Severe Aortic Regurgitation From Sinus of Valsalva Aneurysm Prolapse

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INTRODUCTION

Sinus of Valsalva aneurysm (SVA) results from weakness of the elastic lamina at the junction of the aortic media and annulus fibrosus. Sinus of Valsalva aneurysm is usually asymptomatic until rupture. We present a case of an unruptured SVA causing severe aortic regurgitation and decompensated heart failure.

CASE PRESENTATION

A 32-year-old man with no significant medical history presented with several months of shortness of breath and bilateral lower extremity edema. On admission, temperature was 37.7°C, blood pressure was 152/76 mm Hg, heart rate was 114 beats/min, and oxygen saturation was 98% on room air. Physical examination revealed jugular venous distention, a systolic murmur at the apex, a diastolic murmur at the sternal border, diminished bilateral breath sounds, and bilateral +1 lower extremity edema.

The patient's history and physical examination suggested new-onset heart failure. The differential diagnosis in a young patient with heart failure includes toxin-induced cardiomyopathy, myocarditis, endocarditis, hypertrophic cardiomyopathy, infiltrative heart disease, congenital heart disease, severe valvular stenosis or regurgitation, or pulmonary embolism. Other potential noncardiac causes of the patient's presentation include cirrhosis, renal failure, and primary pulmonary diseases.

The electrocardiogram on admission showed sinus tachycardia and left ventricular hypertrophy. Pertinent laboratory values are listed in Table 1. Computed tomography angiogram of the chest showed cardiomegaly, pulmonary venous congestion, pulmonary edema, bilateral pleural effusions (right greater than left), and a calcified granuloma in the right upper lobe. The transthoracic echocardiogram (TTE) showed left ventricular cavity dilation (diastolic dimension 6.6 cm, systolic dimension 5.6 cm), impaired biventricular systolic function with a left ventricular ejection fraction <20%, global hypokinesis, and a large unruptured SVA with diastolic prolapse into the left ventricular outflow tract over the right coronary cusp (Figure 1, Videos 1 and 2). There was visually estimated severe aortic regurgitation (Figures 1 and 2, Videos 2 and 3), severe mitral regurgitation (Video 4), and moderate tricuspid regurgitation with elevated right ventricular systolic pressure (63 mm Hg). Right heart catheterization findings are listed in Table 2.

The patient was treated with intravenous diuretics and continuous intravenous milrinone. In the first 8 days of admission, the patient achieved a cumulative net fluid balance of −22 L, and the cardiac index improved to an estimated 3.2 L/min/m^2. Infectious disease specialists were consulted to evaluate the patient in light of the positive interferon-gamma release assay. The patient underwent bronchoscopy with bronchoalveolar lavage. Two acid

VIDEO HIGHLIGHTS

Video 1: Two-dimensional TTE, parasternal long-axis view before surgery, demonstrates the dilated left ventricle with severe systolic dysfunction and the SVA with prolapse through the aortic valve.

Video 2: Two-dimensional TTE, parasternal long-axis view with color-flow Doppler before surgery, demonstrates the SVA with prolapse and severe aortic regurgitation.

Video 3: Two-dimensional TTE, parasternal short-axis view, without (*left*) and with (*right*) color-flow Doppler at the level of the aortic valve before surgery, demonstrates the severe aortic regurgitation.

Video 4: Two-dimensional TTE, zoomed apical 4-chamber view, without (*left*) and with (*right*) color-flow Doppler before surgery, demonstrates the normal mitral valve with systolic tenting and severe secondary mitral regurgitation.

Video 5: Two-dimensional transesophageal echocardiogram, midesophageal aortic valve long-axis (122°) view, demonstrates the SVA prolapsing into the left ventricular outflow tract in diastole. The image is overgained likely due to the intraoperative environment with bright operating room lights.

Video 6: Two-dimensional TTE, parasternal long-axis view following surgery, demonstrates the bioprosthetic aortic valve and aortic root repair. Also noted is the mitral valve repair with annuloplasty ring.

Video 7: Two-dimensional TTE, parasternal long-axis view, with color-flow Doppler following surgery, demonstrates the normally functioning bioprosthetic aortic valve, aortic root repair, and mitral valve repair with residual mitral regurgitation.

View the video content online at www.cvcasejournal.com.

From the Cooper University Health Care, Camden, New Jersey.

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Table 1 Admission laboratory values

Laboratory test	Actual value	Normal value
N-terminal pro b-type natriuretic peptide	11,548 pg/mL	1-93 pg/mL
High-sensitivity troponin (0, 1, and 3 hours)	67, 67, 70 ng/L	0-19 ng/L
Creatinine	1.4 mg/dL	0.6-1.2 mg/dL
ALT	804 U/L	6-45 U/L
AST	602 U/L	10-35 U/L
Alkaline phosphatase	112 U/L	39-117 U/L
Bilirubin	1.2 mg/dL	0-1.2 mg/dL
HIV	Nonreactive	Nonreactive
RPR	Nonreactive	Nonreactive
Blood parasite smear	Negative	Negative
Blood cultures	Negative	Negative
Interferon-gamma release assay	Positive	Negative
Acute hepatitis panel	Negative	Negative

fast bacilli smears and cultures were negative. The specialists concluded that the patient had a latent tuberculosis infection (which was unlikely contributing to the clinical presentation) and recommended outpatient treatment.

Following medical stabilization in the cardiac intensive care unit, cardiothoracic surgery was consulted. After a heart team discussion that considered institutional experience with bioprosthetic versus mechanical valves in patients of this age and experience with valve-in-valve transcatheter aortic valve replacement, as well as the patient's unwillingness to take anticoagulation therapy, the patient underwent a biological Bentall procedure for aortic root replacement and bioprosthetic aortic valve replacement. The patient also underwent mitral valve repair with ring annuloplasty and reimplantation of the left and right main coronary arteries. Intraoperative transesophageal echocardiogram revealed an ejection fraction 20% to 25%, severe eccentric aortic regurgitation with a large mobile right coronary SVA, and moderate mitral regurgitation (Figure 3, Video 5). There was no significant mitral valve leaflet pathology, and the mitral regurgitation was attributed to left ventricular cavity dilation. Intraoperatively the aortic valve showed chronic inflammatory changes. The aortic valve tissue culture was negative for infectious etiology. The pathology report of the SVA and ascending aorta revealed focal myxoid degeneration.

One week after surgery, repeat TTE showed a left ventricular ejection fraction <20%, mitral valve annuloplasty ring with mild mitral regurgitation, normally functioning bioprosthetic aortic valve with no significant aortic regurgitation, and intact aortic root replacement (Figure 4, Videos 6 and 7). As the patient's hemodynamics improved, appropriate heart failure therapy was started. Six months later, repeat echocardiogram showed an ejection fraction of 15%. Given the patient's lack of improvement in ejection fraction despite adhering to guideline-directed medical therapy for heart failure with reduced ejection fraction, an implantable cardioverter-defibrillator was placed for primary prevention.

DISCUSSION

Unruptured SVA usually does not cause symptoms and may remain undetected.[1] However, in some cases unruptured SVA can impair nearby structures. Unruptured SVA can compress and obstruct the cardiac chambers, outflow tracts, great vessels, and coronary arteries. It can also cause valvular abnormalities, most commonly aortic regurgitation. It can ultimately lead to myocardial ischemia, heart failure, arrhythmias, and conduction defects.[2] Here we present a highly unusual case of an unruptured SVA that caused severe aortic regurgitation with severe left ventricular systolic failure and ultimately cardiogenic shock. Echocardiography showed that the SVA prolapsed through the aortic valve during diastole. This led to aortic regurgitation, left ventricular dilation, mitral regurgitation, and systolic dysfunction of the left ventricle. In this case, echocardiography was essential for establishing the cause of heart failure and the need for surgical management. Additionally, echocardiography was necessary to monitor for improvement in ejection fraction, which led to the decision to place an implantable cardioverter-defibrillator for primary prevention of sudden cardiac death.

The etiology of this patient's SVA remains unknown. Most cases of SVA are congenital. Pressure on the aortic root during embryological development can cause formation of a diverticulum that develops into SVA. Congenital SVA is associated with connective tissue diseases. Acquired SVA results from secondary weakening of the elastic connective tissue. Acquired SVA may occur as a result of connective

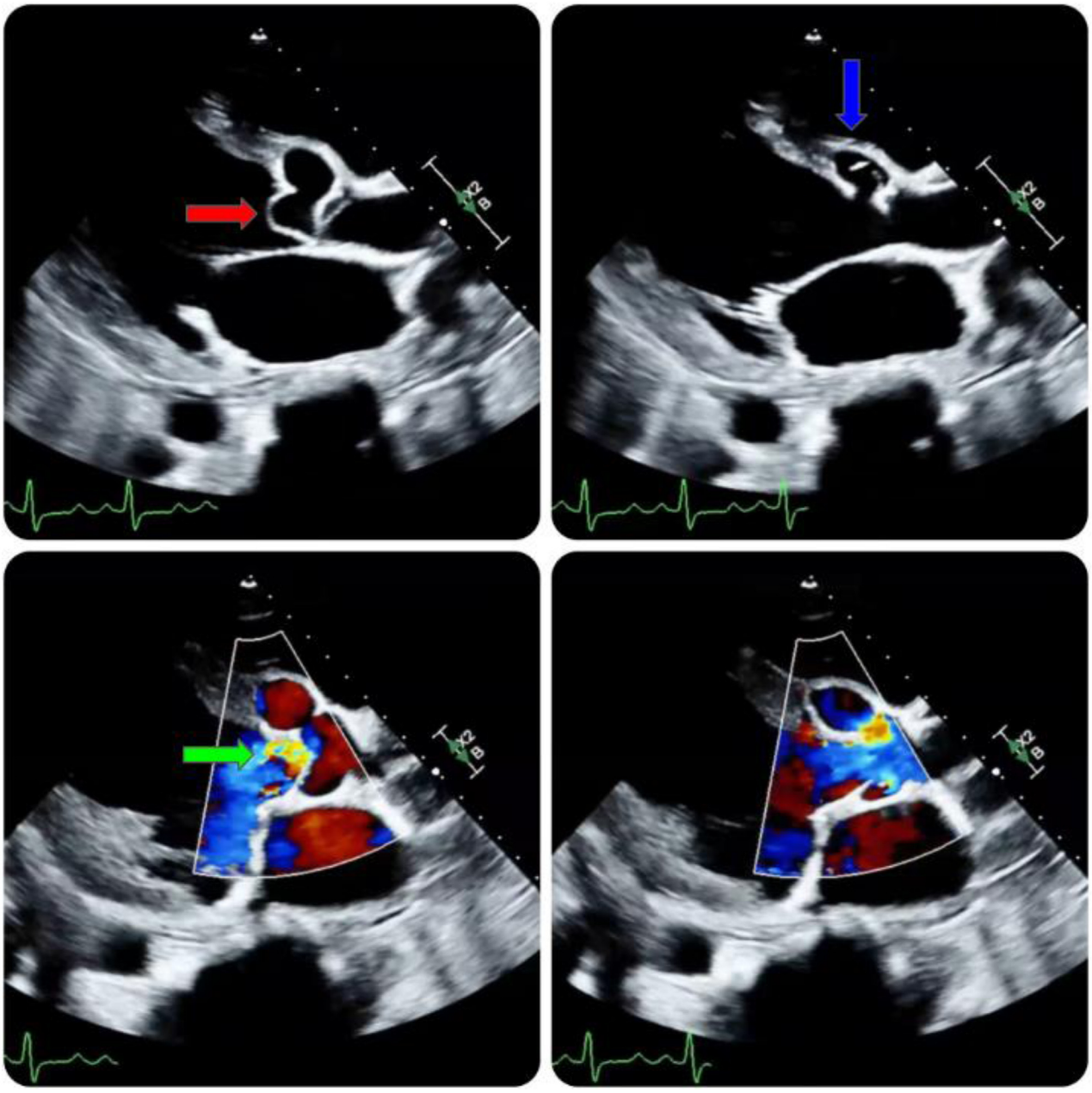

Figure 1 Two-dimensional TTE, parasternal long-axis view, without (*top*) and with (*bottom*) color-flow Doppler in diastole (*left*) and systole (*right*) demonstrates the SVA prolapsed into the left ventricle (*red arrow*), positioned in the right ventricular outflow tract (*blue arrow*) and aortic regurgitation (*green arrow*) without systolic obstruction.

tissue diseases, infections (such as tuberculosis, syphilis, and bacterial endocarditis), atherosclerosis, and trauma.[1] Although this patient was suspected to have latent tuberculosis, the infectious disease specialists concluded that tuberculosis likely did not cause the SVA, as there were no other signs of systemic tuberculosis. It was unclear whether this patient's SVA was congenital or acquired.

Ruptured SVA requires prompt surgical repair. Unruptured SVA should undergo repair if it is large or associated with significant symptoms.[1,2] Given the severity of the clinical picture, this patient underwent surgery for aortic root replacement, aortic valve replacement, and mitral valve repair. The patient initially required vasopressor and inotropic support but improved after surgery and initiated an

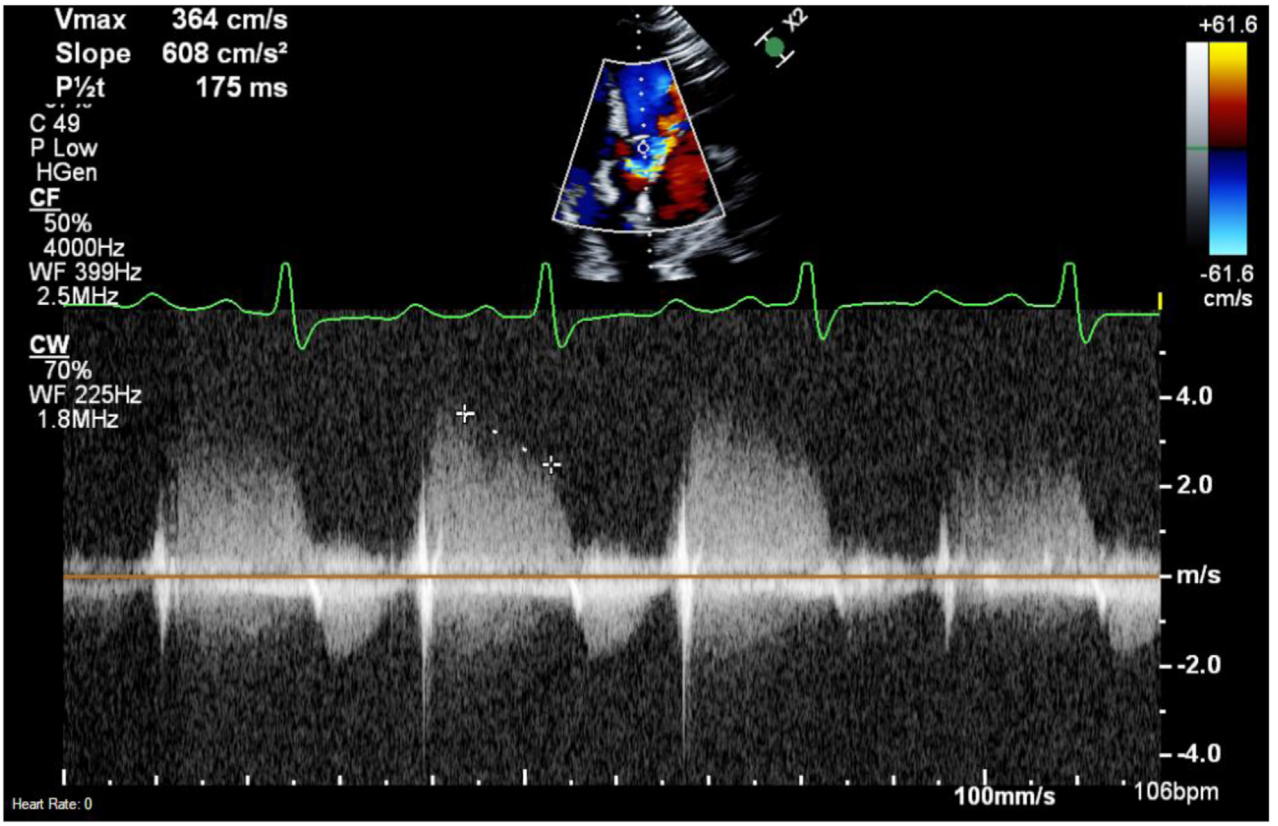

Figure 2 Two-dimensional TTE apical 5-chamber view with color-flow Doppler-guided continuous-wave spectral Doppler display demonstrates the short aortic pressure half time (175 msec).

Table 2 Right heart catheterization values

	Actual value	Normal value
Pulmonary capillary wedge pressure	51 mm Hg	6-15 mm Hg
Cardiac output	3.26 L/min	4-8 L/min
Cardiac index	1.9 L/min/m^2	2.8-4.2 L/min/m^2
Pulmonary artery systolic pressure	60 mm Hg	15-25 mm Hg

oral regimen of guideline-directed medical therapy for heart failure with reduced ejection fraction

CONCLUSION

Sinus of Valsalva aneurysm is a rare and challenging disease process. Although unruptured SVA is usually asymptomatic, this patient presented with an unruptured SVA causing severe aortic regurgitation, decompensated heart failure, and cardiogenic shock. Early surgical intervention was necessary to prevent worsening cardiogenic shock and ultimately death. Echocardiography was vital as it played a key role in diagnosis and management.

ETHICS STATEMENT

The authors declare that the work described has been carried out in accordance with The Code of Ethics of the World Medical Association (Declaration of Helsinki) for experiments involving humans.

CONSENT STATEMENT

Complete written informed consent was obtained from the patient (or appropriate parent, guardian, or power of attorney) for the publication of this study and accompanying images.

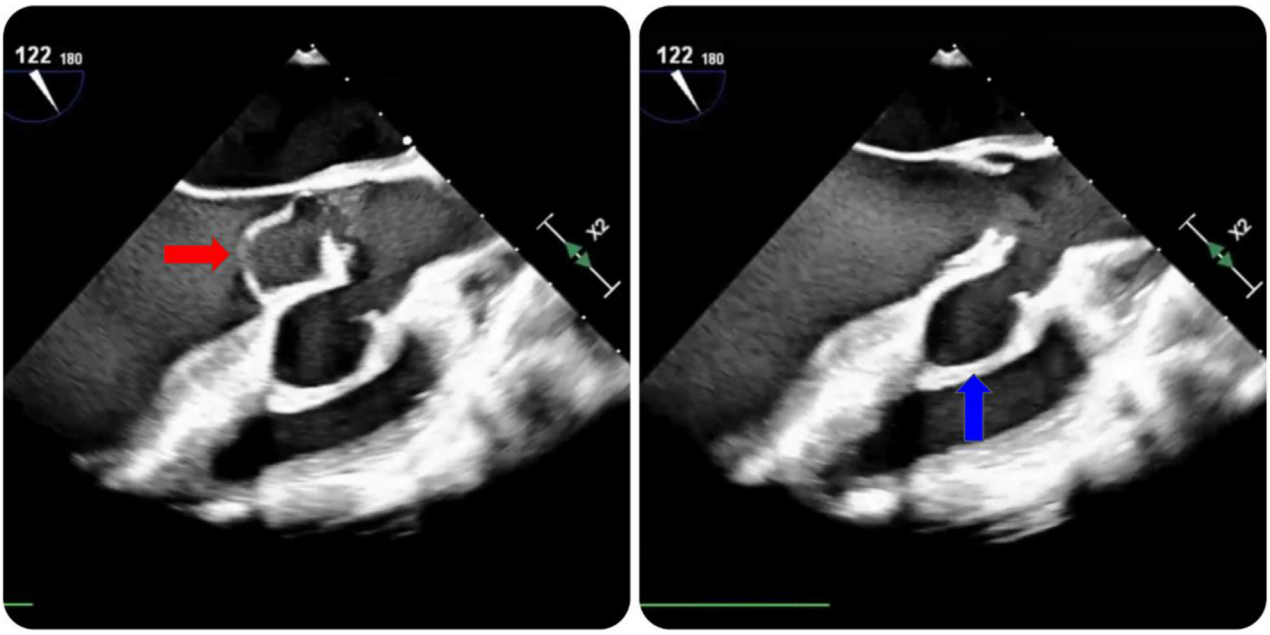

Figure 3 Two-dimensional transesophageal echocardiogram, midesophageal aortic valve long-axis (122°) view, demonstrates the SVA prolapsed into the left ventricular outflow tract in diastole (*left*, *red arrow*) and the SVA position during systole (*right*, *blue arrow*). The image is overgained likely due to the intraoperative environment with bright operating room lights.

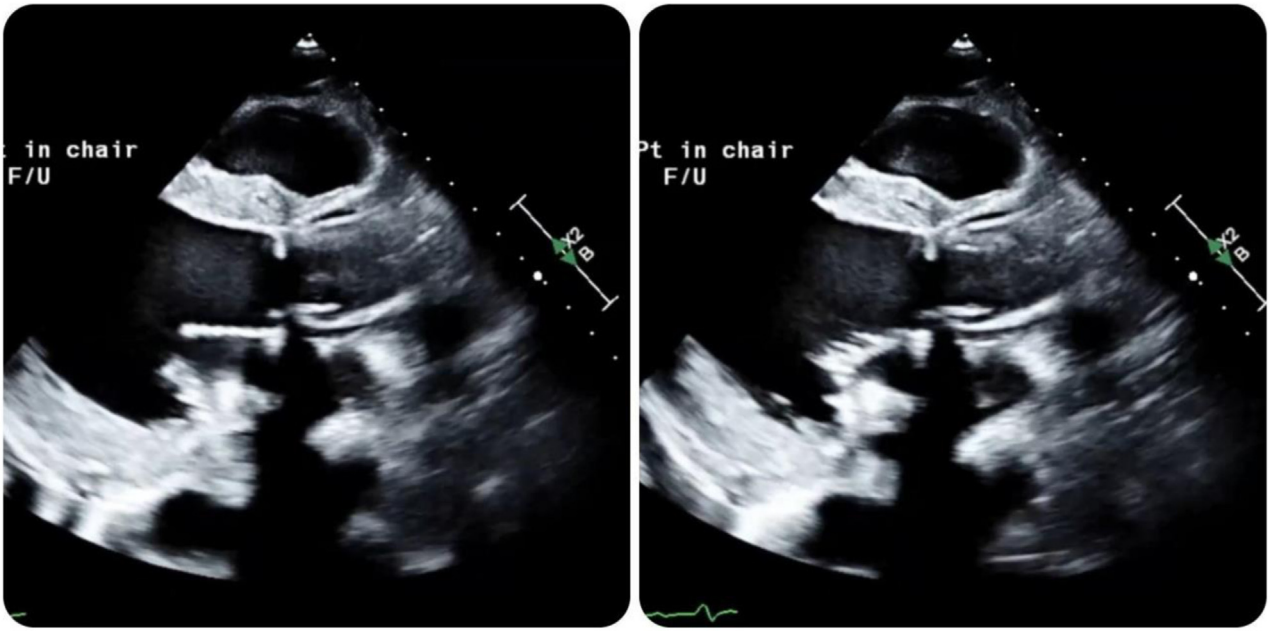

Figure 4 Two-dimensional TTE, parasternal long-axis view following surgery demonstrates the bioprosthetic aortic valve and aortic root repair in diastole (*left*) and systole (*right*). Also noted is the mitral valve repair with annuloplasty ring.

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DISCLOSURE STATEMENT

The authors report no conflict of interest.

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SUPPLEMENTARY DATA

Supplementary data to this article can be found online at https://doi.org/10.1016/j.case.2023.08.005.

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