GIANT LEFT ATRIAL APPENDAGE ANEURYSM WITH SUPRAVENTRICULAR TACHYCARDIA

Giant Congenital Left Atrial Appendage Aneurysm Presenting With Recurrent Supraventricular Tachycardia and Chest Pain

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INTRODUCTION

Left atrial appendage aneurysm (LAAA) is a rare cardiac anomaly that involves the progressive dilatation of the left atrial appendage, predisposing the patient to a number of complications, including tachyarrhythmias and thromboembolic phenomena. This lesion may be either congenital or acquired and is typically diagnosed in the third decade of life, often as an incidental finding during echocardiography. Surgical management is the preferred therapy for this lesion.

Case Presentation

We present the case of a 43-year-old African American woman who presented with chest discomfort. She had a history of recurrent deep venous thrombosis and pulmonary embolism treated with warfarin, supraventricular tachycardia (SVT), and idiopathic pericardial effusion requiring a pericardial window. The patient described an exertional burning discomfort in her chest that radiated to the left arm, symptoms that were not typical of her prior episodes of SVT. The patient denied recent palpitations, syncope, dyspnea, or edema. A physical examination demonstrated normal vital signs, regular heart rate and rhythm, and no signs of heart failure. The remainder of the examination was normal. Results of basic laboratory examination, including metabolic panel, complete blood count, and brain natriuretic peptide, were normal. D-Dimer was mildly elevated. Serial troponins were unremarkable. Twelve-lead electrocardiography (Figure 1) demonstrated sinus bradycardia with poor R-wave progression and a bifid P wave consistent with prolonged left atrial activation and enlargement. No arrhythmias were noted on telemetry.

Chest radiography demonstrated mild cardiomegaly. The patient’s electronic records from a recent admission at a nearby hospital for SVT revealed that coronary computed tomography (CT) had demonstrated a coronary calcium score of zero and normal coronary arteries without evidence of extrinsic compression; however, a large dilated structure associated with the left upper pulmonary vein, which was assumed to be a pulmonary vein varix, was noted. The patient was sent for computed tomographic pulmonary embolism protocol, which did not show pulmonary embolism but was remarkable for a 7.0 × 6.9 × 5.9 cm lobular structure suspected to be arising from or taking the place of the left atrial appendage (Figures 2A and 2B).

Comprehensive two-dimensional and three-dimensional (3D) transthoracic echocardiography (TTE) with contrast and Doppler imaging showed normal ventricular size and function (normal ejection fraction and normal global longitudinal strain 1–19%), moderate left atrial enlargement, and normal valvular structures. In the apical views, a large, nonmobile, lobular, echo-free cavity measuring 9.1 × 6.7 cm was seen adjacent to the left ventricular antralateral and left atrial free walls; only mild impingement of the left ventricle was noted. Focused imaging of the left atrium demonstrated the communication between this structure and the left atrium and revealed a giant LAAA (Figures 3A, 3B, and 4; Videos 1 and 2).

Of note, the thrombus was not apparent within the LAAA on contrast echocardiographic images (the absence of thrombus was subsequently confirmed at the time of surgery; Figure 3B, Video 2), and pulsed-wave Doppler interrogation of the ostium demonstrated brisk flow into and out of the LAAA (Figure 5, Video 3).

Follow-up was arranged with our adult congenital cardiology clinic, and the patient was discharged from the hospital. At the time of follow-up, the patient reported intermittent, nearly daily, episodes of sharp left-sided chest pain. Given the large size of her aneurysm and highly symptomatic status, she was referred for open heart surgery. Before the scheduled surgery, the patient presented emergently to the hospital with palpitations and chest discomfort and was found to be in SVT. Following hospital admission, telemetry demonstrated SVT and paroxysmal atrial fibrillation. She was started on amiodarone, and her β-blocker dose was increased. In further discussion with electrophysiology and cardiothoracic surgery, a combined left atrial appendectomy and maze procedure was planned. Intraoperatively, the diagnosis was confirmed on transesophageal echocardiography, and the patient successfully underwent resection of a large intrapericardial left atrial appendage and uncomplicated maze procedure (Figures 5-7, Videos 3 and 4). She made a complete recovery and at 6-month follow-up had no recurrence of her chest discomfort or atrial arrhythmia.

DISCUSSION

LAAA is a rare abnormality of the left atrial appendage characterized by either local or diffuse outpouching and enlargement of the left atrial appendage. Although the published literature is limited, there is suggestion that giant LAAA can be either congenital or acquired. Far rarer are cases of right atrial appendage aneurysm. Giant LAAA cases tend to present in the third to fourth decade of life, most commonly in female patients. Patients can present with a variety of symptoms, including dyspnea, chest discomfort, palpitations...
secondary to recurrent atrial arrhythmia, angina, or cerebrovascular accident.

In recent years, CT has become increasingly useful in assessing a wide variety of cardiac pathologies as a result of more sophisticated gating. A number of cases in the literature have highlighted the diagnostic utility of CT in giant LAAA, including the use of volume-rendered 3D reconstructions. Despite this, the standard for diagnosis remains echocardiography, and TTE is useful as the initial evaluation tool in suspected LAAA. Our case is illustrative of this, as comprehensive TTE was instrumental in reconciling discordant computed tomographic interpretations, confirming the diagnosis, and planning for surgery. Novel to our study, we demonstrated the important role of newer echocardiographic technologies (i.e., contrast and 3D imaging) in making the diagnosis. Furthermore, with fetal echocardiography, cases have been diagnosed as early as during the intrauterine period. In a large review, Aryal et al. reported that atrial appendage aneurysm size by TTE was highly variable, with the smallest measuring 2.2 × 1.1 cm and the largest 13 × 10 cm. The average size of LAAAs was 7.08 ± 3.03 × 5.75 ± 2.36 cm.

**Figure 1** Twelve-lead electrocardiogram revealing sinus bradycardia with poor R-wave progression and left atrial enlargement.

**Figure 2** Computed tomographic images. (A) Axial computed tomography demonstrates a 7.0 × 6.9 × 5.9 cm giant aneurysmal left atrial appendage. (B) Computed tomographic 3D reconstruction of the LAAA. Ao, Aorta; LA, left atrium; LPV, left pulmonary vein; LV, left ventricle; RV, right ventricle; RVOT, right ventricular outflow tract.
However, they did not correlate their measurements with pathologic specimens. Of note, in our case, transthoracic echocardiographic measurements of the left atrial appendage proved to be highly accurate, as they were subsequently confirmed at the time of surgery, further supporting the diagnostic role of TTE. Additionally, the use of echocardiographic contrast was effective in ruling out the presence of thrombus, a key element in surgical planning.\(^12\)

Surgical resection is the treatment of choice given the potential serious complications associated with this anomaly. Multiple surgical approaches have been used in the treatment of LAAA.\(^13\)

**CONCLUSION**

This case represents a unique presentation of a giant LAAA that presented with recurrent chest pain and frequent SVT. Despite multiple prior images of the patient, the cause of the patient’s symptoms remained elusive until a comprehensive multimodal imaging approach including TTE, CT with volume-rendered 3D...
reconstructions, and transesophageal echocardiography was undertaken at our institution. Giant LAAA has the potential to cause significant morbidity for the patient and should be managed definitively with surgical resection. As in the case of our patient, surgery is usually uncomplicated and can provide complete resolution of symptoms.

**SUPPLEMENTARY DATA**

Supplementary data related to this article can be found at https://doi.org/10.1016/j.case.2019.01.003.

**REFERENCES**

1. Aryal MR, Hakim FA, Ghimire S, Ghimire S, Giri S, Pandit A, et al. Left atrial appendage aneurysm: a systematic review of 82 cases. Echocardiography 2014;31:1312-8.
2. Aryal MR, Hakim FA, Giri S, Ghimire S, Pandit A, Bhandari Y, et al. Right atrial appendage aneurysm: a systematic review. Echocardiography 2014;31:534-9.
3. Krueger SK, Fertic RM, Mooring PK. Left atrial appendage aneurysm: correlation of noninvasive with clinical and surgical findings: report of a case. Circulation 1975;52:732-8.
4. Wagshal AB, Applebaum A, Crystal P, Goldfarb B, Erez A, Tager S, et al. Atrial tachycardia as the presenting sign of a left atrial appendage aneurysm. Pacing Clin Electrophysiol 2000;23:283-5.
5. Frambach PJ, Geskes GG, Cheriex EC, Wellens HJ, Penn OC. Giant intrapericardial aneurysm of the left atrial appendage. Eur Heart J 1990;11:848-53.
6. Pomerantzeff PM, Freyre HM, de Almeida Brandao CM, Pereira Barreto AC, Almeida de Oliveira S. Aneurysm of the left atrial appendage. Ann Thorac Surg 2002;73:1981-3.
7. Ulucam M, Muderrisoglu H, Sezgin A. Giant left atrial appendage aneurysm: the third ventricle! Int J Cardiovasc Imaging 2005;21:225-30.
8. Wagdy K, Samaan A, Romeih S, Simry W, Afifi A, Hassan M. Giant left atrial appendage aneurysm compressing the left anterior descending coronary artery. Echocardiography 2016;33:1790-2.
9. Zeng H, Yu J, Xu Z, Luo Y, Chen H, Zhu H. Giant congenital left atrial appendage aneurysm. J Card Surg 2015;30:646-7.
10. Hoffmann U, Hamed N, Herold C, Globits S. Radiological signs of a left atrial aneurysm. Eur Radiol 2000;10:1332-4.
11. Cho MJ, Park JA, Lee HD, Choo KS, Sung SC. Congenital left atrial appendage aneurysm diagnosed by fetal echocardiography. J Clin Ultrasound 2010;38:94-6.
12. DiBardino DJ, Aggarwal A, Knudson JD. Off-pump snare technique for congenital left atrial appendage aneurysm. Cardiol Young 2014;24:555-8.
13. Vagefi PA, Choudhry M, Hilgenberg AD. Excision of an aneurysm of the left atrial appendage. J Thorac Cardiovasc Surg 2007;133:822-3.