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CASE REPORT

BEGINNER

CLINICAL CASE

Now You See Me Now You Don't

Subaortic Membrane Causing a Diagnostic Dilemma

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ABSTRACT

Subaortic stenosis secondary to subaortic membrane is the second most common form of left ventricular outflow tract obstruction. We present the case of a 70-year-old male patient who presented with a 6-week history of progressive signs of heart failure. Multimodality imaging was required to confirm the presence of a subaortic membrane. (**Level of Difficulty: Beginner.**) (J Am Coll Cardiol Case Rep 2023;18:101916) Published by Elsevier on behalf of the American College of Cardiology Foundation. This is an open access article under the CC BY-NC-ND license (http://creativecommons.org/licenses/by-nc-nd/4.0/).

HISTORY OF PRESENTATION

A 70-year-old man presented to the emergency department with progressive worsening dyspnea on exertion and weight gain (40 lbs over the previous 3 weeks). On admission, he had a blood pressure of 101/78 mm Hg with increased respiratory effort (respiratory rate, 20 breaths/min) requiring 5 L/min oxygen through a nasal cannula. On physical examination, the patient was ill-appearing, his abdomen was distended, he had elevated jugular venous pressures, and he had bilateral lower pitting edema. A grade 3/4 systolic murmur was appreciated along the left midsternal border. Subsequently, the patient was admitted to the intensive care unit and started on a continuous intravenous furosemide infusion for decompensated heart failure.

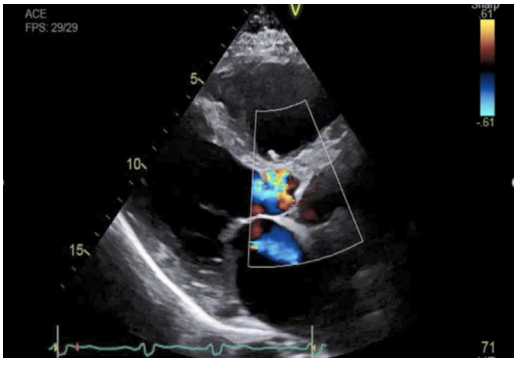
LEARNING OBJECTIVES

- To demonstrate that multimodality imaging is crucial in establishing a more concrete diagnosis of valve disorders.
- To demonstrate the use of invasive pressure wire as a technique in confirming subaortic stenosis by using pressure gradients.

PAST MEDICAL HISTORY

The patient had a history of mild aortic stenosis (AS) diagnosed 8 months previously, atrial fibrillation,

FIGURE 1 Transthoracic Echocardiography on Admission, Parasternal Long-Axis View Showing Severe Aortic Stenosis



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ABBREVIATIONS AND ACRONYMS

AS = aortic stenosis

CMR = cardiac magnetic resonance

CT = computed tomography

LVOT = left ventricular outflow obstruction

TTE = transthoracic echocardiography

nonobstructive coronary artery disease, interstitial lung disease, chronic kidney disease stage III, obstructive sleep apnea, type 2 diabetes, hypertension, hyperlipidemia, and hypothyroidism.

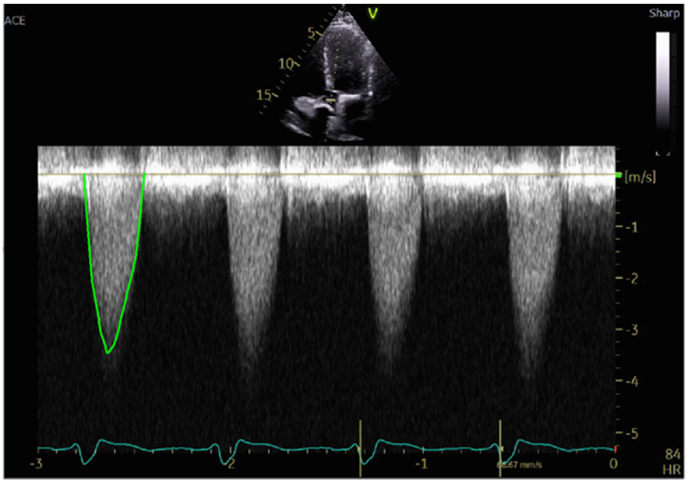
DIFFERENTIAL DIAGNOSIS

Given his rapidly deteriorating condition, a possible diagnosis included progression of AS to the severe range with or without cardiomyopathy.

INVESTIGATIONS

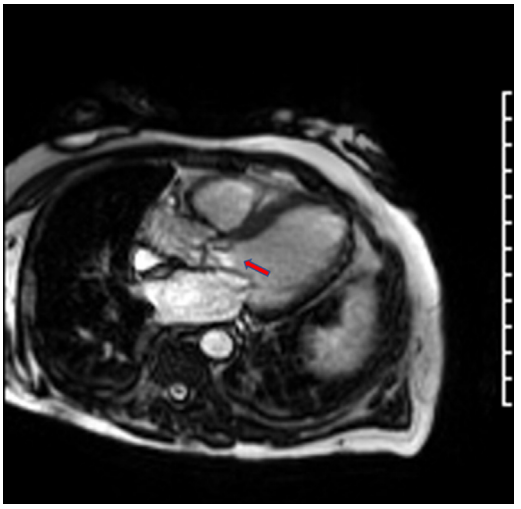
The patient's creatinine level on admission was 2.4 mg/dL, in comparison with a previous normal baseline of 1.0 mg/dL. His B-type natriuretic peptide value was 1,174 pg/mL. The electrocardiogram showed sinus rhythm with right bundle branch block. The transthoracic echocardiography (TTE) on admission showed a left ventricular ejection fraction of 25% to 30% and low-flow low-gradient severe AS with a mean aortic valve gradient of 33 mm Hg and a calculated aortic valve area of 1 cm^2 (**Figure 1**). After diuresis to a nearly euvolemic state, dobutamine stress echocardiography confirmed the diagnosis of severe AS with a mean gradient of 51 mm Hg and a peak velocity of 4.86 m/s (**Figure 2**). In addition, his stay was complicated by COVID-19. The patient's condition soon deteriorated, he started showing signs of confusion, he had elevated lactic acid levels (5 mmol/L), and he was in cardiogenic shock requiring inotropic support. A Swan-Ganz catheter showed elevated filling pressures and low cardiac output. After a 2-week course, he was able to be weaned from inotropic therapy.

FIGURE 2 Dobutamine Stress Echocardiography



The imaging shows a 4.4 m/s maximal velocity across the aortic valve with a mean gradient of 41 mm Hg.

FIGURE 3 Cardiac Magnetic Resonance



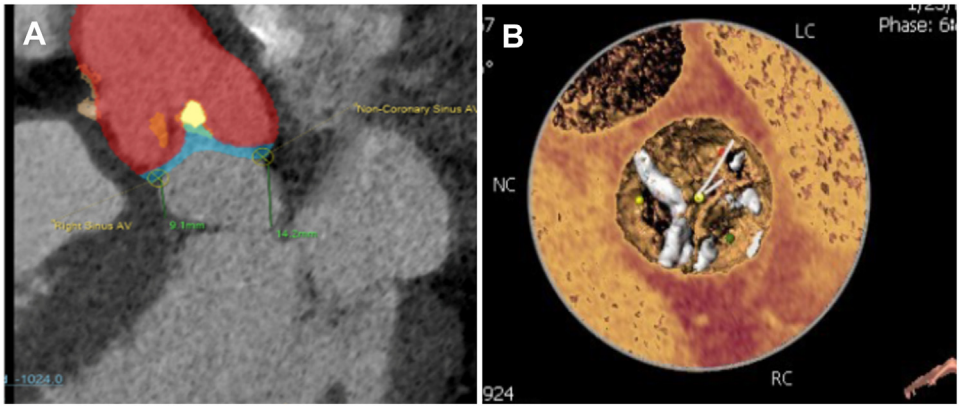
The **red arrow** shows the subaortic membrane.

Cardiac magnetic resonance (CMR) was performed to assess for myocarditis as a possible cause of his rapidly deteriorating heart failure. CMR showed severe AS and a discrete subaortic membrane (**Figure 3**, Video 1). The presence of the subaortic membrane was later confirmed by gated cardiac computed tomography (CT), which also showed a heavily calcified trileaflet aortic valve (**Figures 4A and 4B**, Video 2). Coronary angiography showed no significant obstructive coronary artery disease. Invasive pressure wire (Verrata wire, Philips) testing verified that the pressure gradient was mostly subaortic and not transvalvular (**Figures 5 and 6**). After a multidisciplinary heart team meeting, the decision was made to perform surgical resection of the subaortic membrane and surgical aortic valve replacement.

MANAGEMENT

The patient was discharged to cardiac rehabilitation and medical optimization for 1 month before surgery. He subsequently underwent resection of the subaortic membrane along with aortic valve replacement with a 25-mm Avalus pericardiac valve (Medtronic) (**Figures 7 and 8**). The patient did not have any immediate complications. He was extubated on postoperative day 1 and was transferred to the cardiology

FIGURE 4 Cardiac Computed Tomography



(A) Cardiac computed tomography transcatheter aortic valve replacement protocol confirming the presence of a subaortic membrane in the left ventricular outflow tract. **(B)** Computed tomography showing the calcified trileaflet aortic valve.

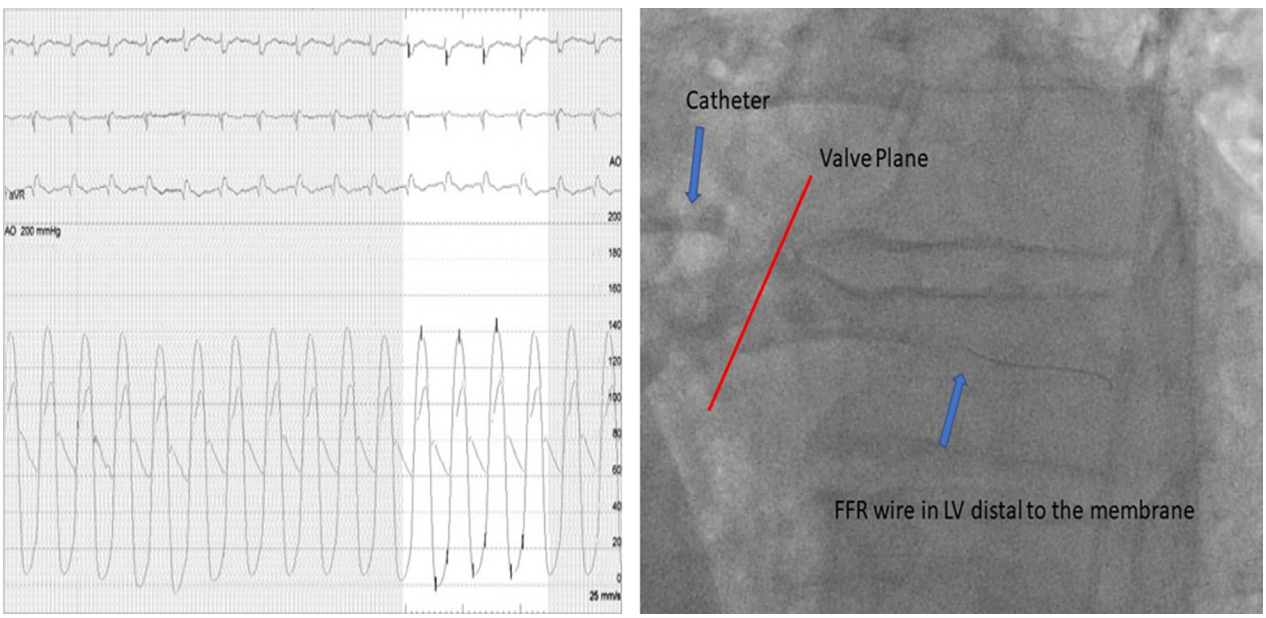
floor on postoperative day 2. He was discharged home 1 week later.

DISCUSSION

Subaortic stenosis, a type of left ventricular outflow tract obstruction (LVOT), is a rare entity in adults that has an unknown origin, with a range of clinical outcomes and symptoms.[1] We present a challenging diagnostic case in a patient with concomitant AS. Although the patient had several echocardiograms, they did not show the subaortic membrane. Eventually, the diagnosis was made by CMR.

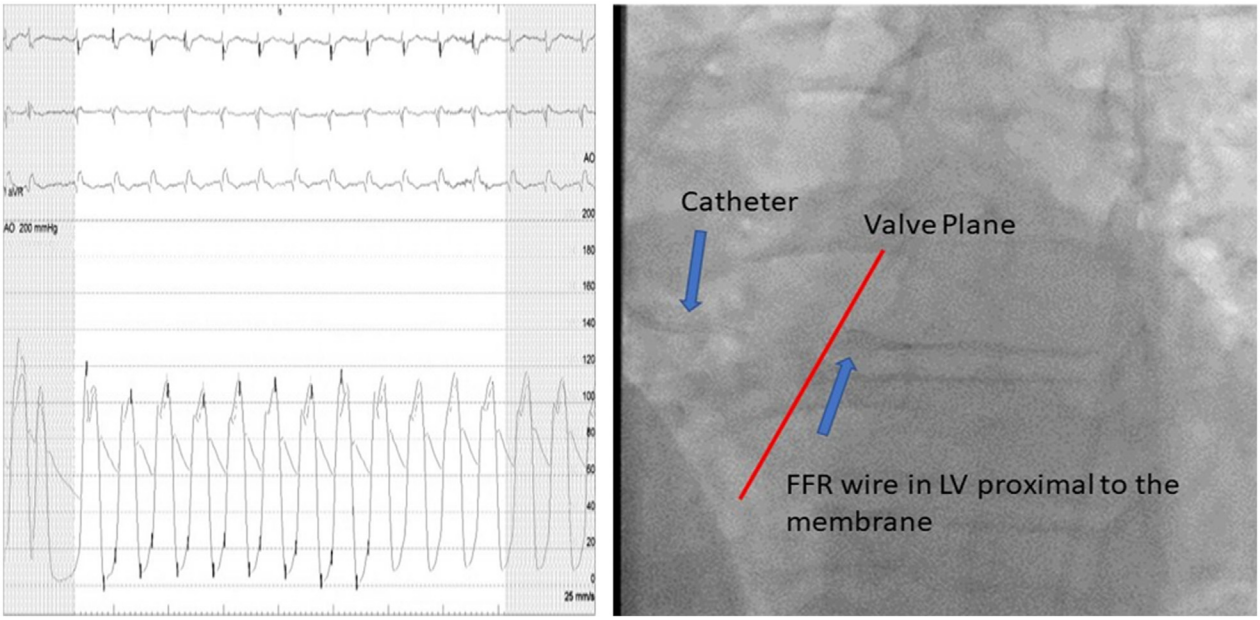
The diagnosis of subaortic membrane is usually established by TTE with color Doppler assistance.

FIGURE 5 Pressure Gradient Wire Measuring 30 mm Hg Across the Subaortic Membrane Area



FFR = fractional flow reserve; LV = left ventricle.

FIGURE 6 Pressure Gradient Wire Measuring 0 mm Hg Across the Aortic Valve, Supra-Membrane Area

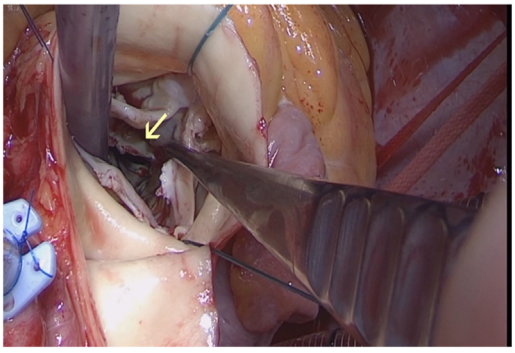


Abbreviations as in Figure 5.

Although TTE remains the first modality of choice, it is not sensitive in diagnosing subaortic membrane because of the thinness of the membrane.[2]

The newer imaging techniques for diagnosing various causes of LVOT include CMR and CT.[3] CMR may be used to measure flow velocity and elucidate anatomy. One drawback of CMR is that the region of interest is frequently hidden by the spin dephasing artifact.[3] This issue makes visualization challenging, especially when taking into consideration the subaortic membrane's thinness. Conversely, cardiac CT complements the role of TTE in diagnosing a subaortic membrane.[4] We demonstrated a novel use of a transducer-tipped pressure wire (usually used to measure fractional flow reserve) in detecting the pressure gradient across the subaortic membrane vs the aortic valve. By detecting the pressure gradient at the level of the membrane and no significant gradient

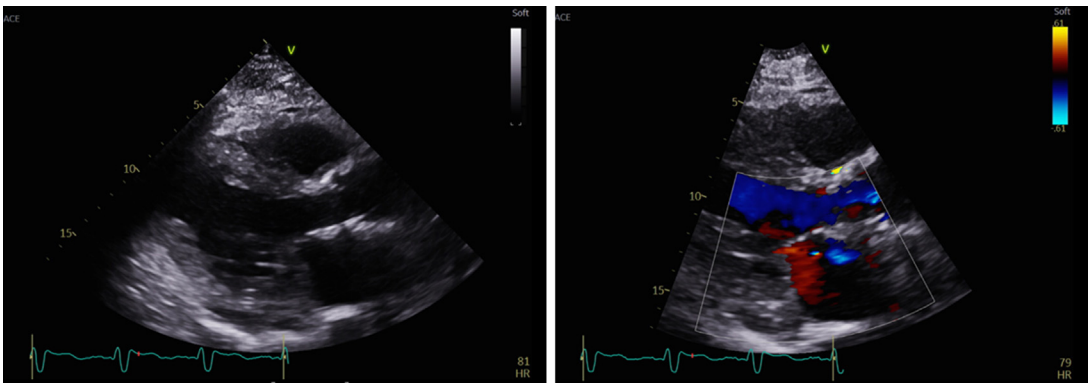
FIGURE 7 The Subaortic Membrane During Surgery



The **yellow arrow** is pointing toward the subaortic membrane.

FIGURE 8 Resected Subaortic Membrane Next to the Native Aortic Valve

FIGURE 9 Postprocedural Echocardiography Showing No Stenosis Across the Aortic Valve



across the aortic valve, we confirmed the diagnosis suggested by CMR and CT.

Surgery remains the gold standard for treating severe and symptomatic subaortic stenosis secondary to a subaortic membrane.[5] Recurrence is not unusual and occurs in up to 30% of patients, especially those with the tunnel variant and those with multiple levels of obstruction.[5,6] Progression of aortic regurgitation from none to mild or mild to moderate is also not uncommon, especially in patients with a preoperative peak LVOT gradient ≥80 mm Hg.[7]

Subaortic membranes are divided into 3 types: discrete ridge, thick fibromuscular ridge, and subaortic tunnel. Although myomectomy can be performed, the risk of recurrence and the overall long-term benefit are controversial.[5-8] Given the discrete ridge anatomy, the patient's age, and the increased risk of complete heart block with myomectomy, the surgeon considered membranectomy alone as adequate removal.

Although the gradient causing the patient's symptoms was mainly the result of the subaortic membrane, the surgeon decided to proceed with a surgical aortic valve replacement during the membranectomy to reduce the need for aortic valve replacement in the near future.

FOLLOW-UP

At 7 months after his surgery, the patient is currently undergoing rehabilitation with gradual resumption of his normal activities. The latest TTE showed an improved left ventricular ejection fraction to 50% to 54% with no AS and normal gradients and velocities across the bioprosthetic aortic valve (**Figure 9**, Video 3).

CONCLUSIONS

The diagnosis of subaortic membrane requires a high index of suspicion. TTE may not be the most sensitive imaging modality for the diagnosis, whereas CMR and cardiac CT are ideal to make this diagnosis. Invasive fractional flow reserve pressure wire pullback can also be helpful in distinguishing the level of obstruction.

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Dr Kleiman has served as a local principal investigator in trials sponsored by Boston Scientific, Medtronic, Abbott, and Edwards Lifesciences. Dr Reardon has served as a consultant for Medtronic, Boston Scientific, Abbott, and W.L Gore & Associates. Dr Goel has served as a consultant for Medtronic and W.L. Gore & Associates; and has served on the Speakers Bureau for Abbott Structural Heart. All other authors have reported that they have no relationships relevant to the contents of this paper to disclose.

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KEY WORDS **aortic stenosis, heart failure, left ventricular outflow obstruction, subaortic membrane**

APPENDIX **For supplemental videos, please see the online version of this paper.**