JACC: CASE REPORTS VOL. 4, NO. 7, 2022
PUBLISHED BY ELSEVIER ON BEHALF OF THE AMERICAN COLLEGE OF CARDIOLOGY FOUNDATION.

CASE REPORT

ADVANCED

CLINICAL CASE

A Rare Case of Post-Mitral Valve Replacement Ventricular Pseudoaneurysm, Bioprosthetic Dehiscence, and Paravalvular Mitral Regurgitation

Pankaj Malhotra, MD,[a,b] Donghee Han, MD,[a] Alan C. Kwan, MD,[a,b] Sabah Skaf, MD,[b] Robert Siegel, MD,[b] Alfredo Trento, MD,[b] Daniel Berman, MD[a,b]

ABSTRACT

We present a rare case of a young patient who underwent a bioprosthetic mitral valve replacement and subsequently experienced a left ventricular pseudoaneurysm complicated by valve dehiscence and paravalvular mitral regurgitation, demonstrated by multimodality imaging and confirmed during surgical repair. (**Level of Difficulty: Advanced.**) (J Am Coll Cardiol Case Rep 2022;4:449–454) 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 27-year old woman presented with new-onset exertional dyspnea and paroxysmal palpitations without prodrome or associated infectious or constitutional symptoms. The patient was afebrile, her blood pressure was 104/61 mm Hg, and her heart rate was 98 beats/min. Her physical examination was notable for a grade III/VI systolic murmur at the fifth intercostal space at the midclavicular line, radiating posteriorly, and bibasilar rales. Her electrocardiogram demonstrated an accelerated junctional rhythm. Initial laboratory results were unremarkable.

LEARNING OBJECTIVES

- To describe the prevalence and risk factors for the development of left ventricular pseudoaneurysms.
- To illustrate the utility of multimodality imaging in the diagnosis of pseudoaneurysms.
- To discuss current management of left ventricular pseudoaneurysms.

MEDICAL HISTORY

The patient's medical history included mitral valve P2/P3 prolapse status post failed valve repair/cleft closure, for which she had undergone bioprosthetic mitral valve replacement (MVR) with a #25 stented bioprosthetic pericardial valve 4 years prior. This bioprosthesis was complicated by valvular stenosis, for which she had undergone a repeated bioprosthetic MVR with a #29 stented bioprosthetic porcine valve 2 months prior to her current presentation. Her history was also notable for paroxysmal atrial fibrillation and atrial flutter, anxiety, and asthma controlled with inhaler therapy.

From the [a]Mark Taper Imaging Center, Cedars Sinai Medical Center, Los Angeles, California, USA; and the [b]Smidt Heart Institute, Cedars Sinai Medical Center, Los Angeles, California, USA.
The authors attest they are in compliance with human studies committees and animal welfare regulations of the authors' institutions and Food and Drug Administration guidelines, including patient consent where appropriate. For more information, visit the Author Center.

Manuscript received November 30, 2021; revised manuscript received February 22, 2022, accepted February 22, 2022.

ISSN 2666-0849 https://doi.org/10.1016/j.jaccas.2022.02.006

ABBREVIATIONS AND ACRONYMS

CMR = cardiac magnetic resonance

CT = computed tomography

MR = mitral regurgitation

MVR = mitral valve replacement

TEE = transesophageal echocardiography

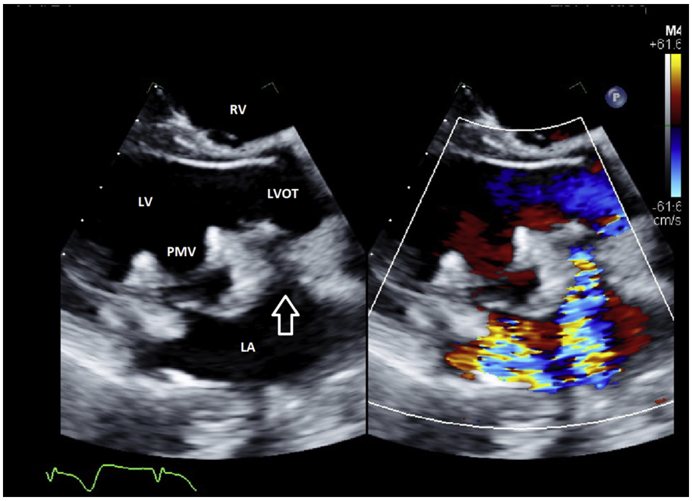
DIFFERENTIAL DIAGNOSIS

The differential diagnosis included bioprosthetic valve dysfunction with mitral regurgitation (MR) or mitral stenosis, tricuspid regurgitation, and symptomatic atrial arrhythmia.

INVESTIGATIONS

A chest radiograph demonstrated prominent pulmonary vasculature markings in the bilateral lung fields. Transthoracic echocardiography exhibited normal left ventricular (LV) systolic function (ejection fraction 56%) with new moderate to severe paravalvular MR and rocking motion of the bioprosthesis (**Figure 1**, Video 1). Subsequent transesophageal echocardiography (TEE) demonstrated a narrow-necked outpouching of the membranous septum that subsequently continued into a wider dimension component of the defect, along with associated anteromedial dehiscence of the bioprosthesis and moderate to severe paravalvular MR (**Figures 2 and 3**, Videos 2 to 6). Cardiac magnetic resonance (CMR) was performed after the TEE primarily to better quantify the paravalvular regurgitation. CMR confirmed the TEE findings and highlighted the presence of the 2.9 × 2.2 cm focal pseudoaneurysm/defect of the membranous septum (**Figure 4**, Video 7) superior to the mitral valve, just posterior to the aorta, with a subsequent associated paravalvular regurgitant jet (Video 8). Indirect quantification of the mitral regurgitant fraction using aortic forward flow volume and LV stroke volume demonstrated regurgitant fraction 44%. Tissue characterization demonstrated normal T1 and T2 relaxation times, normal extracellular volume fraction, and no myocardial or pericardial late gadolinium enhancement. Before surgical repair, CT imaging, performed to evaluate for pulmonary embolism, validated the CMR findings, provided excellent 3-dimensional localization, and demonstrated no further expansion of the pseudoaneurysm and the absence of fistula formation (**Figures 5 to 7**). Owing to the lack of visible vegetations on multimodality imaging studies, no evidence of systemic inflammatory response syndrome criteria, and negative serial blood culture results for evidence of bacteremia, Duke's criteria for infective endocarditis were rejected, and further work-up, including positron emission tomography/CT, was deferred.

FIGURE 1 Transthoracic Echocardiography



Transthoracic echocardiography parasternal long-axis view demonstrating bioprosthetic valvular dehiscence **(arrow)** and paravalvular mitral regurgitation. LA = left atrium; LV = left ventricle; LVOT = left ventricular outflow tract; PMV = prosthetic mitral valve; RV = right ventricle.

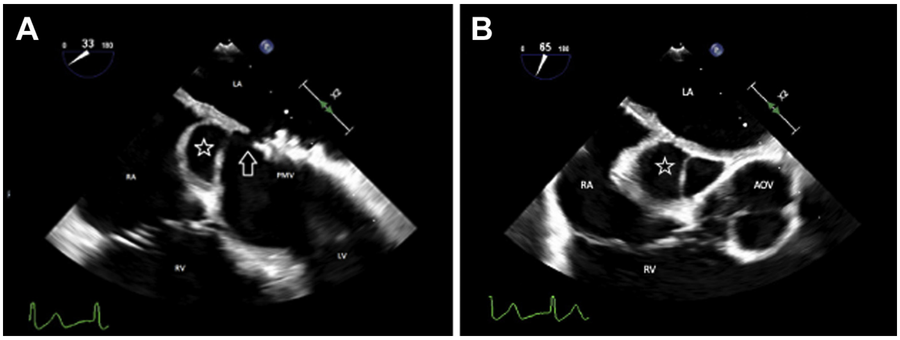
MANAGEMENT

Given the presence of a membranous septum pseudoaneurysm leading to bioprosthetic dehiscence and symptomatic paravalvular MR, the patient required structural repair of the pseudoaneurysm and dehisced valve. Although percutaneous closure was considered, the patient chose to undergo repeat MVR with pseudoaneurysm repair given her age and to limit potential residual outpouching near the LV outflow tract that might remain despite successful plugging. Intraoperatively, the cause of the dehisced bioprosthetic valve was confirmed to be secondary to the 3-cm thin-walled pseudoaneurysmal pouch associated with an anterior defect and subsequent paravalvular leak with no additional evidence of intravalvular pathologic changes. After removal of the dehisced bioprosthetic valve and its pledgets, a new #31 stented bioprosthetic porcine valve was inserted into the level of the true valvular annulus. The outpouching was ligated, eliminating the pseudoaneurysm altogether. Surgical pathology demonstrated no acute intravalvular pathologic changes (**Figure 8**). Postoperatively, the patient's course was unremarkable, with no significant findings on subsequent echocardiography. The patient's condition clinically improved, and she was discharged home.

DISCUSSION

Left ventricular pseudoaneurysm is a rare but serious complication after MVR.[1-4] Unlike true aneurysms, pseudoaneurysms are enclosed within the

FIGURE 2 **2-Dimensional Transesophageal Echocardiography**

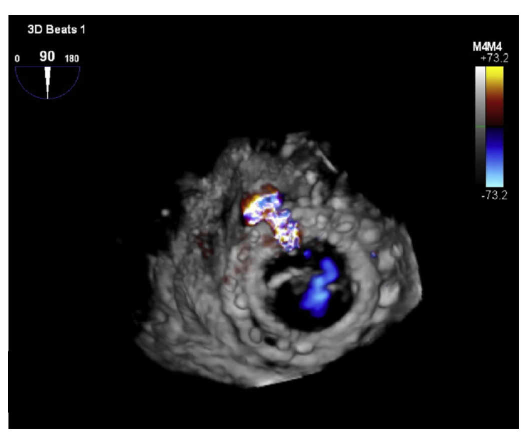


Transesophageal echocardiography illustrating pseudoaneurysm **(star)** as the mechanism of the bioprosthetic valvular dysfunction. **(A)** Midesophageal 5-chamber view. **(B)** Short-axis view at the level of the aortic valve. **Arrow** = valvular dehiscence. AOV = aortic valve; RA = right atrium; other abbreviations as in Figure 1.

pericardium by pericardial adhesions or thrombus.[5] Although their cause remains unclear, pseudoaneurysms may develop whenever there is early separation of the mitral annulus from the fibrous skeleton of the heart.[1] In a previous systematic literature review of 290 patients, myocardial infarction (55%), surgery (33%), and trauma (7%) were the top 3 associations with the development of ventricular pseudoaneurysms.[2] Previous authors have described that reoperation, endocarditis, and valve oversizing have been reported to predispose to its formation.[6]

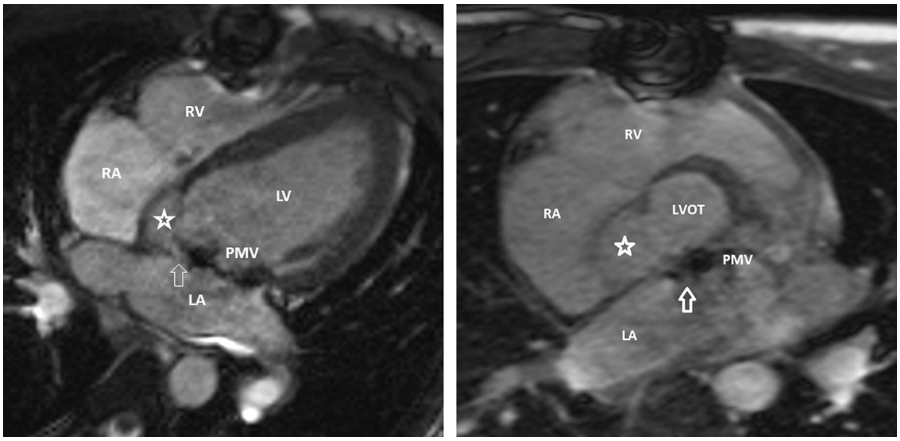
Early diagnosis is of paramount importance for the treatment and prognosis of patients with left ventricular pseudoaneurysm. Recently, the use of multimodality imaging, including transthoracic echocardiography, TEE, CT, and CMR, has superseded the previous gold standard of left ventricular angiography.[7] Specifically, transthoracic echocardiography and TEE have been described as 26% and 75% effective, respectively, in the definitive diagnosis of pseudoaneurysm.[8] Multidetector CT, given its enhanced spatial, contrast, and temporal resolution, has given rise to multiplanar reconstruction, allowing providers to accurately display anatomic and functional information regarding myocardial defects, including disclosing the size and location of LV pseudoaneurysms.[9,10] Specifically, the isotropic reconstruction of CT (compared with the anisotropic reconstruction of CMR) allows for easier manipulation of 3-dimensional multiplanar reconstructions without distortion. In the setting of defects at nonconventional locations or eccentric angulations, this trait may be of significant benefit. Additionally, the ability of CT to delineate the size of the pseudoaneurysm neck provides imperative information that may be difficult to ascertain from echocardiography and contrast ventriculography. Whereas CT may be preferable to CMR in these patients because of to its 3-dimensional imaging, CMR can also be used in these patients, given its high spatial resolution, tissue characterization, and multiplanar acquisition. CMR is also highly useful in the diagnosis of pseudoaneurysm, with reported sensitivity and specificity of 100% and 83%, respectively.[11] Some series have described that delayed pericardial enhancement is another frequent finding in patients with pseudoaneurysm, only visualized with CMR.[12]

FIGURE 3 **3-Dimensional Transesophageal Echocardiography**



Three-dimensional echocardiography from the left atrial perspective depicting paravalvular mitral regurgitation through the anteriorly located dehiscence.

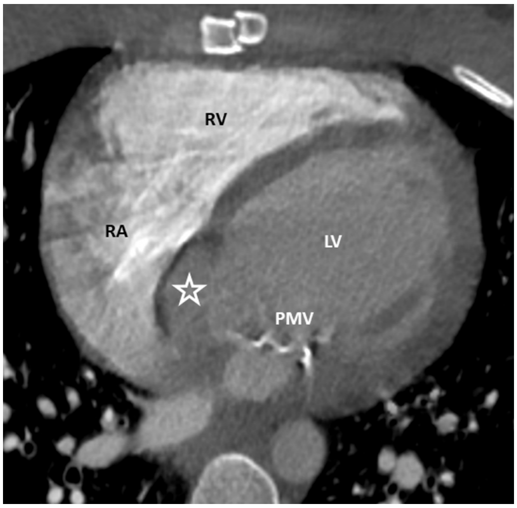
FIGURE 4 Cardiac Magnetic Resonance Imaging Cinematic Sequences



Cardiac magnetic resonance steady-state free precession horizontal long-axis view **(left)** and basal short-axis view **(right)** of the left ventricular outflow tract exhibiting the spatial relationship between the bioprosthetic valve, left ventricular pseudoaneurysm **(star)**, and left ventricular outflow tract. **Arrow** = valvular dehiscence. Abbreviations as in Figures 1 and 2

The primary goal of therapies for pseudoaneurysm is to reduce the frequency of expansion and risk of rupture, which previously have been estimated as high as 30% to 45%.[5] Current practice patterns highly recommend prompt surgical management of pseudoaneurysm with primary closure or patch closure, especially in patients with the symptomatic and acute disease.[13] Although high operative mortality has been reported, improvements in surgical techniques have lowered perioperative mortality rates to ≤10%.[4-5] Moreover, it has been reported that rupture risk in pseudoaneurysms increases when they are managed with medical treatment alone.[14]

FIGURE 5 Contrast-Enhanced Axial Computed Tomography

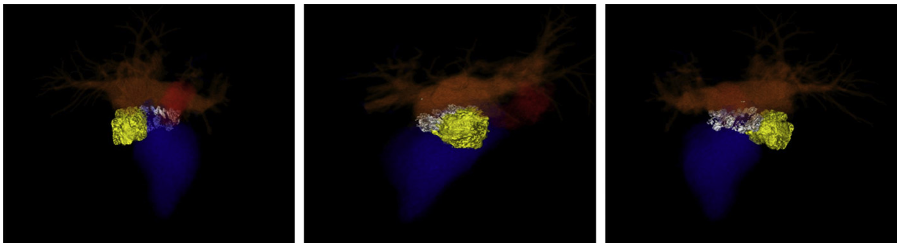


Contrast-enhanced axial computed tomography imaging validating the orientation between the bioprosthetic valve, left ventricular pseudoaneurysm **(star)**, and left ventricular outflow tract. Abbreviations as in Figures 1 and 2.

FIGURE 6 Contrast-Enhanced Coronal Computed Tomography

Contrast-enhanced coronal computed tomography imaging validating the orientation between the bioprosthetic valve, left ventricular pseudoaneurysm **(star)**, and left ventricular outflow tract. Abbreviations as in Figures 1 and 2.

FIGURE 7 3-Dimensional Computed Tomography Reconstructions



Rotated 3-dimensional computed tomography reconstructions demonstrating the spatial relationship between the left ventricular pseudoaneurysm **(yellow)**, stented bioprosthetic mitral valve **(white)**, left atrium **(orange)**, left ventricle **(blue)**, and aorta **(red)**.

For patients who are not candidates for surgical repair, percutaneous closure of pseudoaneurysms has also been performed.[15,16] Currently, the rarity of ventricular pseudoaneurysms has precluded randomized controlled trials comparing surgical and percutaneous repair.[17]

FOLLOW-UP

At 1 month, transthoracic echocardiography demonstrated normal left ventricular systolic function and only trivial MR with normal bioprosthetic function. At the 2-month follow-up visit, the patient's symptoms continued to resolve.

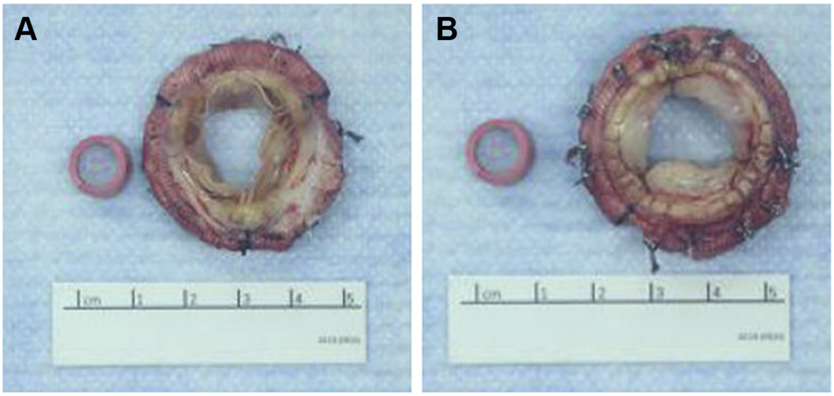
In conclusion, we present a rare case of symptomatic postoperative membranous septum pseudoaneurysm complicated by bioprosthetic mitral valvular dehiscence and paravalvular MR, diagnosed with multimodality imaging. With corrective surgical MVR and pseudoaneurysm repair, the patient's condition improved clinically, with stable echocardiographic findings.

FUNDING SUPPORT AND AUTHOR DISCLOSURES

Dr Kwan has received partial support from the Doris Duke Charitable Foundation Grant 2020059, New York, New York. All other authors have reported that they have no relationships relevant to the contents of this paper to disclose.

ADDRESS FOR CORRESPONDENCE: Dr Daniel S. Berman, Department of Imaging and Medicine, Cedars-Sinai Medical Center, 8700 Beverly Boulevard, Los Angeles, California 90048, USA. E-mail: Daniel.Berman@cshs.org.

FIGURE 8 Surgical Pathology



Surgical pathologic examination of the dehisced valve. **(A)** Ventricular aspect of the bioprosthesis. **(B)** Atrial aspect of the bioprosthesis.

REFERENCES

1. Ono M, Wolf RK. Left ventricular pseudoaneurysm late after mitral valve replacement. *Ann Thorac Surg*. 2002;73:1303-1305.

2. Hirasawa Y, Miyauchi T, Sawamura T, Takiya H. Giant left ventricular pseudoaneurysm after mitral valve replacement and myocardial infarction. *Ann Thorac Surg*. 2004;78:1823-1825.

3. Sanisoglu I, Duran C, Sagbas E, Akpinar B. A left ventricular pseudoaneurysm due to mitral valve replacement. *Eur J Cardiothorac Surg*. 2005;27:713.

4. Yeo TC, Malouf JF, Oh JK, Seward JB. Clinical profile and outcome in 52 patients with cardiac pseudoaneurysm. *Ann Intern Med*. 1998;128:299-305.

5. Frances C, Romero A, Grady D. Left ventricular pseudoaneurysm. *J Am Coll Cardiol*. 1998;32:557-561.

6. Namboodiri N, Dora SK, Thomas B, Misra M. Subannular left ventricular pseudoaneurysm following mitral valve replacement. *J Cardiothorac Surg*. 2008;3:28.

7. Bisoyi S, Dash AK, Nayak D, Sahoo S, Mohapatra R. Left ventricular pseudoaneurysm versus aneurysm a diagnosis dilemma. *Ann Card Anaesth*. 2016;19:169-172.

8. Kupari M, Verkkala K, Maamies T, Härtel G. Value of combined cross sectional and Doppler echocardiography in the detection of left ventricular pseudoaneurysm after mitral valve replacement. *Br Heart J*. 1987;58:52-56.

9. Ko SM, Hwang SH, Lee HJ. Role of cardiac computed tomography in the diagnosis of left ventricular myocardial diseases. *J Cardiovasc Imaging*. 2019;27:73-92.

10. Sharma RK, Gore R, Rosen BD, Arbab-Zadeh A. Diagnosis of left ventricular pseudoaneurysm by cardiac CT angiography. *J Cardiovasc Comput Tomogr*. 2014;8:246-247.

11. Gill S, Rakhit DJ, Ohri SK, Harden SP. Left ventricular true and false aneurysms identified by cardiovascular magnetic resonance. *Br J Radiol*. 2011;84:e35-e37.

12. Konen E, Merchant N, Gutierrez C, Provost Y, Mickleborough L, Paul NS, et al. True versus false left ventricular aneurysm: differentiation with MR imaging-initial experience. *Radiology*. 2005;236:65-70.

13. Atik FA, Navia JL, Vega PR, Gonzalez-Stawinski GV, Alster JM, Gillinov AM, et al. Surgical treatment of postinfarction left ventricular pseudoaneurysm. *Ann Thorac Surg*. 2007;83:526-531.

14. Si D, Shi K, Gao D, Yang P. Ruptured left ventricular pseudoaneurysm in the mediastinum following acute myocardial infarction: a case report. *Eur J Med Res*. 2013;18:2.

15. Dudiy Y, Jelnin V, Einhorn BN, Kronzon I, Cohen HA, Ruiz CE. Percutaneous closure of left ventricular pseudoaneurysm. *Circ Cardiovasc Interv*. 2011;4:322-326.

16. Yoo D, Robertson G, Block P, Babaliaros V, Lattouf O, Pernetz MA, et al. Percutaneous closure of a left atrial appendage pseudoaneurysm. *J Am Soc Echocardiogr*. 2011;24(109):e1-e3.

17. Hulten EA, Blankstein R. Pseudoaneurysms of the heart. *Circulation*. 2012;125:1920-1925.

KEY WORDS mitral valve replacement, multimodality imaging, paravalvular mitral regurgitation, pseudoaneurysm, valvular dehiscence

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