JACC: CASE REPORTS VOL. 17, 2023

CASE REPORT

ADVANCED

CLINICAL CASE

Unroofed Coronary Sinus as Potential Cause of Ischemic Stroke Following Left Atrial Appendage Closure

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ABSTRACT

Unroofed coronary sinus is a rare type of atrial septal defect with a strong association with persistent left superior vena cava. We describe a case with unroofed coronary sinus and persistent left superior vena cava as a cause of recurrent embolic strokes following left atrial appendage closure for atrial fibrillation. (**Level of Difficulty: Advanced.**) (J Am Coll Cardiol Case Rep 2023;17:101892) © 2023 The Authors. Published by Elsevier on behalf of the American College of Cardiology Foundation. This is an open access article under the CC BY-NC-ND license (http://creativecommons.org/licenses/by-nc-nd/4.0/).

HISTORY OF PRESENTATION

A 78-year-old woman was referred for a cardiology consultation following 2 interval strokes after placement of a left atrial appendage (LAA) closure device (Boston Scientific). The patient presented to the hospital with worsening headaches. Blood pressure was 175/95 mm Hg and her heart rate was 95 beats/min. Her physical examination was unremarkable with no neurological deficits. Computed tomography of the head demonstrated interval large right cerebellar infarction (Figure 1).

LEARNING OBJECTIVES

- To be able to make a differential diagnosis of the causes of recurrent stroke in patients with unroofed CS complicated by atrial fibrillation.
- To emphasize that comprehensive imaging is crucial in identifying unroofed CS.
- To understand the limitation of LAA closure in patients with atrial fibrillation and unroofed CS.
- To understand the treatment options in patients with unroofed CS.

PAST MEDICAL HISTORY

Medical history was remarkable for recurrent ischemic strokes, lupus on hydroxychloroquine, diabetes mellitus, peripheral neuropathy, chronic kidney disease, chronic systolic heart failure that recovered, and paroxysmal atrial fibrillation (PAF). She had a right middle cerebral artery infarct in 2019. PAF was diagnosed, and a LAA closure was performed in 2020 at an outside hospital. Based on the available data, she continued apixaban for 45 days after undergoing LAA closure, and then was switched to antiplatelet therapy. In 2021, she had a computed tomography of the head that demonstrated interval left cerebellar infarction. At the time of presentation, she was taking aspirin 81 mg daily.

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The authors attest they are in compliance with human studies committees and animal welfare regulations of the authors' institutions and Food and Drug Administration guidelines, including patient consent where appropriate. For more information, visit the Author Center.

Manuscript received January 8, 2023; revised manuscript received May 1, 2023, accepted May 4, 2023.

ISSN 2666-0849 https://doi.org/10.1016/j.jaccas.2023.101892

ABBREVIATIONS AND ACRONYMS

ASD = atrial septal defect

CS = coronary sinus

LA = left atrium

LAA = left atrial appendage

LSVC = left-sided superior vena cava

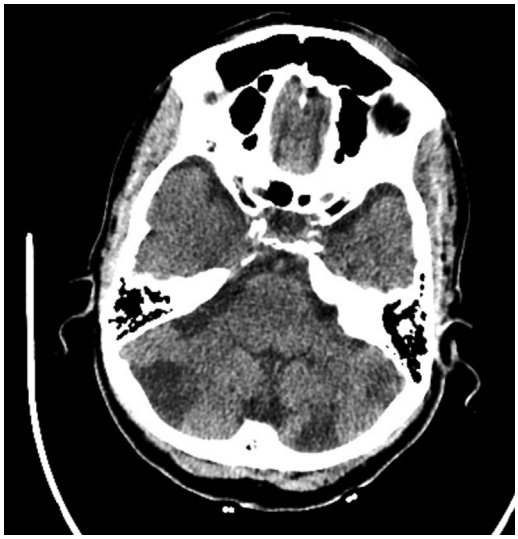
PAF = paroxysmal atrial fibrillation

DIFFERENTIAL DIAGNOSIS. The differential causes of the new right cerebellar stroke (in a new vascular territory) were an LAA peridevice leak, another form of cardioembolism, arteriogenic embolism, paradoxical embolism, or less likely severe stenosis of the basilar artery.

INVESTIGATIONS

Laboratory studies were unremarkable, including a negative anticardiolipin antibody and a low-density lipoprotein of 68 mg/dL. Electrocardiogram demonstrated sinus rhythm (**Figure 2**). Transthoracic echocardiogram demonstrated a dilated coronary sinus (CS) (Video 1), which raised the suspicion of a persistent left-sided superior vena cava (LSVC). Agitated saline injected in the right upper extremity did not demonstrate an intracardiac shunt. However, agitated saline from the left upper extremity demonstrated microbubbles appearing in the CS, left atrium (LA), and left ventricle, confirming a persistent LSVC with an unroofed CS (Video 2).

FIGURE 1 Head Computed Tomography



Computed tomography of the head demonstrates old right cerebellar infarct and new left cerebellar infarct.

With strokes in different vascular territories after LAA closure, a transesophageal echocardiogram was performed, which also demonstrated the persistent LSVC draining to an unroofed CS (Video 3). LAA closure device was well seated, and there was no peridevice leak.

MANAGEMENT (MEDICAL/INTERVENTIONS)

Patient was discharged with apixaban 5 mg twice daily for suspected paradoxical embolism from the left upper extremity through the persistent LSVC and unroofed CS.

DISCUSSION

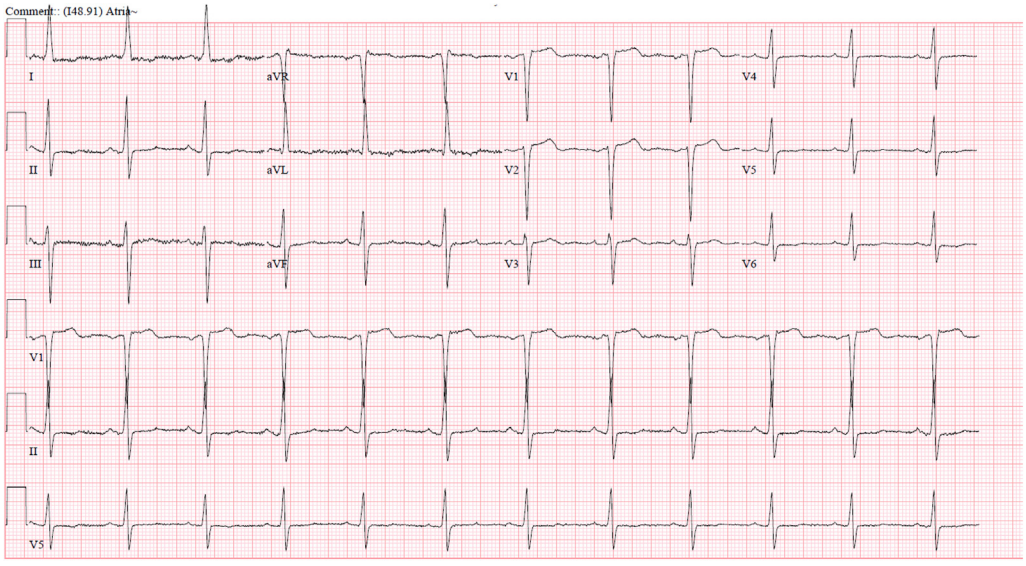
Unroofed CS is a rare congenital anomaly caused by partial or complete absence of the common wall between the CS and the LA, which results in a communication between the CS and the LA.[1] Unroofed CS accounts for <1% of total atrial septal defects (ASDs) and is commonly accompanied by a persistent LSVC,[2] caused by their shared origin from the left anterior cardinal vein during embryonic development.

Cardioembolic strokes associated with ASD may occur caused by either the 2 mechanisms—paradoxical embolism through right-to-left shunting or PAF.[3] ASD is associated with a 6.73× increased risk of an ischemic stroke.[4] In a recent study of 346 patients with ASD, the incidence of stroke prior to ASD closure among patients without atrial arrhythmias was 10%.[5] A prior case of unroofed CS-related cryptogenic stroke has also been reported.[6]

In this case, the first stroke was possibly related to PAF, for which the patient underwent LAA closure. LAA closure has been shown to be noninferior to direct oral anticoagulants for preventing stroke during long-term follow-up in a large clinical trial.[7] In this case, paradoxical embolism from left upper extremity through the persistent LSVC and unroofed CS could be the cause of recurrent stroke, supported by infarcts involving multiple vascular territories.

According to the guidelines, ASD closure should be considered in patients between 18 and 60 years of age for secondary prevention of stroke.[8] However, randomized controlled trial data on ASD closure in patients older than 60 years are limited. In our patient's case, there was a high-risk anatomic feature with a grade 3 shunt bubble through the unroofed CS to LA,

FIGURE 2 **Electrocardiogram**



Electrocardiogram demonstrates normal sinus rhythm with nonspecific T-wave abnormalities.

and she failed antiplatelet therapy. Therefore, she was referred to cardiothoracic surgery for closure.

The role of LAA closure in patients with unroofed CS complicated by PAF is unclear, because the patient may require life-long anticoagulation. Literature is extremely limited because of the rarity of the scenario. LAA closure procedure of this patient was performed at an outside hospital. It is unclear whether a preprocedural transthoracic echocardiogram or transesophageal echocardiogram or intraprocedural transesophageal echocardiogram or intracardiac echocardiography identified the unroofed CS. Which reinforces the need for comprehensive imaging in evaluating patients with strokes, and this is crucial in diagnosing persistent LSVC and unroofed CS, because their identification could have an impact on management.

FOLLOW-UP

Patient will follow-up with cardiology and cardiothoracic surgery.

CONCLUSIONS

Unroofed CS is a rare type of ASD, which could be an unrecognized source of embolic stroke. Dilated CS in the parasternal long-axis view should raise the suspicion of an unroofed CS. An agitated saline intravenous study performed through both arms can increase the diagnostic sensitivity for unroofed CS. Treatment for patients >60 years of age who failed aspirin includes surgical closure vs switching to anticoagulation.

FUNDING SUPPORT AND AUTHOR DISCLOSURES

The authors have reported that they have no relationships relevant to the contents of this paper to disclose.

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KEY WORDS atrial fibrillation, congenital heart defect, stroke

APPENDIX For supplemental videos, please see the online version of this paper.