PERCUTANEOUS PERILS

Aorto-Right Ventricular Fistula Post-Transcatheter Aortic Valve Replacement: Multimodality Imaging of Successful Percutaneous Closure

Alan F. Vainrib, MD, Homam Ibrahim, MD, Kazuhiro Hisamoto, MD, Cezar S. Staniloae, MD, Hasan Jalilahwii, MD, Ricardo J. Benenstein, MD, Larry Latson, MD, Mathew R. Williams, MD, and Muhamed Saric, MD, PhD, New York, New York

INTRODUCTION

Transcatheter aortic valve replacement (TAVR) has transformed aortic stenosis therapy, with greater than 200,000 procedures performed worldwide since the first case was performed by the French interventional cardiologist Alain Cribier in 2002. One rare but dreaded complication is aortic periannular rupture, seen in less than 1% of all TAVR procedures. Although TAVR is a safe procedure, with an average national in-hospital mortality of less than 4%, complications may still occur.

Aortic periannular rupture post-TAVR refers to a spectrum of injuries that occur in the aortic root and left ventricular outflow tract (LVOT) region and portend a poor prognosis. It has been described as having four types: supra-annular, intra-annular, subannular, and combined. These ruptures often lead to flow of blood into the pericardial space, which may lead to pericardial tamponade.

We describe a case of aorto-right ventricular (RV) fistula post-TAVR that is unique due to its flow pathway, Doppler findings, and lack of rupture into the pericardial space. Using a multimodality imaging approach for diagnosis and preprocedure planning, the rupture defect was successfully eliminated by percutaneous implantation of one Amplatz vascular plug (AVP) intravenous (IV) device (St. Jude Medical, St. Paul, MN). To our knowledge, this is the first reported case of the successful use of a plugging device for this type of disease.

CASE PRESENTATION

A 91-year-old man with severe symptomatic high-gradient aortic stenosis (American College of Cardiology/American Heart Association Stage D1) underwent successful percutaneous transcatheter implanta-

From the Leon H. Charney Division of Cardiology (A.F.V., H.I., K.H., C.S.S., H.J., R.J.B., M.R.W., M.S.), Department of Cardiothoracic Surgery (H.I., K.H., H.J., M.R.W.), and Department of Radiology (L.L.), New York University Langone Medical Center, New York, New York.

Keywords: Transcatheter echocardiography, 3D transesophageal echocardiography, Multidetector computed tomography, Percutaneous closure, TAVR

Conflicts of Interest: Dr. Mathew R. Williams is a consultant for and received research funding from Medtronic. Dr. Muhamed Saric is a member of a Philips and Medtronic speakers’ bureau. The remaining authors had no actual or potential conflicts of interest relative to this document.

Copyright 2017 by the American Society of Echocardiography. Published by Elsevier Inc. This is an open access article under the CC BY-NC-ND license (http://creativecommons.org/licenses/by-nc-nd/4.0/)

DISCUSSION

Although rare, aortic periannular rupture TAVR can be life threatening. The diagnosis of periannular rupture is typically made by 2D and Doppler echocardiography at the time of implantation, which can also detect associated complications such as pericardial effusion with tamponade.

The current classification scheme of periannular rupture is based on its location relative to the aortic annulus: above (supra-annular), below (subannular), within (annular), or a combination (combined). However, determining the exact location of rupture can be difficult and often requires advanced imaging such as cardiac computed tomography, magnetic resonance imaging, and 3D echocardiography to provide maximal spatial resolution for defect visualization.

Postrupture, fistulous flow may occur along the following pathways:

1. From the aorta at the level of the sinuses to the lower pressure right atrium (RA) or RV, causing systolic and diastolic (continuous) flow tracings on spectral Doppler.
2. From the LVOT to the RVOT, causing systolic-only flow on spectral Doppler similar to ventricular septal defects.
3. From the aorta at the level of the sinuses to the pericardium.
4. Some combination of the above three types.
Figure 1 Aorto-RV fistula: TTE. (A, B) Post-TAVR TTE with color flow Doppler in the parasternal view demonstrates aorto-RV fistula with fistulous flow (arrow) from the aortic annular area to the RVOT in the long axis (A) and short axis (B). (C) Continuous wave spectral Doppler demonstrates unusual, mostly high-velocity systolic and diastolic flow between the aortic annulus and RVOT. This flow pattern argues against a ventricular septal defect where the high velocity flow would have been in systole only. CW, Continuous wave; LA, left atrium; PLAX, parasternal long axis; SAX, short axis.

Figure 2 Aorto-RV fistula: TEE. (A, B) Post-TAVR TEE in the deep transgastric view demonstrates aorto-RV fistula represented by an echolucent channel (arrow) between the TAVR prosthesis and the RVOT on B mode (A) and color Doppler imaging (B). (C) Mid-esophageal short-axis view with color Doppler demonstrates fistulous flow from the aortic root into the RVOT. Note that on TEE, short-axis visualization of the defect may be more difficult than on TTE since the anteriorly located annular rupture is partly shadowed by the posteriorly located TAVR valve on TEE. This added to the challenge of TEE guidance of percutaneous closure. LA, Left atrium; PA, pulmonary artery; PV, pulmonic valve; SAX, short axis.
In this case, fistulous flow from the aorta at the SOV to the RVOT is demonstrated by the presence of systolic and diastolic spectral Doppler signals. Although conceptually this defect is similar to aneurysm-associated SOV rupture, the spectral Doppler tracings appear unique. In SOV rupture, spectral velocities typically do not drop to baseline, while in our case there was distinct cessation of velocities in late diastole (Figure 1C). A perimembranous ventricular septal defect was also in the differential diagnosis. However, with a ventricular septal defect, the high flow velocities would have been seen in systole only.

High-resolution computed tomography with multiplanar reconstruction points to an entry point defect into the RV at the level of the aortic annulus. Two possibilities may explain this—either the defect is slightly above the aortic annulus or the defect is at or below the aortic annulus but the TAVR skirt creates a seal between the LVOT and the aorta, permitting flow only between the SOV and RVOT.

This case not only demonstrates a unique pathophysiologic mechanism but also a unique treatment approach. Although percutaneous valve-in-valve implantation has reportedly been used to seal post-TAVR annular rupture, ours appears to be the first reported case of a successful elimination of a post-TAVR aorto-RV fistula using a vascular plug. Risk factors for post-TAVR aorto-RV fistula in our patient included heavy calcifications of the native aortic valve and LVOT as well as postdilatation of a self-expanding TAVR valve.

**CONCLUSION**

As the use of TAVR as a treatment for severe aortic stenosis grows, the likelihood of encountering rare but life-threatening complications such as aortic annular rupture becomes greater. As such, multimodality imaging is critical in diagnosing aortic root injury and its
complications, as well as in preprocedure planning for percutaneous therapies.

In summary, this case demonstrates the use of multimodality imaging in the diagnosis of an aorta-RV fistula post-TAVR, a unique disease on the aortic annular rupture spectrum that allowed for an innovative and effective therapeutic approach. The fistula likely developed immediately following postdilatation of the TAVR valve, and its clinical significance was fully recognized 4 months later when the patient presented with congestive heart failure. Risk factors for post-TAVR annular rupture in our patient included heavy calcifications of the native aortic valve and LVOT as well as postdilatation of a self-expanding TAVR valve.

SUPPLEMENTARY DATA

Supplementary data related to this article can be found at http://dx.doi.org/10.1016/j.case.2017.02.002.

REFERENCES

1. Vahl TP, Kodali SK, Leon MB. Transcatheter aortic valve replacement 2016: a modern-day “through the looking-glass” adventure. J Am Coll Cardiol 2016;67:1472-87.
2. Genereux P, Head SJ, Van Mieghem NM, Kodali S, Kirtane AJ, Xu K, et al. Clinical outcomes after transcatheter aortic valve replacement
using valve academic research consortium definitions: a weighted meta-analysis of 3,519 patients from 16 studies. J Am Coll Cardiol 2012;59:2317-26.
3. Edwards FH, Cohen DJ, O’Brien SM, Peterson ED, Mack MJ, Shahian DM, et al. Development and validation of a risk prediction model for in-hospital mortality after transcatheter aortic valve replacement. JAMA Cardiol 2016;1:46-52.
4. Pasic M, Unbehaun A, Buz S, Drews T, Hetzer R. Annular rupture during transcatheter aortic valve replacement: classification, pathophysiology, diagnostics, treatment approaches, and prevention. JACC Cardiovasc Interv 2015;8:1-9.
5. Aksoy O, Paixao AR, Marmagkiolis K, Mego D, Rollefson WA, Cilingiroglu M. Aortic annular rupture during TAVR: Mini review. Cardiovasc Revasc Med 2016;17:199-201.
6. Goldberg N, Krasnow N. Sinus of Valsalva aneurysms. Clin Cardiol 1990;13:831-6.
7. Saric M, Kronzon I. Ventricular septal defect and Eisenmenger syndrome. In: Lang R, Goldstein SA, Kronzon I, Khanderia BK, editors. Dynamic Echocardiography: A Case-Based Approach. Springer; 2010:446-50.
8. Yu Y, Vallely M, Ng MK. Valve-in-valve implantation for aortic annular rupture complicating transcatheter aortic valve replacement (TAVR). J Invasive Cardiol 2013;25:409-10.

Figure 6 Two-dimensional TEE of aorto-RV fistula closure: final result. (A) Biplane TEE demonstrating a well-seated AVP 4 device on both sides of the aorto-RV fistula defect. (B) Color flow Doppler confirms no residual flow across the prior site of aorto-RV fistula. LV, Left ventricle; PA, pulmonary artery.