

Congenital Left Ventricular Diverticulum Complicated by Cardioembolic Stroke



Yazan Daaboul, MD, Ethan J. Rowin, MD, Gregory S. Couper, MD, Knarik Arkun, MD, and Ayan R. Patel, MD, *Boston, Massachusetts*

INTRODUCTION

Congenital left ventricle (LV) diverticulum (LVD) is a rare cardiac malformation with a prevalence of approximately 0.1% of all congenital heart diseases.¹ It is characterized by replacement of normal myocardial tissue with either fibrous tissue that subsequently bulges into an outpouching or muscular tissue that results in the formation of a cavity that communicates with the LV through a short neck. The vast majority of LVDs are asymptomatic, but they have been associated in rare instances with cardiovascular and systemic complications. We present a case of a large congenital apical LVD complicated by thrombus formation and thromboembolic stroke in a young patient with otherwise no prior cardiac disease.

CASE PRESENTATION

We present a case of congenital LVD in a 36-year-old man presenting with a thromboembolic stroke. Initially, the patient presented to the emergency department (ED) with left-sided facial droop and slurred speech. At the time of his arrival, his blood pressure was elevated, measuring 220/90 mm Hg, but his physical exam, including cardiopulmonary exam, was otherwise normal. Notably, he had complete resolution of his neurological deficits on initial examination. However, during his stay in the ED, he developed new-onset delay in verbal expression and stuttering, as well as midline gaze and recurrent facial droop.

The patient had a medical history remarkable for essential hypertension, hyperlipidemia, and type 2 diabetes mellitus. His medications included lisinopril, atorvastatin, metformin, and glargine. He had no family history of stroke or other cardiovascular disease. He did not smoke cigarettes and denied any illicit drug use.

Computed tomography of the head demonstrated no acute territorial infarct, space-occupying lesion, or intracranial hemorrhage. Computed tomography angiography of the head and neck demonstrated a normal appearance of the vasculature of the head and neck, without significant stenosis. Electrocardiogram showed sinus tachycardia with evidence of left ventricular hypertrophy and strain pattern. No major ST-segment changes were observed. Admission labs were notable for normal complete blood count, blood glucose

levels, electrolytes, and renal, liver, thyroid function, and urine and serum drug screening tests. Infectious workup, including COVID-19 testing, was negative. Labs were remarkable for an elevated initial troponin I assay of 1.12 ng/mL and peak troponin I = 1.98 ng/mL within 4 hours of admission. In addition, brain natriuretic peptide was elevated at 112 pg/mL.

In the absence of focal cardiac symptoms or ischemic findings on electrocardiogram, the elevated biomarker on admission was thought to likely be due to demand-associated myocardial injury in the setting of a recent stroke and elevated blood pressure. Initially, his blood pressure was managed with intravenous clevidipine infusion. As the patient had developed new-onset neurological symptoms while in the ED and as his blood pressure had gradually improved, he was subsequently administered intravenous alteplase. Following thrombolytic therapy, his neurological symptoms eventually resolved, and he was then transferred to the neurological intensive care unit for diagnostic workup and further monitoring.

The next day, brain magnetic resonance imaging (MRI) demonstrated an area of T2/FLAIR hyperintensity and restricted diffusion within the right middle and inferior frontal gyri, compatible with acute infarction. Given the distribution of the lesions in multiple territories, the MRI findings raised the suspicion for a possible cardioembolic source of his stroke. Transthoracic echocardiogram (TTE) demonstrated a mildly dilated LV with mildly increased interventricular wall thickness. There was moderate to severe global hypokinesis of the LV, with an LV ejection fraction (LVEF) using the biplane method measuring 30%. Notably, ultrasound-enhancing agent administration demonstrated a large outpouching along the LV lateral wall with a bottleneck appearance (Figure 1, Video 1).

To further characterize this cardiac anomaly, cardiac MRI was performed, demonstrating a large outpouching (measuring 5.5 × 2.8 cm) in the mid to apical lateral wall with a narrow neck (measuring 1.0 cm). The rim of the outpouching was noted to have akinetic thin walls (Figure 2, Video 2). On postcontrast images, there was no clear myocardial late gadolinium enhancement (LGE) in the LV (Figure 3).

In addition, a thrombus within the outpouching was suspected, but its presence could not be entirely confirmed due to the presence of significant artifact during the acquisition of the cardiac MRI. Computed tomography angiography of the abdomen (performed to rule out abdominal masses for evaluation of secondary causes of hypertension) demonstrated similar findings, with a thin-walled outpouching of the LV at the apex measuring 4.7 cm anteroposterior × 3.7 cm transverse × 4.79 cm superior-inferior in dimensions. There was a 1.3 cm filling defect concerning for thrombus formation.

The initial differential diagnosis of the cardiac imaging findings included aneurysm versus pseudoaneurysm, with particular concern for the latter based on the imaging characteristics. With evidence for ongoing thromboembolism and the high risk of recurrent thromboembolic events, cardiothoracic surgery was indicated. Preoperative left heart catheterization overall demonstrated patent epicardial

From the CardioVascular Center, Tufts Medical Center, Boston, Massachusetts.

Keywords: Congenital diverticulum, Cardiac aneurysm, Congenital anomaly, Stroke

Conflicts of Interest: None.

Correspondence: Yazan Daaboul, MD, CardioVascular Center, Tufts Medical Center, Boston, MA 02111 (E-mail: ydaaboul@tuftsmedicalcenter.org).

Copyright 2021 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/>).

2468-6441

<https://doi.org/10.1016/j.case.2021.11.010>

VIDEO HIGHLIGHTS

Video 1: Transthoracic echocardiogram (apical four-chamber view) with ultrasound-enhancing agent demonstrating the LV with a reduced LVEF, as well as a large outpouching along the LV anterolateral/apical lateral wall with a bottleneck connection to the LV.

Video 2: Cardiac MRI in four-chamber cine steady-state free precession showing the LV and the large outpouching in the mid to apical lateral wall with a narrow neck. The rim of the outpouching is remarkable for akinetic thin walls.

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

arteries with the exception of a single area of possible “cutoff” seen on angiography of the very distal segment of the first diagonal artery. Right heart catheterization demonstrated marginally elevated left-sided filling pressures with pulmonary capillary wedge pressure measuring 17 mm Hg but otherwise normal right-sided pressures, pulmonary pressures, and cardiac output.

Eventually, he underwent surgical repair of his cardiac anomaly on day 12 of his hospitalization. Intraoperatively, the heart was free within the pericardial space, that is, there was no evidence of rupture of the LV into the free pericardium, ruling out infarct-related pseudoaneurysm. The aneurysmal cavity was easily visualized, measuring approximately $6.5 \times 4.5 \times 2.5$ cm. The epicardial surface of the aneurysmal cavity appeared to have normal heart tissue, and the endocardial surface of the cavity showed no evidence of infarction. Based on these intraoperative findings, it was evident that the patient’s anomaly was neither a pseudoaneurysm nor an aneurysm and was in fact a congenital LVD (Figure 4).

The diverticulum was successfully excised, and a Peri-Guard bovine pericardial patch (Synovis, St. Paul, MN) was sutured. A polytetrafluoroethylene patch overlay was also sutured to cover the exposed surfaces and the underlying patch. Incision within the diverticulum demonstrated multiple thrombi, confirming the cardioembolic nature

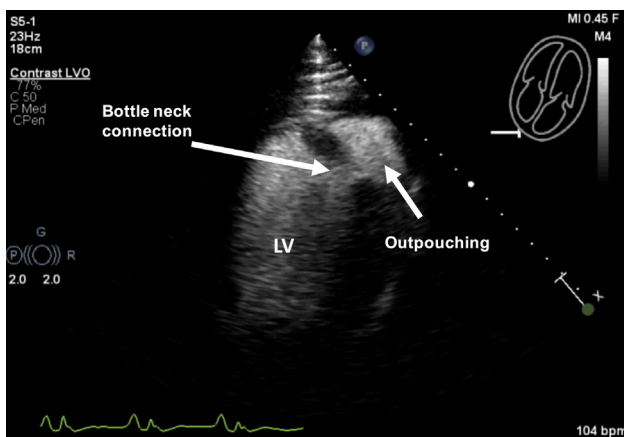


Figure 1 Transthoracic echocardiogram (apical four-chamber view with ultrasound-enhancing agent) demonstrating the LV with a reduced LVEF (see Video 1), as well as a large outpouching along the distal LV anterolateral wall with a bottleneck connection to the LV.

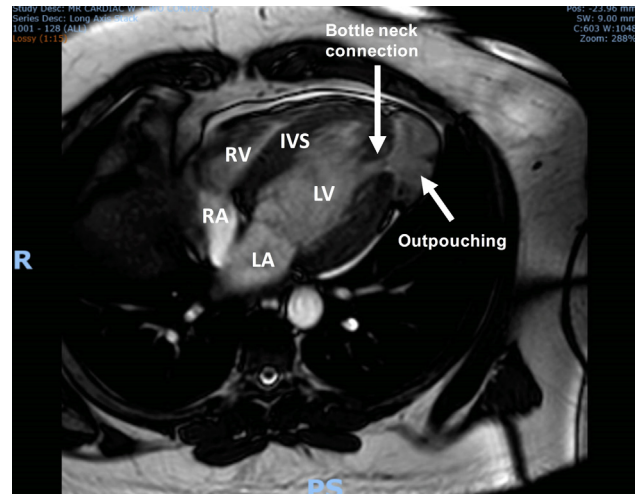


Figure 2 Cardiac MRI in four-chamber still frame of cine steady-state free precession demonstrated a large outpouching (measuring 5.5×2.8 cm) in the mid to apical lateral wall with a narrow neck (measuring 1.0 cm). IVS, Interventricular septum; LA, left atrium; LV, left ventricle; RA, right atrium; RV, right ventricle.

of his stroke. Surgical pathological evaluation confirmed the presence of full endomyocardial tissue within the diverticulum, further establishing the diagnosis of LVD (Figure 5).

The patient’s postsurgical course remained uncomplicated. Repeat TTE at 1 week postop demonstrated no surgical complications, along with gradual improvement in LVEF to 35%-40% using the biplane method (Figure 6). He was eventually discharged on postop day 7. Following his discharge, he visited the cardiology clinic, and he continues to feel well with no further symptoms. Repeat TTE at 6 months postop demonstrated full recovery of LVEF, measuring 60%.

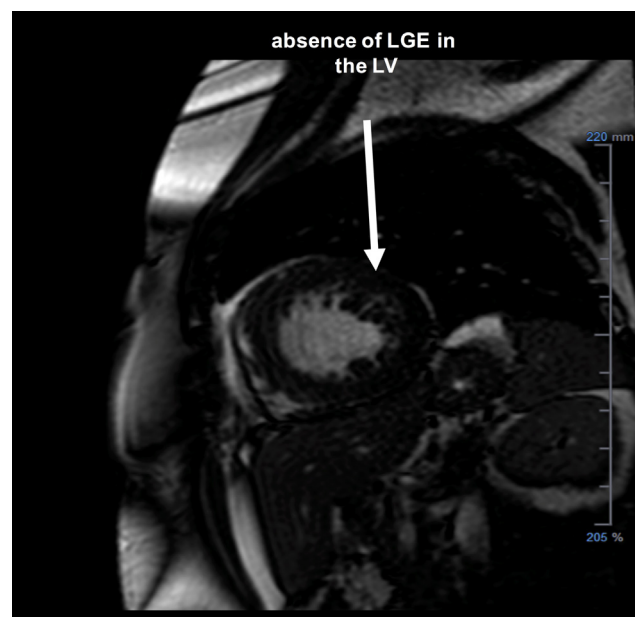


Figure 3 Cardiac MRI with LGE sequencing in short axis demonstrating absence of LGE in the LV.

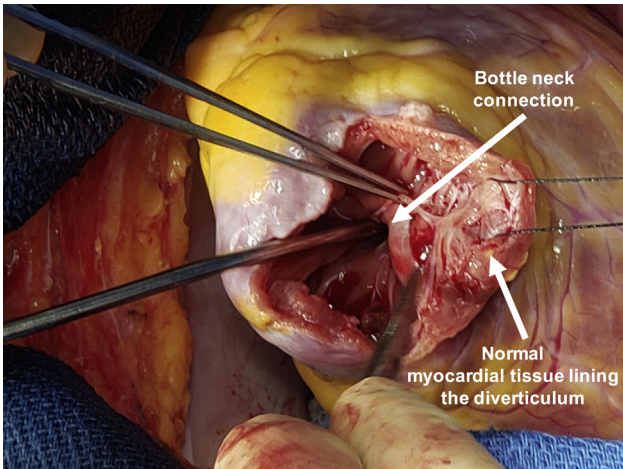


Figure 4 Intraoperative surgical specimen of the diverticulum demonstrating normal myocardial tissue along the wall of the outpouching and a bottleneck connection with the LV cavity.

DISCUSSION

We presented a case of a large congenital apical lateral LVD complicated by thromboembolic stroke. Congenital LVD is a rare cardiac malformation with a prevalence of approximately 0.1% of all congenital heart diseases.¹ It is characterized by replacement of normal myocardial tissue with either fibrous tissue that subsequently bulges into an outpouching or muscular tissue that results in the formation of a cavity that communicates with the LV through a short neck. In both instances, the diverticulum contracts synchronously with the LV cavity, which was discrepant to our case. In contrast, an LV aneurysm is often hypokinetic or contracts dyssynchronously with the LV cavity. While LVDs may be isolated, they may also occur with other congenital cardiovascular anomalies, namely, ventricular septal defects, coronary artery malformations, or Cantrell syndrome.² The

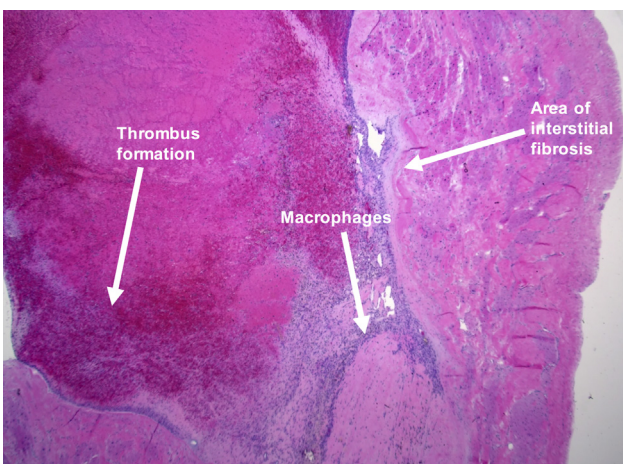


Figure 5 Hematoxylin and eosin–stained section. Full thickness of myocardium showing epicardial and interstitial fibrosis with attached thrombus, consisting of areas with older organization and macrophages at the point of attachment to myocardium and fresh blood clot.

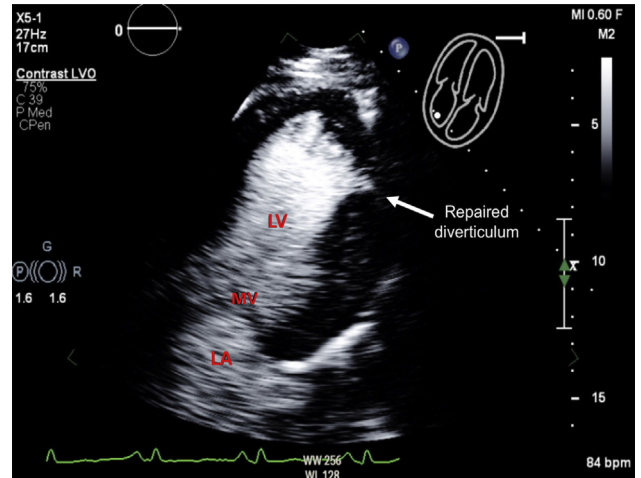


Figure 6 Transthoracic echocardiogram (apical four-chamber view with ultrasound-enhancing agent) demonstrated the LV at the site of the repaired diverticulum postsurgery. LA, Left atrium; MV, mitral valve.

vast majority of LVDs are asymptomatic, but they have been associated in rare instances with diverticular rupture, thrombotic events, and malignant arrhythmias.^{3,4} While there are no randomized data regarding the optimal management, surgical excision is usually reserved for patients who develop complications.^{1,5}

In this case, there was initial concern for pseudoaneurysm based on the appearance of the outpouching on TTE and cardiac MRI, particularly given the absence of synchronous contractility of the outpouching walls, a feature that is inconsistent with a typical diverticulum. Similar to a diverticulum, a pseudoaneurysm may form at the apical LV region and often communicates with the LV through a small neck. In contrast to a diverticulum, a pseudoaneurysm does not contain a true lumen, but this may not be clearly evident on imaging modalities. Therefore, it is common that a definitive diagnosis of pseudoaneurysm versus diverticulum is only made at the time of surgical excision. In this case, considering the patient's young age, relatively low surgical risk, and high risk of recurrent thromboembolic events, it was deemed appropriate that the patient undergo surgical resection of the ventricular abnormality, whether it was a pseudoaneurysm or a diverticulum.

Notably, at the time of presentation, the patient's TTE demonstrated global hypokinesis with severely reduced LVEF of 30%. This, however, was followed with recovery over the subsequent several weeks. It is unclear whether resection of the diverticulum contributed to the improvement of the LVEF. Alternatively, considering the presentation of stroke and hypertensive emergency, it is also possible that the patient sustained stress cardiomyopathy resulting in transient and reversible global hypokinesis.

CONCLUSION

This case highlights cardiac anomalies in the differential diagnosis of stroke and systemic embolism in young patients with otherwise no known cardiac history. While cardiac LVDs are usually asymptomatic, clinical manifestations, including thromboembolic events, may occur, and therefore should be investigated. The distinction between an LVD and LV pseudoaneurysm may be difficult based on imaging alone.

SUPPLEMENTARY DATA

Supplementary data to this article can be found online at <https://doi.org/10.1016/j.case.2021.11.010>.

REFERENCES

1. Yao M, Wang R, Ye W, Ren C. Surgical treatment of congenital left ventricular diverticulum. *J Thorac Dis* 2021;13:291-8.
2. Sakabe K, Fukuda N, Fukuda Y, Wakayama K, Nada T, Morishita S, et al. Isolated congenital left ventricular diverticulum in an elderly patient that was identified because of an incidental finding during a complete medical checkup. *Int J Cardiol* 2008;125:e30-3.
3. Claus I, Adriaenssens B, Van Beeumen K, Timmers L, Bové T. Congenital left ventricular diverticulum: a rare cause of cardiac arrhythmia. *J Card Surg* 2020;35:926-9.
4. Shauq A, Agarwal V, Crawley C. Congenital left ventricular diverticulum. *Heart Lung Circ* 2006;15:272-4.
5. Marijon E, Ou P, Fermont L, Concordet S, Le Bidois J, Sidi D, et al. Diagnosis and outcome in congenital ventricular diverticulum and aneurysm. *J Thorac Cardiovasc Surg* 2006;131:433-7.