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Author for correspondence:
E. Saraçoğlu, Gaziantep Dr Ersin Arslan Training and Research Hospital, Department of Cardiology, Gaziantep 270001, Turkey. Tel: +90 5325561245; E-mail: erhansaracoglu@hotmail.com

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Multiple congenital ventricular diverticula causing heart failure

Erhan Saraçoğlu, Salih Kılıç and Zülfiye Kuzu

Department of Cardiology, Dr. Ersin Arslan education and research Hospital, Gaziantep, Turkey

Abstract

This paper reports a case of multiple congenital ventricular diverticula, a rare anomaly that is associated with heart failure.

Genital ventricular diverticulum is a rare cardiac disease that most commonly involves the left ventricle. It may be associated with serious complications such as systemic thromboembolism, heart failure, cardiac rupture, ventricular arrhythmias, and even sudden cardiac death.[1–3]

Case report

A 19-year-old woman presented with dyspnoea on exertion (NYHA class II). During physical examination, a late systolic murmur of grade 2/6 was heard in the apical area. A 12-lead electrocardiogram showed a sinus rhythm with a heart rate of 74 bpm. Transthoracic echocardiography revealed mild mitral regurgitation and minimally reduced left ventricular systolic functions. In addition, three deep trabecular sinusoids were revealed in the left ventricular apex that were consistent with multiple diverticula (Supplementary Fig A). The patient underwent coronary angiography and left ventriculography. The coronary arteries were normal. A left ventriculogram obtained from a right anterior oblique projection showed three contractile diverticula originating from the apex of the left ventricle (Supplementary Fig B). The diverticula were muscular structures, and they were seen to be synchronously contracting in systole in the left anterior oblique projection and relaxing in diastole with the rest of the left ventricle. Serum T. Cruzi IgG antibody was negative. This prompted us to make a diagnosis of congenital, multiple left ventricular diverticula. The patient was treated with angiotensin converting enzyme-1, metoprolol and spironolactone for heart failure. She improved on medical treatment and was discharged with periodic follow-up.

Discussion

Congenital left ventricular outpouchings have long been classified based on the following three characteristics as narrow connecting neck, synchronous contraction of protrusion, and presence of all three layers – myocardium, endocardium, and pericardium – in the outpouching.[4] However, a review published in 2014 provided a new classification, where congenital left ventricular outpouchings were classified according to left ventricular elliptical shape preservation, wall thickness, and regional wall motion.[5] According to the new classification, our patient was classified as left ventricular outpouching type 1, because the left ventricle could not retain its elliptical shape, and the wall thickness did not decrease.

Ohlow et al investigated a large series of patients from prenatal age to adulthood and identified 664 patients with left ventricular aneurysm or left ventricular diverticulum since the first description of the condition in 1816. Today, around 800 cases have been published.[6,7] However, multiple ventricular diverticula are very rare.[1,8,9] Congenital left ventricular outpouchings should also be distinguished from other causes of left ventricular aneurysm such as post-cardiac surgery, trauma, Chagas disease, tuberculosis, and idiopathic.[10,11]

Although it is a rare cause of non-ischaemic cardiomyopathy, the finding of apical wall dyskinesia or apical ventricular aneurysm in a patient with symptoms of heart failure and no evidence of myocardial ischemia should prompt the consideration for Chagas disease. The left ventricular aneurysms due to Chagas disease are very difficult to distinguish from congenital left ventricular aneurysms without the knowledge of past history and the serology. Therefore, when apical aneurysm is seen, Chagas disease needs to be ruled out by doing serological tests. In addition, travel to places where the disease is common needs to be questioned.[12]

Although the exact aetiology of the left ventricular diverticula remains unclear, an embryological developmental defect has been proposed. An outpouching can occur due to maldevelopment of myocardial intra-trabecular sinusoids in the first 2–3 weeks of embryonic

life.[13] Most left ventricular diverticula are located at the left ventricular apex.[7] When left ventricular diverticulum is associated with midline thoracoabdominal congenital abnormalities, diaphragmatic and sternal defects, and partial absence of the inferoapical pericardium, the condition is called Cantrell syndrome.[14]

The natural history of left ventricular diverticulum has not been studied systematically. The age of diagnosis may range from the prenatal period to the eighth decade. Interestingly, the size of the protrusion might increase, decrease, or remain unchanged with time. If Cantrell syndrome is not accompanied by any other condition, it may remain silent and go unnoticed for a long time. However, several complications have been reported in association with ventricular diverticulum, including cardiac rupture, embolic events, congestive heart failure, endocarditis, ventricular arrhythmias, and valvular abnormalities. In our case, a patient who had no complaints for up to 2 years presented with increasing effort dyspnoea. Echocardiography revealed mild impairment of left ventricular systolic function. A small number of adult cases of heart failure due to ventricular diverticulum have been reported in the literature.[15,16] These cases may be related to the size of the diverticulum or aneurysm. In our case, although the diverticular dimensions were small, they were multiple, and apical settlement may lead to heart failure.

Currently, there is no consensus on treatment of the left ventricular diverticula due to rarity of the condition. Some authors have recommended immediate surgical resection to prevent complications.[17] However, the decision depends on parameters such as diverticulum size, presumed risk of rupture, occurrence of a thromboembolic incident, or serious ventricular arrhythmia.[18] Chesini et al reported a successful surgical intervention in a patient with multiple left ventricular diverticula who presented with multiple embolic episodes. No recurrent embolism was observed during 1 year follow-up. However, surgical treatment could only be performed on one of four diverticula.[19] Harris et al reported a symptomatic young lady who presented with multiple diverticula and was followed up with heart failure treatment and anticoagulant therapy; the patient's symptoms were seen to improve.[20] We treated our patient with β-blockers, spironolactone, ramipril + hydrochlorothiazide, and warfarin. On follow-up, she remains asymptomatic. Therefore, surgical treatment was not considered.

Conclusions

Congenital left ventricular diverticula are rare diseases. They can remain clinically silent until the end of life, and can cause serious complications. Our patient presented with heart failure that was caused by multiple congenital left ventricular diverticula. We demonstrated the clinical benefit of a medical treatment approach.

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Conflict of interest. The authors have no conflicts of interest.

Ethical standards. The authors assert that all procedures performed in this case comply with the ethical standards of the Helsinki Declaration of 1975, as revised in 2008.

Supplementary materials. To view supplementary material for this article, please visit https://doi.org/10.1017/S104795111800241X

References

1. Alkan MB, Bilgin M, Zihni B, Nalbantgil S. Unusual association of multiple congenital left ventricular diverticulum and cerebrovascular events in an adult. Turk Kardiyol Dern Ars 2015; 43: 269–271.
2. Pitol R, Cardoso CO, Cardoso CR, et al. Congenital ventricular diverticulum associated with ventricular tachycardia. Arq Bras Cardiol 2005; 84: 173–175.
3. Cooke CT, Nolan JR, Kilburn CJ. Sudden death associated with a cardiac diverticulum. Am J Forensic Med Pathol 1991; 12: 340–343.
4. Zhang YL, Shen E, Wu ZH, Xue XP, Zhang WX. Echocardiographic findings on congenital left ventricular diverticulum and aneurysm: two rare cases and literature review. Echocardiography 2010; 27: 344–347.
5. Malakan Rad E, Awad S, Hijazi ZM. Congenital left ventricular outpouchings: a systematic review of 839 cases and introduction of a novel classification after two centuries. Congenit Heart Dis 2014; 9: 498–511.
6. Ohlow MA, Brunelli M, Lauer B. Characteristics and outcome of primary congenital left ventricular aneurysm and diverticulum: analysis of cases from the literature. Prenat Diagn 2014; 34: 893–899.
7. Ozaki N, Sato M, Inoue K, Wakita N. Congenital left ventricular diverticulum in an adult patient. Asian Cardiovasc Thorac Ann 2017; 25: 388–390.
8. Hubacek J, Brydie A, Jackson S. Multiple congenital left ventricular diverticula and aneurysm. J Am Coll Cardiol 2009; 53: 1087.
9. Tandar A, Meredith KG, Anderson JL. Multiple left ventricular diverticuli in an asymptomatic adult: case report and review of the literature. Catheter Cardiovasc Interv 2007; 69: 453–458.
10. Nunes MC, Kreuser LJ, Ribeiro AL, et al. Prevalence and risk factors of embolic cerebrovascular events associated with Chagas heart disease. Glob Heart 2015; 10: 151–157.
11. Wei W, Hui C, Sheng-Yu W, Xin D. Uncommon cause of left ventricular pseudoaneurysm: case report and review of literature. Thorac Cardiovasc Surg 2014; 62: 375–377.
12. Andrade JP, Marin Neto JA, Paola AA, et al. I. Latin American Guidelines for the diagnosis and treatment of Chagas' heart disease: executive summary. Arq Bras Cardiol 2011; 96: 434–442.
13. Papagiannis J, Van Praagh R, Schwint O, et al. Congenital left ventricular aneurysm: clinical, imaging, pathologic, and surgical findings in seven new cases. Am Heart J 2001; 141: 491–499.
14. Makkuni P, Kotler MN, Figueredo VM. Diverticular and aneurysmal structures of the left ventricle in adults. Tex Heart Inst J 2010; 37: 699–705.
15. Handler CE, Walker JM. Congenital diverticulum of the left ventricle presenting as heart failure and diagnosed by magnetic resonance imaging. Int J Cardiol 1989; 22: 115–119.
16. Lachaud M, Tivane A, Menete A, Narayanan K, Sidi D, Ferreira B. Isolated giant congenital diverticulum of the left ventricle in adulthood. Int J Cardiol 2014; 171: e107–e109.
17. Parthenakis FI, Kochiadakis GE, Patrianakos AP, Hamilos MI. Peripheral arterial embolism due to a left ventricular diverticulum in a young adult. Chest 2005; 127: 1452–1454.
18. Marijon E, Ou P, Fermont L, et al. Diagnosis and outcome in congenital ventricular diverticulum and aneurysm. J Thorac Cardiovasc Surg 2006; 131: 433–437.
19. Chesini G, Sangera R, Howard T, et al. Rare case of recurrent cardioembolic stroke attributed to multiple congenital cardiac malformations. J Case 2017; 2: 42–46.
20. Harris GA Jr, Wong AK, Lantz DA. Multiple idiopathic left ventricular aneurysms in a Japanese woman. Mil Med 1999; 164: 448–450.

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