhead)*, SAM of the mitral valve became unclear *(yellow arrow)*. *IVS*, Interventricular septum; *LA*, left atrium; *LV*, left ventricle.

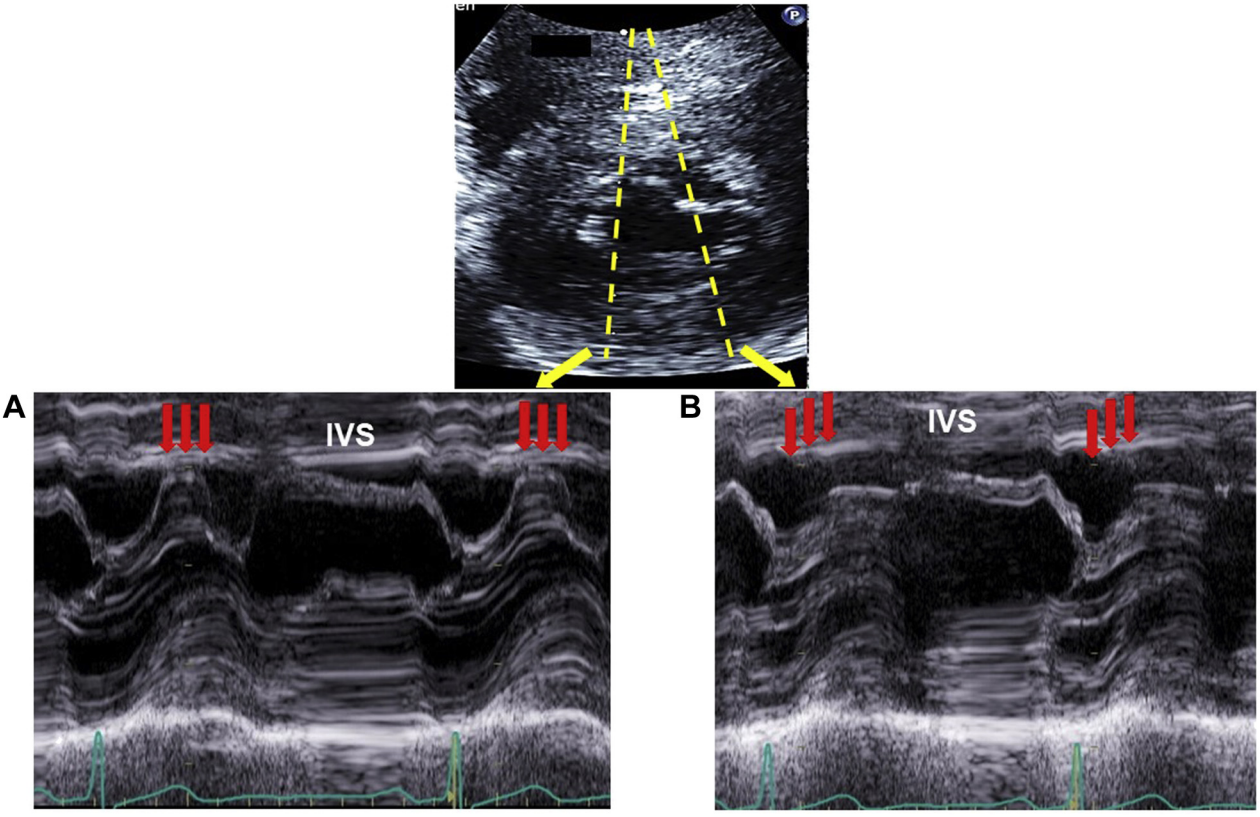

Figure 5 M-mode imaging at the level of the mitral valve in the parasternal short-axis view. **(A)** SAM of the anterior mitral leaflet with complete systolic septal contact was observed on the medial side of the mitral valve *(red arrows)*. **(B)** SAM was not observed on the lateral side of the mitral valve *(red arrows)*. IVS, Interventricular septum.

LVOT PG on the medial side. We should be aware of the existence of asymmetry of SAM in patients with HOCM to avoid underestimation of the LVOT PG.

Clinical Significance of LVOT Obstruction in Patients with HCM

In patients with HCM, assessment of the LVOT PG is a central component of the management algorithm[11] because LVOT obstruction is

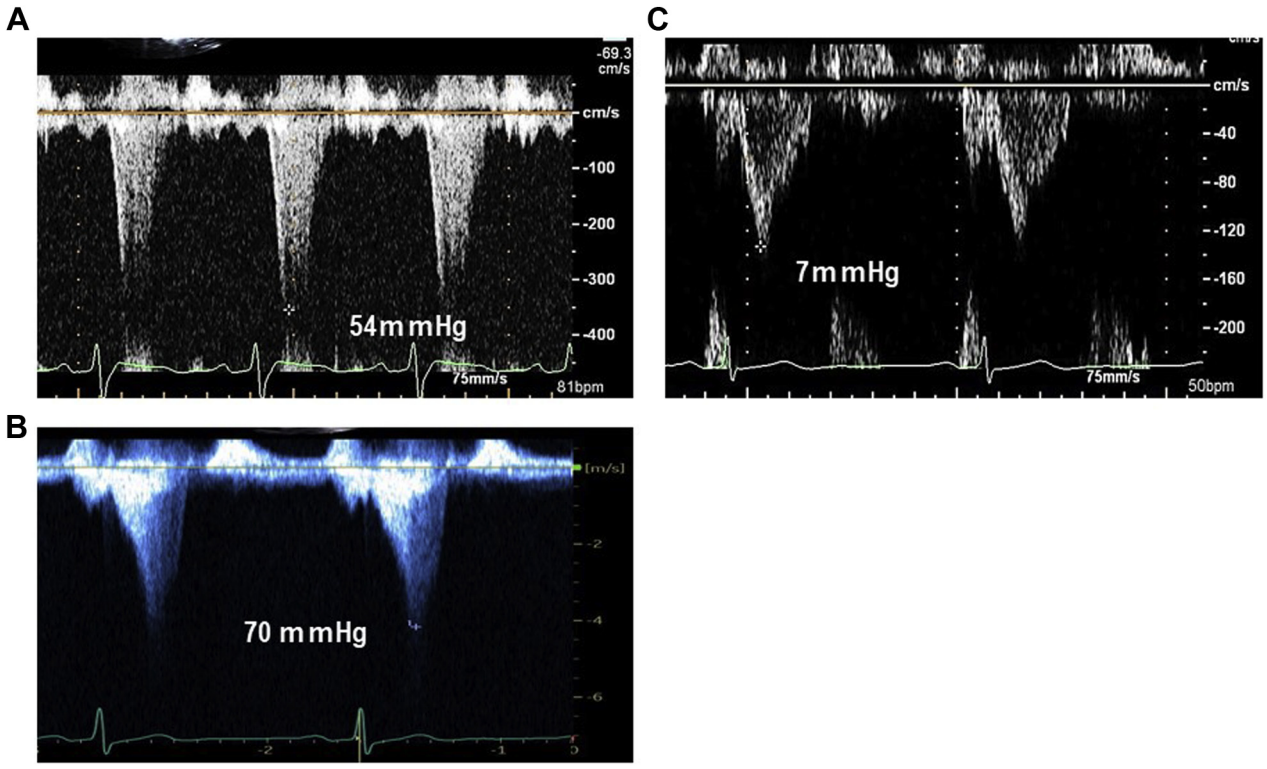

Figure 6 Doppler imaging in the parasternal long-axis view. **(A)** In the medial plane, high pulse repetition frequency Doppler echocardiography showed a peak LVOT gradient of 54 mm Hg, and **(B)** continuous-wave Doppler echocardiography showing a peak LVOT gradient of 70 mm Hg. **(C)** In the lateral plane, pulsed-wave Doppler echocardiography showed a peak LVOT gradient of 7 mm Hg.

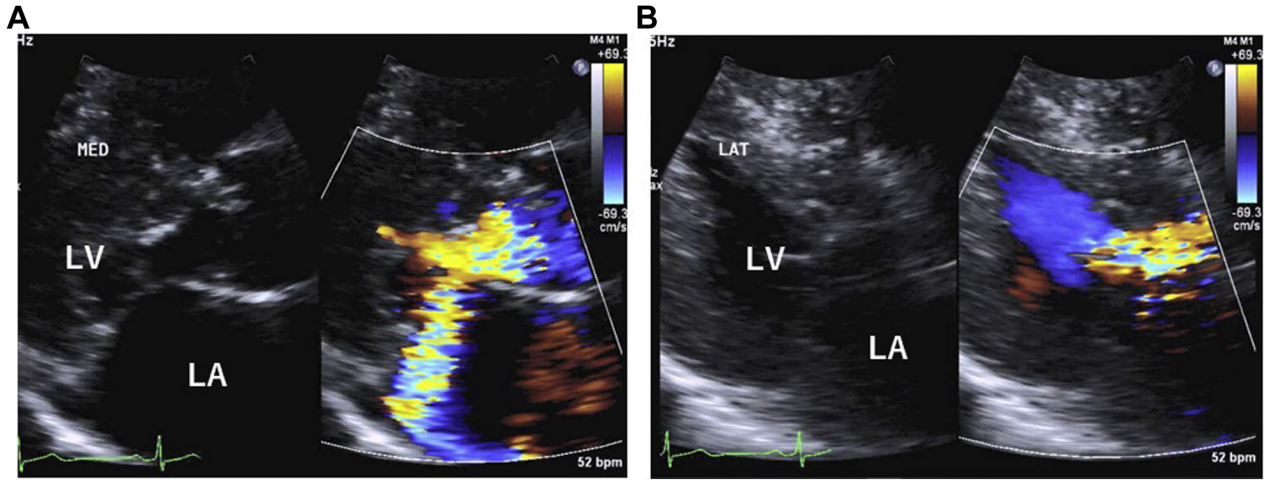

Figure 7 Color Doppler imaging. **(A)** In the medial plane, moderate mitral regurgitation due to SAM was observed. **(B)** In the lateral plane, significant mitral regurgitation was not observed. *LA*, Left atrium; *LV*, left ventricle.

associated with cardiac morbidity and mortality.[11,12] Furthermore, in patients with medically refractory HOCM, detailed anatomic information of SAM can provide important guidance for the surgeon in planning the surgical treatment.[13,14]

Appropriate Evaluation of LVOT PG on Echocardiography

Echocardiography can be very useful for evaluating the presence and severity of LVOT obstruction.[11,13,15] However, technical factors may influence the measurement of LVOT PG. The sonographer should reproducibly align the interrogating Doppler beam parallel to the LVOT systolic flow and take care to avoid contamination of the LVOT flow signal with the mitral regurgitation jet.[15] In addition to those points, the presented case suggested that asymmetry or laterality of SAM could be a cause of underestimation of LVOT PG by Doppler imaging obtained from a standard apical view. If asymmetry or laterality of SAM is observed on a two-dimensional image, adequate evaluation of the LVOT PG

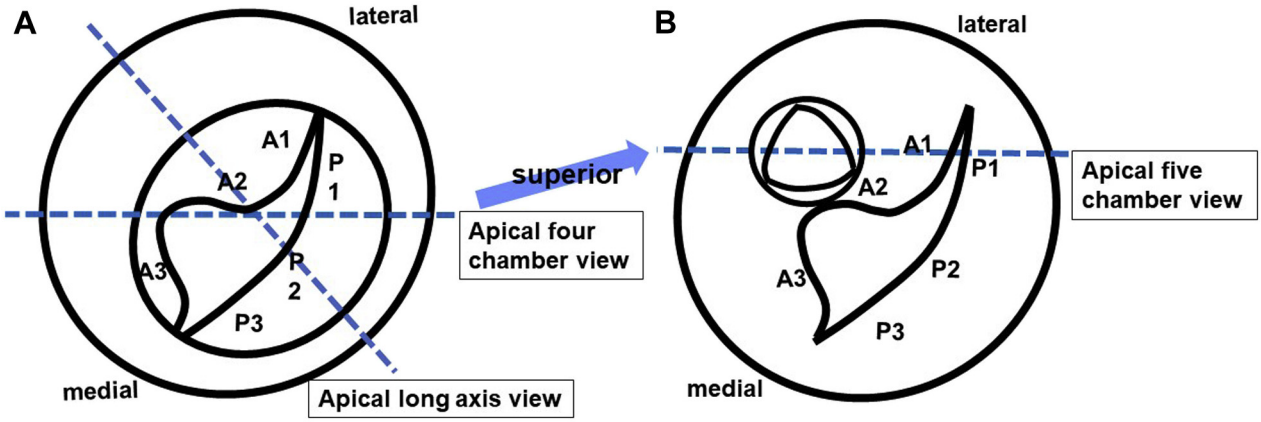

Figure 8 Schema showing mitral valve imaging in the short-axis view in the presented case. **(A)** Mitral valve scallops A3, A2, and P1 in the standard four-chamber view and mitral valve scallops A2 and P2 in the apical long-axis view are observed. **(B)** Then we angle the transducer in the superior direction from the apical four-chamber view, obtaining an apical five-chamber view, and mitral valve scallops A1 and P1 are observed.

on an out-of-standard apical view is required for accurate estimation of the LVOT PG.

A cardiac anatomic understanding is essential for optimal two-dimensional image acquisition. Figure 8 is a schema showing the left ventricle and mitral valve in a short-axis view in this case and the lines in the imaging planes in the standard apical view. We can visualize mitral valve scallops A3, A2, and P1 in the standard four-chamber view and A2 and P2 in the apical long-axis view (Figure 8A). When we slightly angle the transducer toward in the superior direction to obtain an apical five-chamber view (Figure 8B) from the apical four-chamber view, anterolateral segments of the mitral valves (generally scallops A1 and P1) are visualized. Therefore, if we identify that the portion of the SAM is located unevenly in the mitral valve, we need for an appropriate scanning maneuver to the direction of the presence of SAM from the standard apical view to obtain an accurate LVOT PG. In this presented case, we could obtain the LVOT-PG successfully by an angling maneuver toward the medial side from the standard apical five-chamber view.

Asymmetry of SAM of the Mitral Valve in Patients with HOCM

SAM can be seen in patients with HCM with various etiologies and reasons for elongation of the mitral valve, papillary muscle displacement or anomaly, or calcification of the mitral annulus or leaflets.[6] Previously, a variety of patterns of SAM were described, and pathophysiologic or morphologic factors of SAM were investigated.[3-8] However, there have been very few reports that focused on asymmetry of SAM in patients with HCM,[9,10] and it is not a well-known echocardiographic finding in clinical practice.

Song *et al.*[10] investigated the exact features of SAM and the presence of its asymmetry using real-time three-dimensional echocardiography in 39 patients with HOCM.[10] They demonstrated that SAM occurred dominantly on the medial side in 33 of the patients (85%) and in the center in six of the patients (15%). They discussed the mechanism of the asymmetry of SAM. In patients with asymmetry of SAM, significant differences in the length of the mitral valve and interventricular septum wall thickness between the medial side and the lateral side were not shown in their study. Also in our case, there was no significant difference in the length of the mitral valve or the ventricular septum wall thickness between the medial side and the lateral side (data not shown).They speculated that higher flow velocity on the medial side of the LVOT could enhance the generate a larger dragging force to bend the anterior mitral valve leaflet on the basis of previous reports showing higher flow velocity on the medial side of the LVOT adjacent to the interventricular septum than that in the lateral side.

To our knowledge, there have been no reports on asymmetry of SAM to date since the report by Song *et al.*[10] in 2006. As other potential factors, the following points can be considered: (1) LVOT shape is not round but elliptical and (2) abnormal LV septal curvature is seen in patients with hypertrophic cardiomyopathy. These morphologic factors may relate to the direction of systolic flow. The mechanism of asymmetry of SAM is still controversial. Additional analysis with a large cohort using transesophageal echocardiography, comprehensive three-dimensional echocardiographic imaging, vector flow mapping, three-dimensional computed tomography, and four-dimensional flow cardiac magnetic resonance may be needed to elucidate the mechanism of asymmetry of SAM.

CONCLUSION

We reported a case of HOCM masquerading as no significant LVOT PG on color Doppler imaging in the standard apical view due to asymmetry of SAM. If asymmetry of SAM or uneven SAM is identified, the standard plane should be modified to enable scanning of the anatomic point of SAM for accurate estimation of the LVOT PG.

SUPPLEMENTARY DATA

Supplementary data related to this article can be found at https://doi.org/10.1016/j.case.2021.03.001.

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