Left Ventricular Pseudoaneurysm in an Adult With a Repaired Partial Atrioventricular Canal Defect

Kathryn Wershing, DO, Joseb Colon, BS, Jorge Alegria, MD, Joseph Paolillo, MD, Daniel Wallihan, MD, and Matthew Schwartz, MD, *Charlotte, North Carolina*

INTRODUCTION

We describe a 39-year-old man with a history of a partial atrioventricular canal (AVC) repair that presented with an embolic stroke and was found to have a left ventricular (LV) pseudoaneurysm in the lateral wall. Imaging showed an occluded left circumflex coronary artery (LCX), and it was hypothesized that circumflex injury occurred at the time of AVC repair, with resultant myocardial infarction and eventual development of pseudoaneurysm in the LCX territory. The patient was a poor candidate for surgical repair and was managed with chronic anticoagulation, with no recurrent events at 3-year follow-up.

CASE PRESENTATION

A 39-year-old man presented with sudden onset of right-sided weakness in the upper and lower extremities, aphasia, and facial droop. This patient had a history of trisomy 21 as well as a partial AVC defect (isolated primum atrial septal defect) that was repaired at 26 years of age with patch closure of the primum atrial septal defect and suture closure of the cleft in the left atrioventricular valve. The patient had never been seen at our center and had not seen a cardiologist in years. On physical examination, the patient was afebrile, with a heart rate of 88 beats/min, blood pressure of 126/69 mm Hg, and an oxygen saturation of 97% on room air. Cardiac examination showed a normal precordium with a regular rate and rhythm, normal S1, a physiologically split S2, and no murmurs. Neurologic examination showed significant aphasia with left-sided facial droop and right upper and lower extremity weakness (3/5 throughout both extremities). The left upper and lower extremities had normal strength. Urgent head computed tomographic angiography showed a thrombus in the left main cerebral artery. Blood supply was restored to the distal branches via suction thrombectomy of the left main cerebral artery and then intravenous alteplase therapy. Transthoracic echocardiography showed a large aneurysm affecting the lateral LV wall with possible thrombus (Figure 1, Videos 1-3). LV systolic function was mildly reduced and estimated at 45% by visual assessment, and mild mitral regurgitation was present. Cardiovascular magnetic resonance (CMR) was limited by inadequate breath holding but showed an estimated LV ejection fraction of 40% (volumes not obtained) and suspected pseudoaneurysm affecting the lateral LV wall. CMR showed an abrupt myocardial thickness "cutoff" at the neck of the aneurysm that was strongly suggestive of a pseudoaneurysm instead of a true aneurysm (Figure 2).[1] Late gadolinium enhancement imaging showed diffuse pericardial enhancement surrounding the area, which further supported a diagnosis of pseudoaneurysm.[2] Delayed contrast imaging also showed a transmural infarction of the inferolateral LV wall. Contrast-enhanced cardiac computed tomography (CCT) was then performed and showed a large pseudoaneurysm affecting the lateral LV wall measuring 9.7 × 6.0 cm, with an intramural thrombus (Figures 3 and 4). CCT also showed a small proximal LCX that appeared occluded as it entered the lateral atrioventricular groove (Figure 5). The left main, left anterior descending, and right coronary arteries were all without stenosis or calcification. It was hypothesized that the patient had experienced LCX injury during the prior incomplete AVC repair, with resultant transmural myocardial infarction and LV pseudoaneurysm in the affected LCX territory.

Surgical treatment of the LV pseudoaneurysm was not performed, because the base of the pseudoaneurysm was broad, making surgical resection challenging and risky. Surgical resection could potentially

VIDEO HIGHLIGHTS

Video 1: Two-dimensional transthoracic echocardiography (TTE), parasternal long-axis view, demonstrates the large inferolateral pseudoaneurysm and its relationship to the mitral valve and left atrium.

Video 2: Two-dimensional TTE, parasternal long-axis view, demonstrates mildly reduced LV systolic function with a large inferolateral pseudoaneurysm with dyskinetic motion.

Video 3: Two-dimensional TTE, parasternal long-axis view with color flow Doppler, demonstrates low-velocity flow from the LV pseudoaneurysm to the left ventricle in diastole and from the left ventricle to the LV pseudoaneurysm in systole.

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

From the Department of Pediatrics, Levine Children's Hospital, Atrium Health, Charlotte, North Carolina (K.W.); Congenital Cardiac 3D Printing Program, Levine Children's Hospital, Atrium Health, Charlotte, North Carolina (J.C.); Adult Congenital Heart Disease Program, Sanger Heart and Vascular Institute, Atrium Health, Charlotte, North Carolina (J.A., J.P., M.S.); Division of Pediatric Cardiology, Levine Children's Hospital, Atrium Health, Charlotte, North Carolina (J.P., M.S.); and Division of Radiology, Levine Children's Hospital, Atrium Health, Charlotte, North Carolina (D.W.).

Keywords: Left ventricle, Pseudoaneurysm, Atrioventricular canal

Correspondence: Matthew Schwartz, MD, Division of Pediatric Cardiology, Levine Children's Hospital, Atrium Heath, 1001 Blythe Boulevard, 5th Floor, Charlotte, NC 28203. (E-mail: *matthew.c.schwartz@atriumhealth.org*).

2468-6441

https://doi.org/10.1016/j.case.2023.12.009

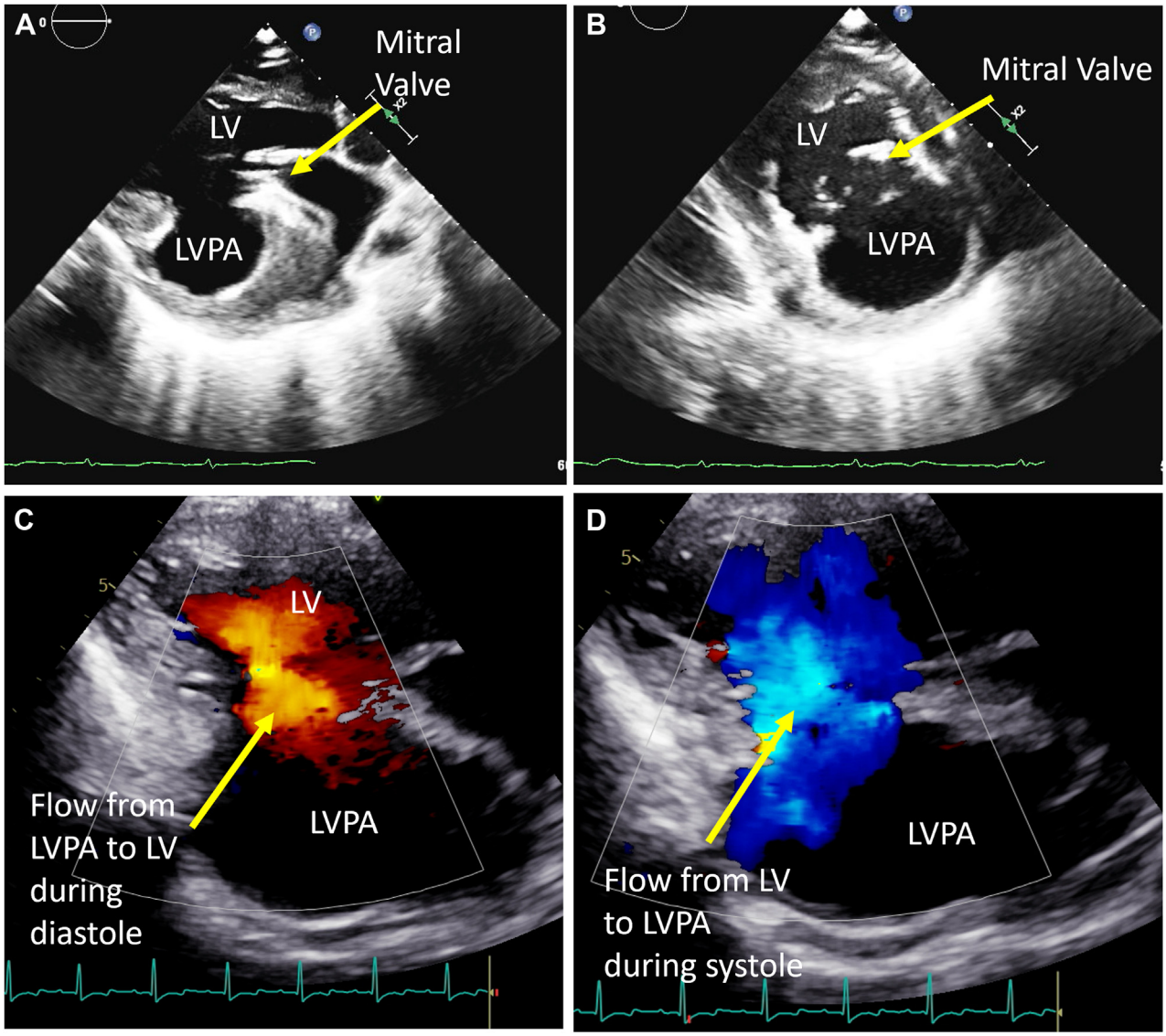

Figure 1 Two-dimensional transthoracic echocardiography (TTE), parasternal long-axis **(A)** and parasternal short-axis **(B)** diastolic views, demonstrates the left ventricle (LV), left ventricular pseudoaneurysm (LVPA), and mitral valve (*arrow*). Two-dimensional TTE, parasternal long-axis zoom view with color flow Doppler in diastole **(C)** and systole **(D)**, demonstrates to-and-fro low-velocity flow.

compromise the integrity of the LV lateral wall and cause systolic dysfunction. In addition, the pseudoaneurysm was felt to be old and chronic and thus associated with lower risk for rupture. Given the likely chronicity of the defect and the patient's embolic stroke with thrombus found in the pseudoaneurysm, the patient was anticoagulated with apixaban.

One month later, the patient's neurologic function had recovered, without any residual deficits. At follow-up 3 years later, the patient was doing well, with no recurrent events. Transthoracic echocardiography showed stable appearance of the pseudoaneurysm without intramural thrombus.

DISCUSSION

We describe the case of a 39-year-old man with a history of repaired partial AVC who presented with embolic stroke and was found to have a large LV pseudoaneurysm with intramural thrombus. An LV pseudoaneurysm results when rupture of the ventricular myocardium is contained by either the epicardial wall or pericardial adhesions. This is a rare complication following myocardial infarction, but it can also occur after cardiac surgery, trauma, or cardiac infection. Frances *et al.*[3] reviewed and summarized 273 patients with LV pseudoaneurysm reported in the literature. In their series, 55% were associated with myocardial infarction, 33% after prior cardiac surgery, 7% with cardiac trauma, and 5% with cardiac infection. Of the patients, 6% presented with stroke and 12% were asymptomatic.[3] Reports of LV pseudoaneurysms in patients with congenital heart disease also exist.[4]

Distinguishing a ventricular pseudoaneurysm from a true ventricular aneurysm is very important because an acute LV pseudoaneurysm is at a much higher risk for rupture. Both CMR and CCT can be very helpful in accurately diagnosing a ventricular pseudoaneurysm. On CMR or CCT, patients with pseudoaneurysms will often show the

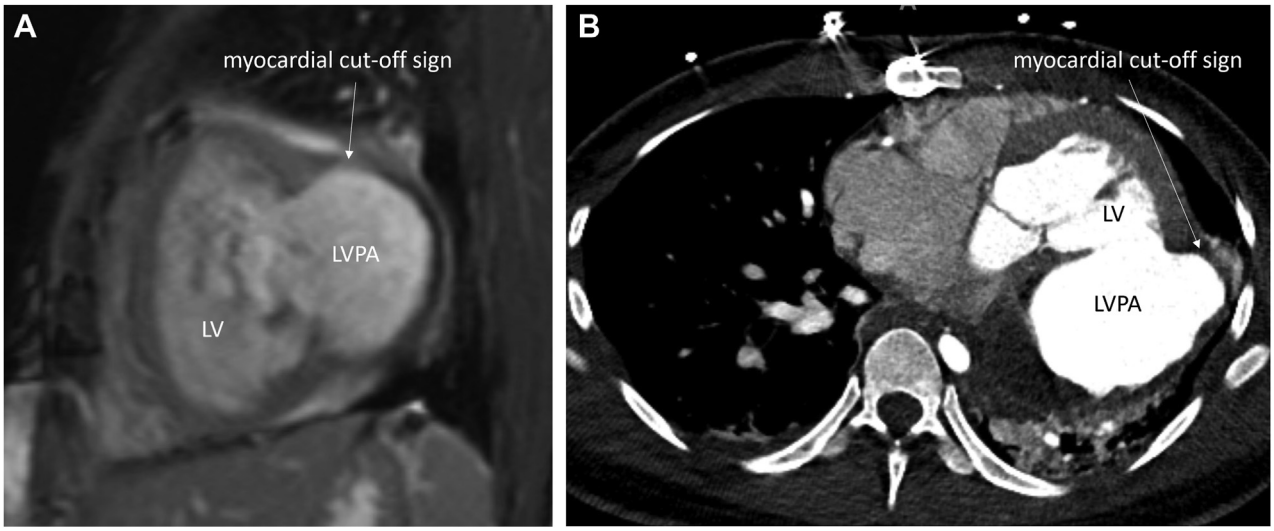

Figure 2 CMR, balanced steady-state free precession sequence in a sagittal LV short-axis view **(A)** is of low quality but demonstrates the relatively small "mouth" and large "body," the myocardial cutoff sign, and the thin, low–signal intensity lateral wall of the left ventricular pseudoaneurysm (LVPA). CCT, multiplanar reconstruction, axial display **(B)**, similarly demonstrates typical LVPA features, including the myocardial cutoff sign. *LV*, Left ventricle.

abrupt "cutoff" of ventricular myocardium near the neck of the pseudoaneurysm (myocardial cutoff sign). Patients with true aneurysms will often have a gradual tapering of the aneurysmal sac thickness. The myocardial cutoff sign is defined as a >50% drop in the aneurysm's wall thickness at 1 cm from the aneurysm neck.[1] As shown in Figure 2, our patient had a positive cutoff sign on CMR. In addition, our patient's CMR showed pericardial enhancement around the aneurysm, which strongly supported a diagnosis of pseudoaneurysm.[2,5] A three-dimensional model was made on the basis of our patient's CCT scan. We felt that the model enhanced our three-dimensional understanding of the pseudoaneurysm's relationship to cardiac structures. We did not use surgical resection, but a three-dimensional model may help with surgical planning and strategy. The model confirmed a very broad-based pseudoaneurysm, which made surgery much less appealing.

In our patient, the LCX was found to be occluded, and the pseudoaneurysm was in the territory of this vessel. Also, CMR showed transmural infarction in the lateral LV wall. Thus, we hypothesized that the pseudoaneurysm developed after a silent myocardial infarction. We suspect that there was subclinical LCX injury at the AVC repair in childhood. It is also possible that the patient experienced a thrombotic LCX occlusion in the weeks to months before the stroke, but this would be very uncommon in a 39-year-old man. The left anterior descending and right coronary arteries were also without stenosis or calcification. Although we suspected that the pseudoaneurysm resulted from LCX territory infarction, we cannot be absolutely sure of a causal association. Given the lack of interval imaging between the original cardiac repair and the pseudoaneurysm diagnosis, we cannot definitively conclude that the LCX occlusion was related to the pseudoaneurysm formation.

The treatment of choice for an acute LV pseudoaneurysm is surgical resection, although resection can be associated with significant risk and may not always be feasible. Resection is preferred, as it lowers the risk for rupture. In a series by Yeo *et al.*,[6] 81% of patients underwent surgical resection, and 19% did not undergo surgery. The surgical mortality rate was 7%, and the death rate was higher among those who had myocardial infarction (13%) compared with histories of cardiac surgery (4%). In this series, 48% of patients had pseudoaneurysms diagnosed incidentally, and thus the ages of the pseudoaneurysms were unknown.[6] In rare cases, percutaneous device closure has been used to treat LV pseudoaneurysms.[7] Our patient likely had an old, chronic LV pseudoaneurysm. The patient had not been seen by a cardiologist in years, and thus the exact timing of the pseudoaneurysm formation was not clear. A chronic LV pseudoaneurysm should theoretically have a lower risk for rupture because organized scar tissue would likely form around the pseudoaneurysm over time. Also, if our patient developed a pseudoaneurysm after the partial AVC repair, then the absence of rupture during the interval of 13 years between repair and acute presentation would support a low risk for rupture in the future. Medical management of a chronic LV pseudoaneurysm may be more acceptable. However, even chronic pseudoaneurysms have some risk for rupture, and series that describe surgical resection of chronic LV pseudoaneurysms exist.[8]

Our patient was managed with anticoagulation and had no recurrent events and stable pseudoaneurysm appearance at 3-year follow-up. Of the 31 patients treated medically in the large series reported by Frances *et al.*,[3] 48% died at median follow-up of 1 week. The surviving patients remained alive at a median of 3 years; of this subset of patients, 12 lived ≥1 year, five lived ≥5 years, and two patients survived ≥10 years. Zhong *et al.*[9] reviewed 17 cases of post–myocardial infarction LV pseudoaneurysms, 10 of which were managed surgically and the remainder medically. All patients who were treated surgically survived, with a mean follow-up duration of 18.5 months. Of the medically treated patients, one died soon after diagnosis, another died suddenly 3 weeks after discharge, and a third died suddenly 2 years after discharge. The overall survival was higher among the surgical group compared with the medically managed group (100% vs 57%, $P = .02$).[9] Medical therapy alone is associated with risk, but long-term

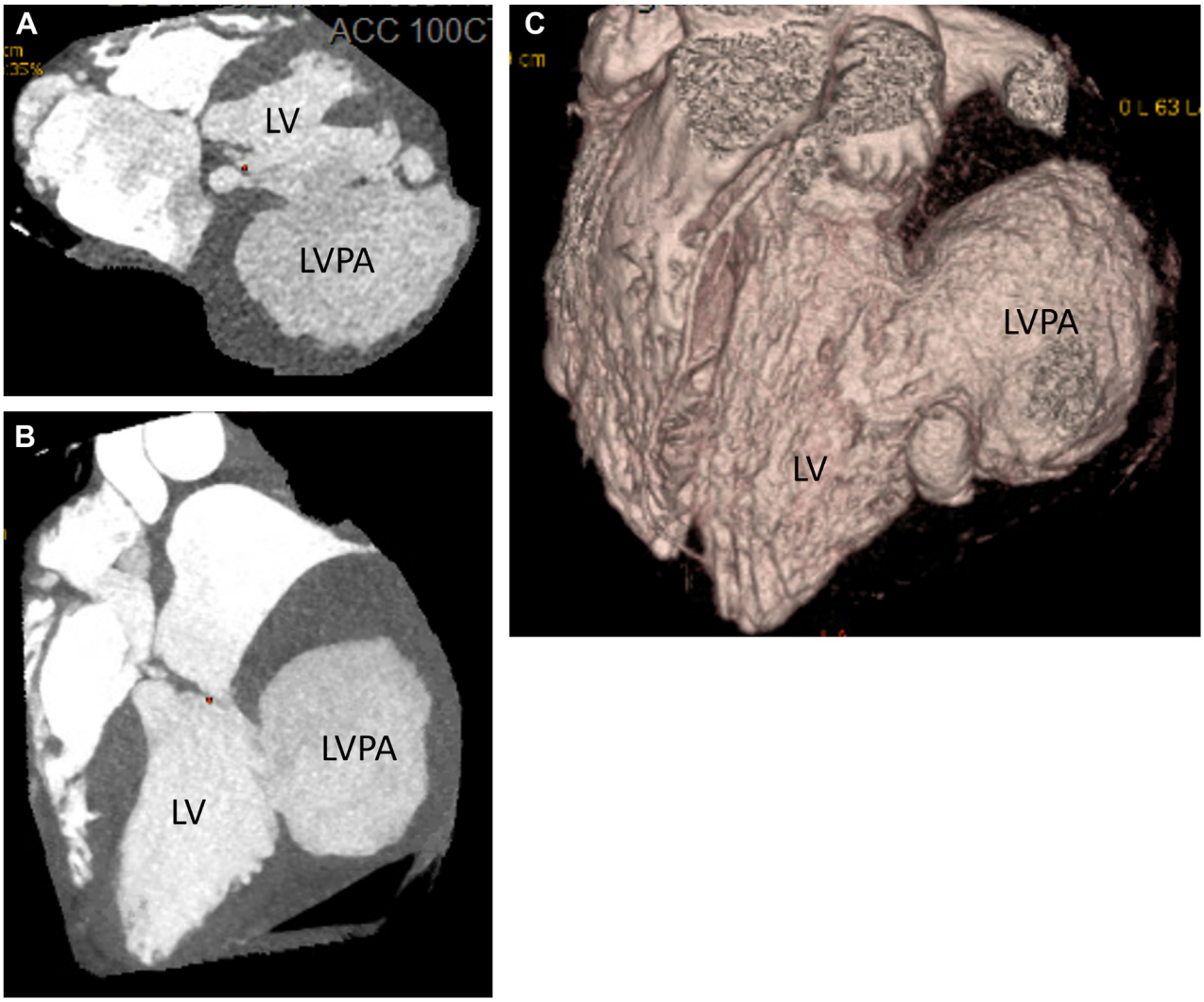

Figure 3 CCT, multiplanar reconstruction views in the short-axis **(A)**, long-axis **(B)**, and volume-rendered whole-heart three-dimensional reconstruction **(C)** view, demonstrates the LVPA arising from the lateral wall of the LV, with the typical narrow neck and broad base.

survival has been described. In our patient, the surgical risk was believed to be prohibitive. To lower the risk for recurrent neurologic event, we anticoagulated our patient with apixaban. Our patient has not had a recurrent neurologic event but does remain at risk.

CONCLUSION

We present the case of a patient who developed an embolic stroke from thrombus that formed in a large pseudoaneurysm in the LV lateral wall. The patient had undergone partial AVC repair many years earlier. We hypothesized that LCX injury at time of the repair led to an infarction and resultant pseudoaneurysm. Our patient was managed conservatively, with no evidence of rupture or recurrent neurologic event at medium-term follow-up.

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

The authors declare that since this was a non-interventional, retrospective, observational study utilizing de-identified data, informed consent was not required from the patient under an IRB exemption status.

FUNDING STATEMENT

The authors declare that this report did not receive any specific grant from funding agencies in the public, commercial, or not-for-profit sectors.

DISCLOSURE STATEMENT

The authors report no conflict of interest.

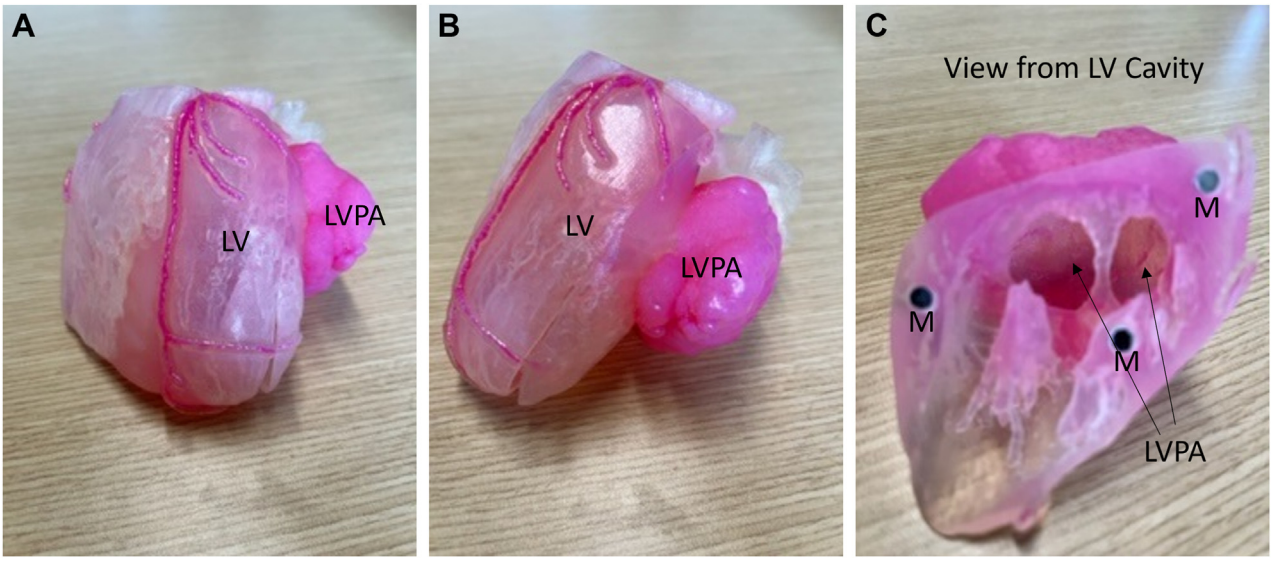

Figure 4 Apical long-axis orientations **(A, B)** of a three-dimensional printed model demonstrating the large LVPA arising from the lateral wall of the LV. When viewed from the inside of the LV cavity **(C)**, the two orifices connecting the LV cavity to the LVPA are well visualized and are separated by a small strand of tissue. The model was constructed with magnets (M) to allow the lateral LV wall to be separated, thus facilitating this view.

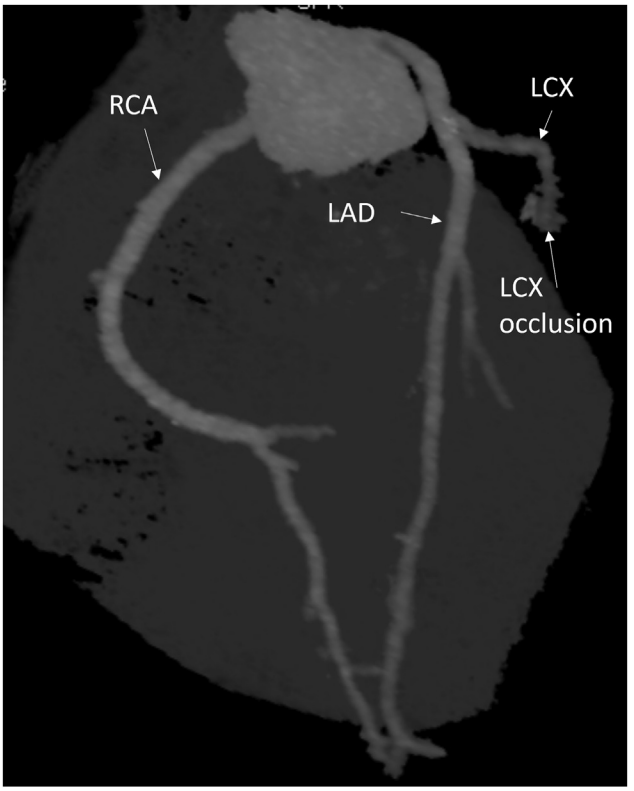

Figure 5 CCT, maximum-intensity projection, left anterior oblique view of the coronary tree, demonstrates the occluded LCX as it approaches the area of the pseudoaneurysm. The left anterior descending coronary artery (LAD) and right coronary artery (RCA) are unobstructed.

SUPPLEMENTARY DATA

Supplementary data to this article can be found online at https://doi.org/10.1016/j.case.2023.12.009.

REFERENCES

1. Ballard DH, Jokerst C, Raptis CA, Pilgram TK, Woodard PK. Myocardial cut-off sign is a sensitive and specific cardiac computed tomography and magnetic resonance imaging sign to distinguish left ventricular pseudoaneurysms from true aneurysms. J Thorac Imaging 2022;37:58-65.
2. Scagliola R, Rosa GM, Seitun S. Cardiac outpouchings: definitions, differential diagnosis, and therapeutic approach. Cardiol Res Pract 2021; 2021:6792643.
3. Frances C, Romero A, Grady D. Left ventricular pseudoaneurysm. J Am Coll Cardiol 1998;32:557-61.
4. Gelehrter S, Wright G, Gless T, Ludomirsky A, Ohye R, Bove E, et al. Left ventricular outflow tract pseudoaneurysms in congenital heart disease. Am J Cardiol 2002;90:806-9.
5. 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.
6. 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.
7. Dudiy Y, Jelnin V, Einhorn BN, Kronzon I, Cohen HA, Ruiz CE. Percutaneous closure of left ventricular pseudoaneurysm. Circ Cardiovasc Interv 2011;4:322-6.
8. Perek B, Jemielity M, Dyszkiewicz W. Clinical profile and outcome of patients with chronic postinfarction left ventricular false aneurysm treated surgically. Heart Surg Forum 2004;7:E132-5.
9. Zhong Z, Song W, Zheng S, Liu S. Surgical and conservative treatment of post-infarction left ventricular pseudoaneurysm. Front Cardiovasc Med 2022;9:801511.