Giant subaortic left ventricular diverticulum with aortic regurgitation and stenosis

Rui Hu², Zhiwei Wang³, Jinling Chen³ and Zhiyong Wu²

² Department of Cardiovascular Surgery, Renmin Hospital of Wuhan University, China
³ Department of Echocardiography, Renmin Hospital of Wuhan University, China

* Corresponding author: Department of Cardiovascular Surgery, Renmin Hospital of Wuhan University, Wuchang District, Wuhan, China, 430064; e-mail: wangzhiwei@whu.edu.cn (Z. Wang).

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Abstract

A subaortic left ventricular diverticulum (SLVD) represents an extremely rare congenital anomaly. It can be asymptomatic but sometimes develops fatal complications. Treatment has been debated due to limited experience. We present the successful treatment of a giant SLVD with aortic regurgitation and stenosis and ascending aorta dilatation. Our goal is to improve understanding of this rare entity.

Keywords: Subaortic left ventricular diverticulum • Aortic regurgitation • Aortic stenosis

INTRODUCTION

A ventricular diverticulum is a rare congenital anomaly. Treatment is a matter of debate because of limited experience and an incomplete understanding of its aetiology [1]. A subaortic left ventricular diverticulum (SLVD) is a special subgroup. It is located just beneath the aortic valve and seems to be associated with more frequent complications [2]. We present the successful treatment of a giant SLVD together with aortic regurgitation and stenosis and ascending aorta dilatation.

CASE REPORT

A 66-year-old man was referred to our hospital with unexpected findings of aortic stenosis and regurgitation. He had no chest distress or pain, nor did he report any other abnormal history. A physical examination indicated a 3/6 biphasic murmur in the left second to third intercostal space with no other positive signs.

The patient was admitted to the inpatient department for further examination. Coronary CT angiography (CTA) revealed a trilobated structure between the aortic root and the left and right atria, communicating to the left ventricular outflow tract through a small orifice. The ascending aorta was dilated with a maximal diameter of 46 mm (Fig. 1A, B). Positive findings from transthoracic echocardiography included aortic stenosis and regurgitation (both moderate to severe). An irregular multicystic structure between the aortic root and the left atrium was also seen, with an to and fro shunt through an orifice beneath the noncoronary cusp shown by colour Doppler (Fig. 1C).

The patient underwent surgery for subaortic ventricular protrusion and aortic valve and ascending aorta pathology. When the aorta was cut open after cross-clamping, we noted a small opening beneath the noncoronary sinus (Fig. 2A). The outpouching was then dissected: It was multilocular with a fibroid septum and thrombus inside. The inner orifice was closed with a continuous suture, and the cyst was thoroughly resected and covered by its own wall. A 21-mm Hancock II bioprosthetic valve was deployed to replace the aortic valve, and an Intergard 26-mm artificial conduit was used to replace the ascending aorta. After defibrillation, intraoperative transoesophageal echocardiography showed a normally functioning valve with no leak. Pathological results suggested fibrous proliferation, thrombus formation and the absence of myocardium, confirming the diagnosis of an SLVD.

The patient recovered uneventfully. Postoperative coronary CTA and echocardiography demonstrated that the SLVD had disappeared, that the aortic valve was functioning normally and that the ascending aorta was well reconstructed (Fig. 2B, C). The patient was discharged after 2 weeks and has been well during the follow-up period.

DISCUSSION

A left ventricular diverticulum is an uncommon congenital anomaly; the reported incidence is 0.42% by ventriculography [1]. The SLVD is a rare subgroup; based on a Chinese population report, it accounts for approximately 17% of the total number of patients with a left ventricular diverticulum [3]. They are mostly fibrous, lack myocardium and seldom coexist with other
congenital anomalies. The aetiology is not well understood; it is possibly due to a local defect in embryologic development.

The clinical presentation of an SLVD is varied. The reported symptoms include exertional angina and exercise intolerance due to coronary artery compression, chest distress and dyspnoea due to congestive heart failure, fever caused by endocarditis and even shock. However, a substantial percentage of patients exhibit no positive symptoms. Thus, SLVD are mostly found during routine examinations or diagnostic procedures for other diseases. A clear diagnosis of SLVD has been difficult due to multiple easily confounding conditions such as subaortic left ventricular aneurysm, which shares a similar morphology but has a different prognosis. Echocardiography, CTA and ventriculography are useful tools for differentiation, whereas a histological analysis remains the gold standard for confirmation.

Management of SLVD is still controversial. Some physicians recommend conservative monitoring with intensive imaging surveillance because most cases are benign during long-term follow-up periods, and unnecessary surgical procedures might pose addition risks and lead to more deaths. However, Li et al. found that SLVD is a major independent risk factor for developing complications, including aortic regurgitation, infective endocarditis, arrhythmia or even diverticulum rupture, which may require emergency treatment, as reported by Deng [2, 3]. Thus, resection should be advocated for obvious symptoms or with other comorbidities, including a rapidly enlarging diverticulum observed by imaging. Overall, we feel that SLVD treatment is individual and flexible. The guiding principle should be carefully tailored based on diverticulum pathology, clinical condition and associated comorbidities. A recent report about SLVD percutaneous closure with a patent ductus arteriosus (PDA) occluder offered an easier and safer way to treat appropriate patients, but the long-term prognosis should be carefully monitored [4].

CONCLUSION

SLVD requires proper, individual treatment, especially when it is associated with other cardiac comorbidities or complications. Further clinical and etiological investigations are warranted to obtain a better prognosis.

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