Echocardiographic Diagnosis of a Subaortic Membrane Attached to the Free Edge of the Right Coronary Cusp of the Aortic Valve

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INTRODUCTION

Subvalvular aortic stenosis is a diagnosis that can include several types of anomalies. These range from a fibrous web below the aortic valve in the left ventricular outflow tract (LVOT) to a long fibrous tunnel with aortic annular hypoplasia.[1] Although transthoracic echocardiography (TTE) is often adequate for diagnosis, transesophageal echocardiography (TEE) may be required for definitive diagnosis.[2] We present a case in which a patient was diagnosed with a typical subaortic membrane on the basis of the presence of a fibromuscular ridge on TTE; further interrogation with TEE, however, showed that the membrane had an unusual attachment to the right coronary cusp of the aortic valve.

CASE PRESENTATION

A 52-year-old woman with a medical history significant for hypertension and intermittent palpitations presented to her cardiologist reporting increasing palpitations and dyspnea on exertion. Although atrial fibrillation was suspected, electrocardiography showed sinus bradycardia. A Holter monitor was ordered, which also could not confirm atrial fibrillation. TTE was subsequently ordered, which demonstrated normal biventricular function, moderate basal septal hypertrophy, a subaortic ridge (Figure 1, Video 1), and severe subvalvular stenosis with a mean gradient of 38 mm Hg, a peak gradient of 70 mm Hg, and a peak velocity of 4.2 m/sec (Figure 2). Velocities of about 3.8 m/s were also obtained using a Pedoff probe from the right sternal border (Figure 3). Although the septal hypertrophy was overall moderate, the area where the subaortic membrane attached was measured at 2.1 cm (Figure 4). There was also mild tricuspid regurgitation, moderate mitral regurgitation, and mild to moderate eccentric aortic regurgitation. The patient was then referred for surgical removal of the subaortic membrane. In addition, she was scheduled for septal myectomy at the site of membrane attachment. A maze procedure and left atrial excision were planned, as a subsequent Zio patch confirmed paroxysmal atrial fibrillation.

On the day of surgery, intraoperative TEE showed normal biventricular function, basal septal hypertrophy, mild to moderate mitral regurgitation, and mild aortic regurgitation. Significant turbulence was seen through the LVOT on color flow Doppler (Figure 5, Video 2). The LVOT was small, measuring 1.7 cm at its maximal diameter, and was even smaller where the subaortic membrane attached. Although the subaortic membrane was seen along the basal anteroseptal wall, it was also noted to be attached to the free edge of the right coronary cusp of the aortic valve (Figures 6 and 7, Videos 3 and 4). The right coronary cusp was possibly restricted in systole as a result of this connection. Overall, the findings on TEE were most consistent with subvalvular stenosis. The surgical team was informed of these findings. After institution of cardiopulmonary bypass and cardiac arrest, the surgical team confirmed the connection of the subaortic membrane to the free edge of the right coronary cusp. The membrane was carefully resected, and the aortic valve did not need to be replaced. In addition, the patient underwent septal myectomy at the site of subaortic membrane, radiofrequency maze procedure, and left atrial appendage excision. Postbypass TEE showed successful removal of the subaortic membrane (Figure 8), with some unresolved turbulence across the LVOT and aortic valve in the setting of hyperdynamic heart function (Figures 9 and 10) and similar aortic regurgitation. As a result of the high velocities, continuous-wave Doppler was used to evaluate the gradient across the LVOT and aortic valve, which demonstrated that the mean gradient had decreased to 10 mm Hg (Figure 11). The patient was subsequently transferred to the intensive care unit at the conclusion of the case. She was extubated on the same day and had an uneventful recovery. During her follow-up visit, she was recovering well and reported improved exercise tolerance.

DISCUSSION

A subaortic membrane is a potential cause for LVOT obstruction. Although it has traditionally been considered a congenital abnormality, it can also be an acquired abnormality.[3] It is thought to result from a variety of factors, including genetic predisposition as well as geometric and anatomic variations in the LVOT that can lead to local turbulence.[4] This turbulence has the potential to damage the endothelium and promote fibrin deposition, which could subsequently lead to fibroelastic obstruction.

Subaortic stenosis is often associated with other congenital abnormalities, including ventricular septal defect, patent ductus arteriosus, and coarctation of the aorta.[5] In some cases, systolic anterior motion of the mitral valve can be seen as in hypertrophic cardiomyopathy. In this case, the patient was noted to have basal septal hypertrophy, although she had not been previously given the diagnosis of hypertrophic cardiomyopathy. Nevertheless, she did not appear to have any of these associated congenital anomalies.

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VIDEO HIGHLIGHTS

Video 1: TTE, parasternal long-axis view, showing a subaortic membrane.

Video 2: TEE, midesophageal long-axis view, with color flow Doppler showing turbulent flow through the LVOT.

Video 3: TEE, midesophageal long-axis view, showing subaortic membrane attached to the interventricular septum and right coronary cusp of the aortic valve.

Video 4: Three-dimensional TEE, midesophageal aortic valve short-axis view, showing subaortic membrane attached to the right coronary cusp of aortic valve.

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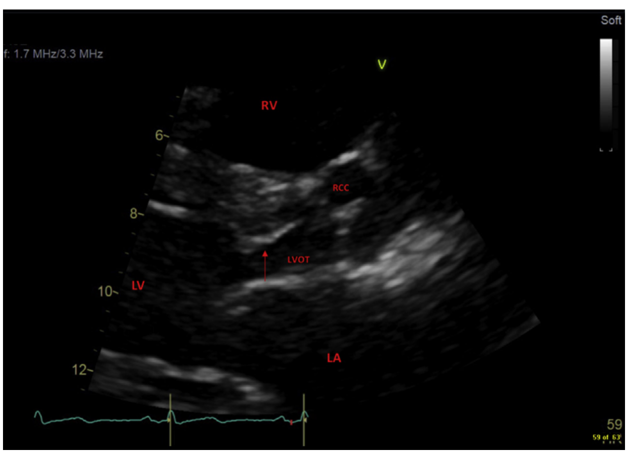

Figure 1 TTE, parasternal long-axis view, showing subaortic membrane (*arrow*). The membrane does not appear to involve the aortic valve. *LA*, Left atrium; *LV*, left ventricle; *RV*, right ventricle; RCC, right coronary cusp.

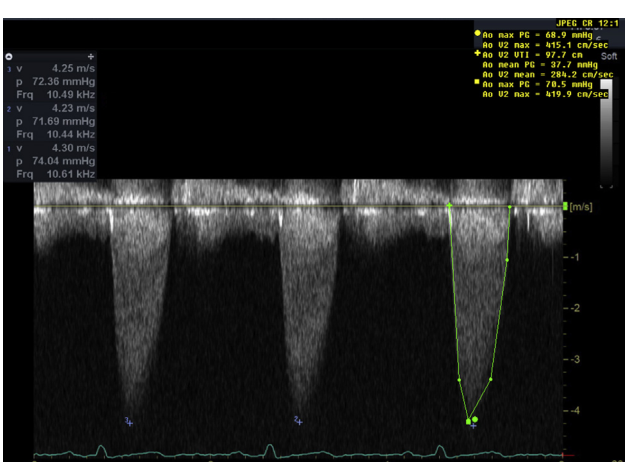

Figure 2 TTE, continuous-wave Doppler profile from an apical five-chamber view through the LVOT and aortic valve, showing velocities >400 cm/sec.

Echocardiography is the test of choice for diagnosis.[6] Furthermore, it can be used to characterize the anatomy of the membrane, determine the severity of obstruction, and evaluate the integrity of the mitral and aortic valves. If additional information is needed, cardiac magnetic resonance imaging, computed tomography, and cine angiography may also play a role.[7]

There are several different types of subaortic membranes: (1) thin fibrous membrane, (2) thick funnel-shaped fibrotic funnel ring, (3) irregular fibromuscular tissue, and (4) long tunnel type.[8] Patients with tunnel-type membranes and those with LVOT obstruction at multiple levels tend to have a worse prognosis in terms of risk for recurrence and reoperation.[6] In this case, TTE demonstrated a seemingly classic appearance of a fibromuscular ridge. However, TEE has superior resolution and clearly showed a highly unusual attachment to the free edge of the right coronary cusp. Although attachment to the base of the aortic leaflets has been reported, we are unaware of reports describing attachment to the free edge of the aortic cusp.

In our case, it is unclear why TEE was not sought after diagnosis of the subaortic membrane to better characterize the anatomy. TEE may provide a more reliable and accurate diagnosis compared with TTE and is especially valuable when the findings on TTE are inconclusive.[6] Had TEE been performed earlier, this unusual variant would likely not have been an intraoperative surprise, possibly allowing additional time for better surgical planning. Interestingly, close inspection of the transthoracic echocardiographic parasternal long axis (Video 1) does raise suspicion for attachment to the aortic valve. However, this observation is made in hindsight after the benefit of seeing the transesophageal echocardiography images, which were far clearer.

Interestingly, Figure 7 suggests limited obstruction of the LVOT by the subaortic membrane, thereby calling into question whether the membrane was the sole cause of the elevated gradients. Although the attachment of the subaortic membrane was quite unusual, the overall clinical pictures suggests that the cause of the elevated high gradients was multifactorial. The flow acceleration began proximal to the LVOT, which may have resulted from the thickened septum at the site of membrane attachment, as well as the small size of the LVOT. It is also possible that the right coronary cusp contributed to the gradient, as it appeared restricted (with improved appearance after removal of the subaortic membrane) as a result of the membrane attachment. However, the appearance of the aortic valve in short axis (Figure 7) did not support aortic valvular stenosis as the primary cause of the high gradients. Unfortunately, the high velocity of blood progressing through the aortic valve prohibited obtaining an accurate gradient at the level of the valve with pulsed-wave Doppler, because of aliasing. Nevertheless, removal of the membrane was associated with a significant improvement in the gradient through the LVOT, although the concomitant performance of a septal myectomy probably affected this as well. Overall, it seems likely that the elevated LVOT gradients may have resulted from several factors, including a small LVOT at baseline, asymmetric basal septal hypertrophy, and the subaortic membrane.

Unfortunately, surgical treatment is not necessarily permanently curative, as subaortic membranes may reoccur despite resection. One study determined that independent risk factors for reoperation were closer proximity to the aortic valve (<6 mm), peak Doppler gradient ≥60 mm Hg, and peeling the membrane off the aortic valve.[9] Some advocate for concomitant myectomy, as was done in this case, although this may not offer long-term benefit and does increase the risk for heart block. Therefore, myectomy should be limited to patients with significant left ventricular hypertrophy.[6] Aside from

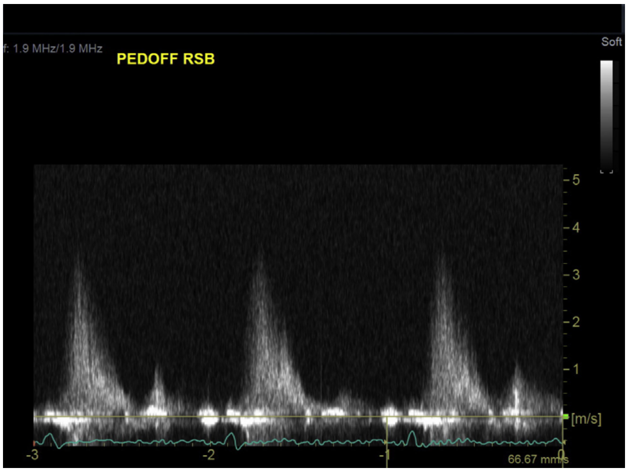

Figure 3 Continuous-wave Doppler profile obtained at the right sternal border using a Pedoff probe with peak velocities of about 3.8 m/sec.

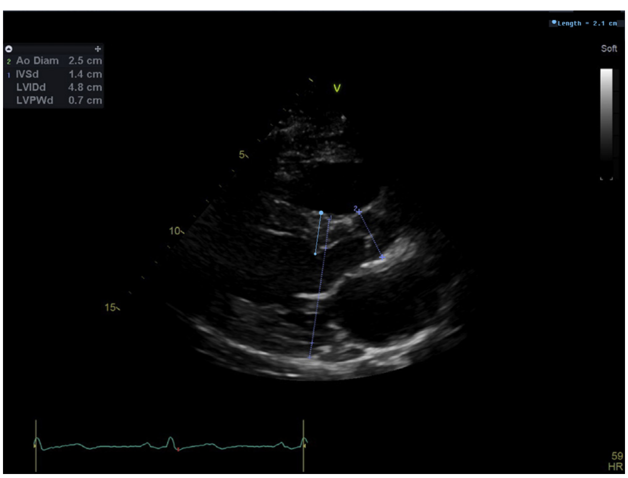

Figure 4 TTE, parasternal long-axis view, with measurements of the left ventricle. The interventricular septum measures 2.1 cm at its maximum width, which is at the site of attachment of the subaortic membrane. The posterior wall of the left ventricle measures 0.7 cm.

reoccurrence of the membrane, it is also possible for aortic regurgitation to develop or progress. However, this is not the case for the majority of patients.[10] Given this information, this patient may be at risk for recurrence given that she exhibited all of risk factors mentioned above. However, it is reassuring that her aortic regurgitation is unlikely to progress.

Although rare, accessory mitral valve tissue should also be considered in the differential diagnosis for LVOT obstruction.[11] It is considered a congenital abnormality resulting from an endocardial cushion defect but has been reported in adults.[11,12] The presentation can be similar to that of a subaortic membrane in terms of associated congenital abnormalities and symptoms such as dyspnea, syncope, chest pain, and palpitations. The echocardiographic appearance can be variable and may resemble a mobile parachute-like leaflet, a fixed structure

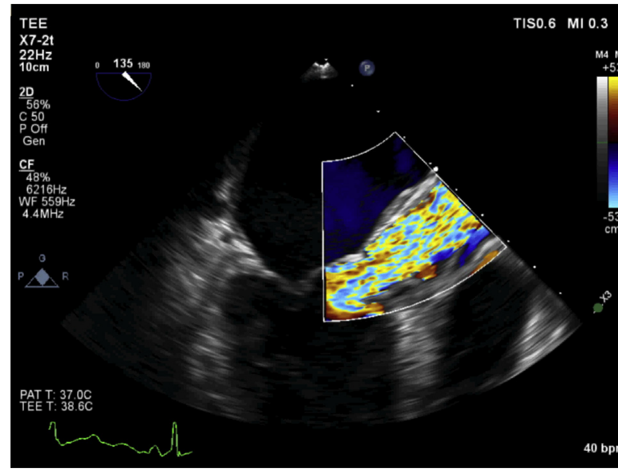

Figure 5 TEE, midesophageal long-axis view, with color flow Doppler showing turbulent flow through the LVOT.

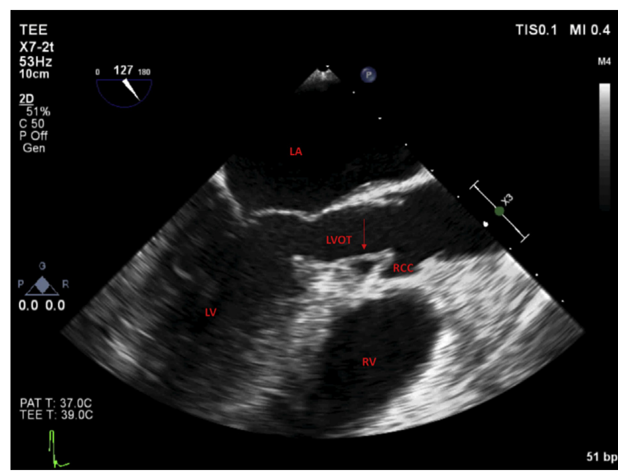

Figure 6 TEE, midesophageal long-axis view, showing a subaortic membrane (*arrow*) attached to the interventricular septum and right coronary cusp (RCC) of the aortic valve. *LA*, Left atrium; *LV*, left ventricle; *RV*, right ventricle.

attached to the interventricular septum by a chordal apparatus, or a globular or cystic mass. Typically, the tissue moves into the LVOT during systole and retracts toward the left ventricle during diastole.[13] In this case, the tissue in the LVOT was not connected to the mitral valve or the mitral subvalvular apparatus, making it unlikely that it represented accessory mitral valve tissue.

CONCLUSION

In summary, we present a patient with a unique variant of a subaortic membrane. Although the preoperative evaluation originally appeared to show a classic fibromuscular ridge, further investigation allowed detailed characterization of the pathology. This case suggests that when echocardiographers diagnose a subaortic membrane, the aortic cusps should be closely examined to determine any involvement. It also highlights that perioperative echocardiographers should complete a thorough examination, even if prior imaging was performed.

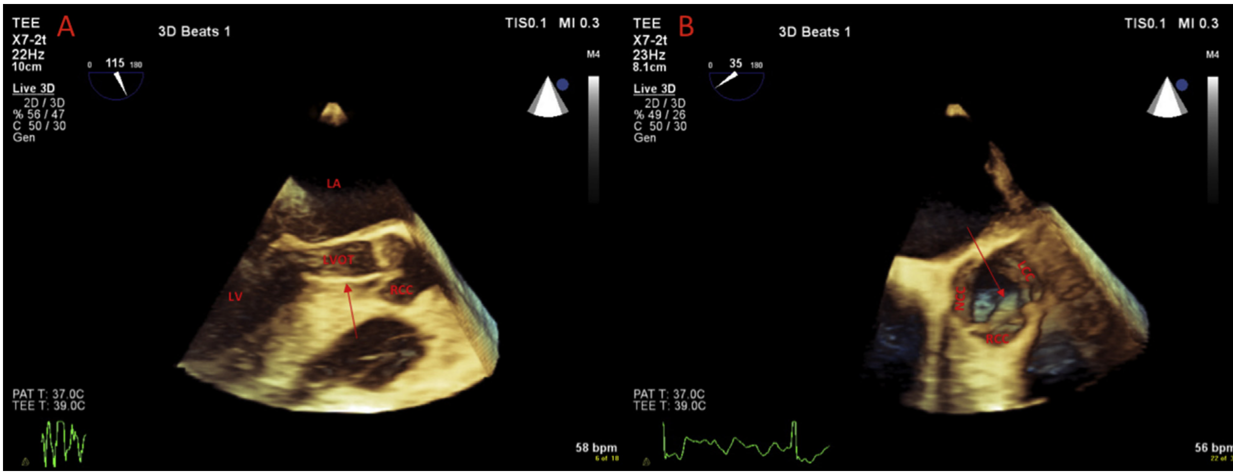

Figure 7 Three-dimensional TEE: midesophageal aortic valve long-axis view **(A)** and short-axis view **(B)** showing a subaortic membrane (*arrow*) attached to the interventricular septum and right coronary cusp (RCC) of the aortic valve. *LA*, Left atrium; *LCC*, left coronary cusp; *LV*, left ventricle; *NCC*, noncoronary cusp.

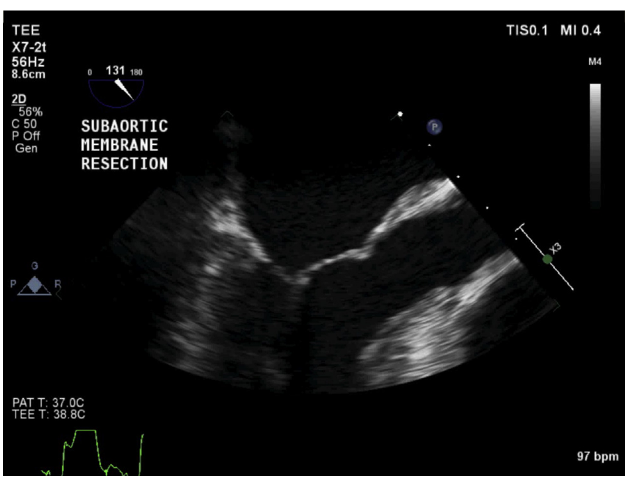

Figure 8 TEE, midesophageal long-axis view, showing successful removal of the subaortic membrane as well as unrestricted opening of the right coronary cusp.

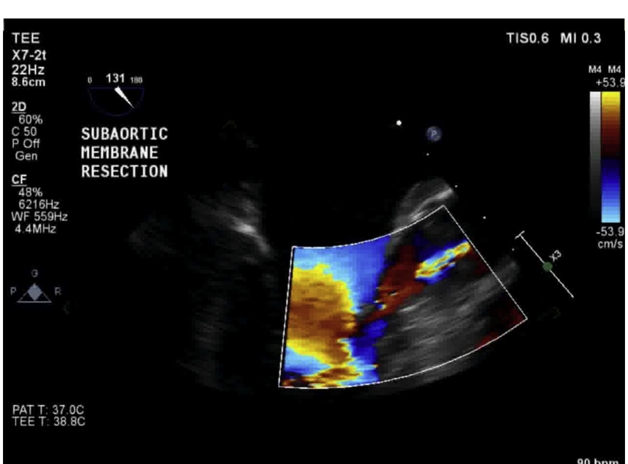

Figure 9 TEE, midesophageal long-axis view, showing similar mild aortic regurgitation after removal of the subaortic membrane. The mitral inflow is characterized by high velocities in the setting of a hyperdynamic heart.

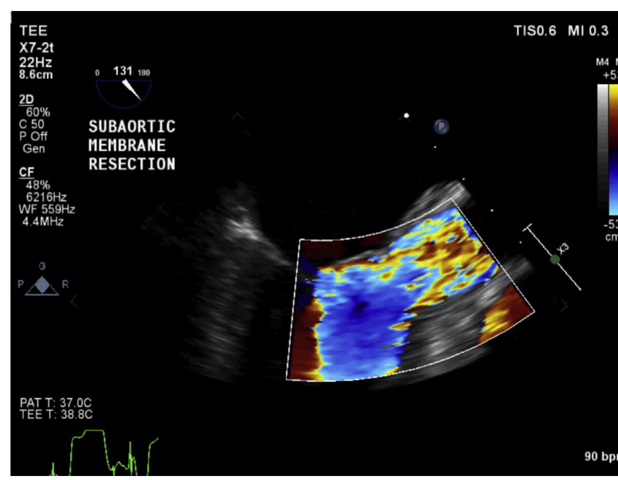

Figure 10 TEE, midesophageal long-axis view, showing some residual turbulence in the LVOT.

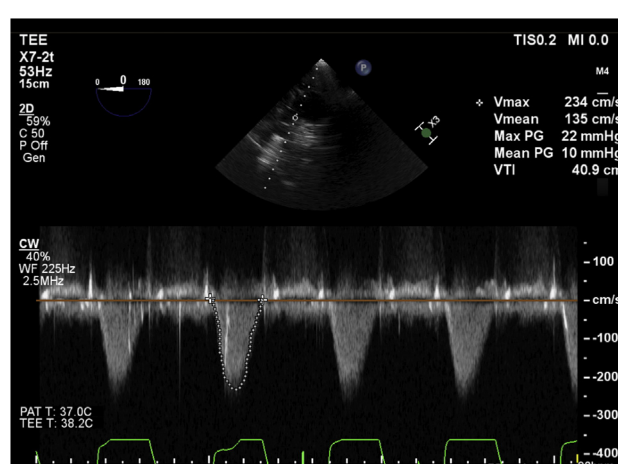

Figure 11 Continuous-wave Doppler profile obtained from a transesophageal deep transgastric long-axis view showing a mean gradient of 10 mm Hg across the LVOT and aortic valve.

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SUPPLEMENTARY DATA

Supplementary data related to this article can be found at https://doi.org/10.1016/j.case.2020.01.004.

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