Left ventricular outflow tract pseudoaneurysms and severe paravalvular aortic regurgitation treated by percutaneous approach in a Marfan syndrome patient

Dean C. Paz BS, Timothy Hegeman DO, Onur Nadi Varli, Cihan Cevik MD

Abstract

A 66-year-old man with a history of Marfan syndrome and resolved infective endocarditis was found to have shortness of breath after a previous aortic valve replacement in 2010. Several severe paravalvular leaks were seen on imaging, and left ventricular outflow tract pseudoaneurysms were identified as the cause of his symptoms, which warranted treatment. Due to this patient's risk of surgical intervention, a retrograde transaortic approach was used.

Keywords: Marfan syndrome, infective endocarditis, paravalvular leaks, left ventricular outflow tract pseudoaneurysm, Amplatzer vascular plug

Introduction

A left ventricular outflow tract (LVOT) pseudoaneurysm carries significant risks, including severe aortic insufficiency, heart failure, rupture, compression of nearby vital structures, and death.[1] Early identification and diagnosis can be achieved with non-invasive imaging modalities, including transthoracic echocardiogram, transesophageal echocardiogram, and computed tomography angiogram. The cornerstone of medical management for patients with these pseudoaneurysms has been open heart surgery. In patients who are high risk surgical candidates, using alternative methods to close paravalvular leaks and LVOT pseudoaneurysms is imperative.

We present a case of a 66-year-old man with a history of Marfan syndrome and a previous diagnosis of infective endocarditis of the aortic valve followed by aortic valve replacement. The patient developed severe paravalvular leak and left ventricular outflow tract pseudoaneurysms after cardiac surgery. He was found to be at prohibitive risk for repeat surgical repair and was managed with transcatheter aortic paravalvular leak and LVOT pseudoaneurysm closure using a retrograde transaortic approach.

Corresponding author: **Dean Paz**
Contact Information: Dean.paz@rvu.edu
DOI: 10.12746/swrccc.v10i45.1081

Case

A 66-year-old man with a history of Marfan syndrome with previous, resolved, infective endocarditis required aortic valve replacement with a 27 mm Freestyle Delta conduit bioprosthesis in 2010. He also had replacement of his ascending aorta with a 24-mm Hemashield graft in the same procedure. The patient developed severe bioprosthetic aortic valvular and paravalvular regurgitation in 2022 and underwent valve in valve transcatheter aortic valve replacement (TAVR) with an Edwards Sapien S3 Ultra 26 mm valve. His transvalvular aortic regurgitation resolved post TAVR, but his severe paravalvular aortic regurgitation persisted. Figure 1 illustrates the patient's paravalvular leaks. Due to his complex medical history, he was found to be at prohibitive risk for redo sternotomy. Cardiac computed tomography scan also demonstrated multiple pseudoaneurysms arising from the left ventricular outflow tract (Figure 2). He reported

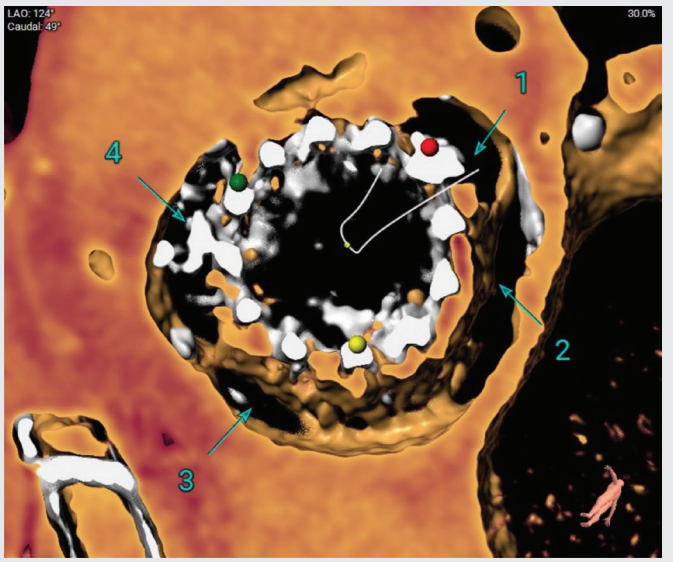

Figure 1. Transverse section through the aortic valve illustrating the multiple paravalvular leaks as indicated by each numbered arrow.

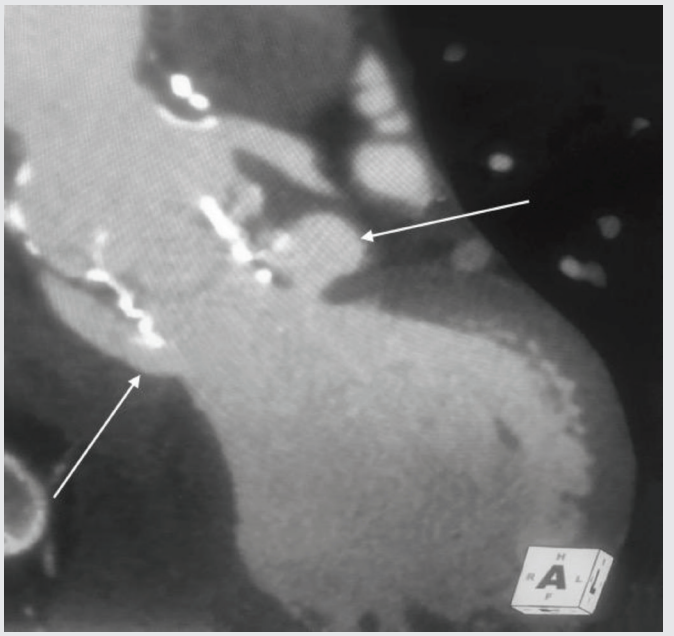

Figure 2. Cardiac computed tomography revealing two pseudoaneurysms laterally extending from the left ventricular outflow tract bilaterally.

shortness of breath with minimal activities, and he was classified as NYHA Class 3. The patient was seen by our cardiac service in this facility, and it was agreed that transcatheter closure of the LVOT pseudoaneurysm and paravalvular leaks should help reduce the symptoms impairing his daily activities.

Procedure

With the guidance of the transesophageal echocardiogram, a femoral arterial access was utilized for the retrograde paravalvular leak closure. A 5 Fr multipurpose catheter was telescoped within a 6F AL-1 guide catheter, and the paravalvular leak was crossed with a 0.035 Glidewire (Boston Scientific, Marlborough, MA). The wire was exchanged for an extra-stiff soft tip Amplatz wire in the left ventricle. A 110 cm shuttle sheath (Cook, Bloomington, IN) was then advanced into the left ventricular cavity through the pseudoaneurysm over this wire. The neck of the anterolateral pseudoaneurysm was then occluded using an Amplatzer II 16 mm vascular plug (Abbott, Abbott Park, IL) with TEE guidance. Similar steps were performed for the posterior pseudoaneurysm which was then closed using an Amplatzer II 10 mm vascular plug (Abbott), as shown in Figure 3. Postprocedural TEE demonstrated significant reduction from severe to mild paravalvular aortic regurgitation.

Discussion

In this report, we describe the management of a 66-year-old man with Marfan syndrome, who developed left ventricular outflow tract pseudoaneurysms which caused severe paraprosthetic aortic regurgitation following infective endocarditis and aortic valve replacement 12 years ago. He underwent successful transcatheter paravalvular leak closure by vascular plugs since he was not an appropriate candidate for surgical repair.

A left ventricular outflow tract pseudoaneurysm is a pooling of blood into a newly formed cavity, which can result secondary to blood flow at high velocities. Left ventricular outflow tract pseudoaneurysms can also develop secondary to infective endocarditis,

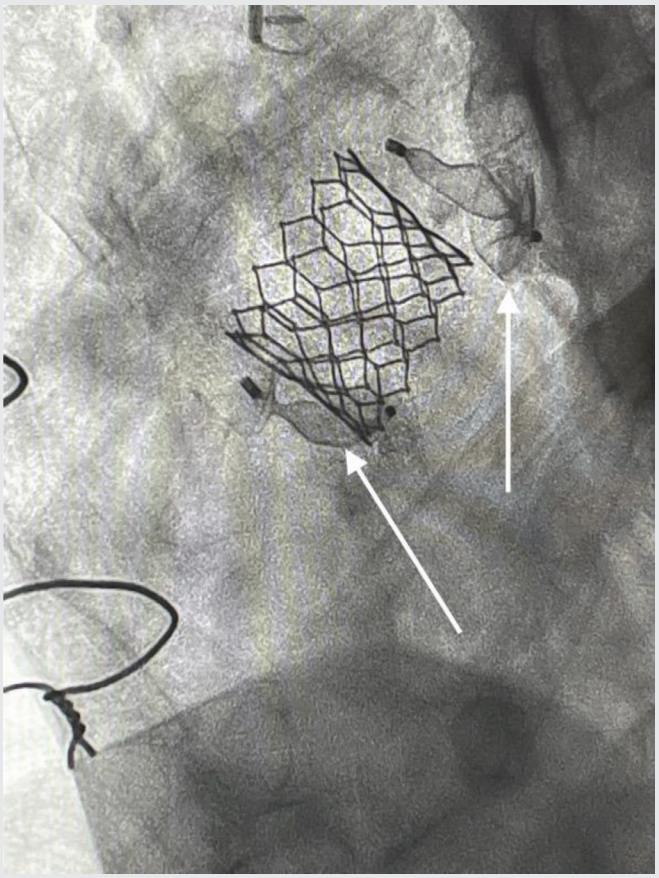

Figure 3. Post-procedural coronary angiogram revealing placement of the Amplatzer II 16 mm vascular plug (above) and Amplatzer II 10 mm vascular plug (below).

suture dehiscence post-aortic valve repair, and other surgical interventions directed towards the aorta.[2] They are more common in patients with connective tissue disorders such as in our patient with Marfan syndrome. His history of infective endocarditis and prior aortic valve replacement could have also contributed to his pseudoaneurysm formation. Detecting pseudoaneurysms is important to minimize the risk of developing rupture, embolism, compression to nearby structures, and infection.[1] While many patients with an LVOT pseudoaneurysm may be asymptomatic, others can present with symptoms similar to those experienced with coronary artery disease secondary to compression of the main coronary arteries.[3] LVOT pseudoaneurysms can also often cause aortic regurgitation, which may lead to left ventricular enlargement, systolic dysfunction and congestive heart failure.

The primary treatment of LVOT pseudoaneurysms and associated aortic regurgitation is surgical repair. Surgical techniques include sutures to close the pseudoaneurysm that are reinforced by Teflon felt. Another method involves the use of xenopericardium to close off the defect yielding the pseudoaneurysm.[4] However, some of these patients are not suitable candidates for surgical intervention. With the newly developing technology and techniques, transcatheter LVOT pseudoaneurysm exclusion is achievable. Transcatheter LVOT pseudoaneurysm closure could be performed with an antegrade technique (transseptal), retrogradely (transaortic), and transapical approaches by utilizing vascular plugs, coils, Amplatzer septal occluders, and VSD occluders with TEE guidance.[5,6] Our case is an illustration of large, multiple LVOT pseudoaneurysms treated via retrograde approach percutaneously. Our patient's shortness of breath improved dramatically in his one-month follow-up.

In conclusion, transcatheter treatment of LVOT pseudoaneurysms and associated paravalvular leak closure is an alternative and feasible option to a select cohort of patients who are prohibitive or at high risk for surgical repair.

Article citation: Paz DC, Hegeman T, Varli ON, Cevik C. Left ventricular outflow tract pseudoaneurysms and severe paravalvular aortic regurgitation treated by percutaneous approach in a Marfan syndrome patient. The Southwest Respiratory and Critical Care Chronicles 2022;10(45):71–74
From: College of Osteopathic Medicine (DCP), Rocky Vista University, Parker, Colorado; Department of Medicine-Cardiology (TH, CC), UC Health Memorial Hospital Central, Colorado Springs, Colorado; Uludag University School of Medicine (ONV), Bursa, Turkey
Submitted: 9/9/2022
Accepted: 10/9/2022
Conflicts of interest: none

References

1. Takayama, H., Otsuka, T., Kubota, H. et al. Pseudoaneurysm developed after aortic root homograft implantation. Jpn J Thorac Caridovasc Surg. 2004;52,158–160. https://doi.org/10.1007/s11748-004-0135-7
2. Gelehrter S, Wright G, Gless T, et al. Left ventricular outflow tract pseudoaneurysms in congenital heart disease. Am J Cardiol. 2002;90(7):806–809. doi:10.1016/s0002-9149(02)02621–8
3. Shariff MA, Martingano D, Khan U, et al. Left ventricular outflow tract pseudoaneurysm after aortic valve replacement. Aorta (Stamford). 2015 Oct 1;3(5):167–71. doi: 10.12945/j.aorta.2015.15.001.
4. Prêtre R, Linka A, Jenni R, et al. Surgical treatment of acquired left ventricular pseudoaneurysms. Ann Thorac Surg. 2000;70(2):553–557. doi:10.1016/s0003-4975(00)01412–0
5. Kar B, Gholkar G, Gregoric ID, et al. Percutaneous closure of a left ventricular pseudoaneurysm in a high-risk surgical candidate. Tex Heart Inst J. 2012;39(5):680–682.
6. Romaguera R, Slack MC, Waksman R, et al. IMAGE CARDIO MED: Percutaneous closure of a left ventricular outflow tract pseudoaneurysm causing extrinsic left coronary artery compression by transseptal approach. Circulation. 2010;121(4):e20–e22. doi: 10.1161/CIR.0b013e3181cf2fe2