Congenital Subaortic Left Ventricular Muscular Diverticulum

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INTRODUCTION

Congenital subaortic left ventricular diverticulum (LVD) is a rare congenital malformation consisting of a localized outpouching from the free wall of the left ventricular outflow tract. The majority of cardiac diverticula arises from the apex of the left ventricle (LV); however, nonapical LVD also occurs, but infrequently. The diagnosis of a muscular diverticulum should be distinguished from that of an aneurysm of the LV.

CASE PRESENTATION

A baby boy was delivered by full-term vaginal delivery, with no significant family history. The parents denied any symptoms on a routine prevaccination visit at 14 months of age. Physical examination revealed a heart rate of 100 beats/min, blood pressure of 85/45 mm Hg, oxygen saturation of 97%, and a grade 3/6 systolic-diastolic ("to-and-fro") murmur at the left upper sternal border. Chest radiographic examination showed cardiomegaly and normal pulmonary vascular markings. Electrocardiography showed sinus rhythm with a normal axis, left ventricular enlargement, and inverted T waves in leads V_1, V_2, and V_3. Holter electrocardiography recorded sinus rhythm, with no ST-segment or T-wave changes.

Transthoracic echocardiography (TTE) showed situs solitus and levocardia with atrioventricular and ventriculoarterial concordance. An isolated subaortic LVD was diagnosed, with an outpouching of the left ventricular outflow tract that measured 16 × 25 mm (Figure 1). The LVD was located below the right atrioventricular groove, on the anterolateral aspect of right ventricle, and communicated with the left ventricular outflow tract through a 4-mm narrow channel. There was a wall thickness of 5.1 mm with synchronized contraction noted, and biventricular systolic function was normal (Video 1). There was aliasing bidirectional flow in and out of the outpouching's cavity visualized by color flow mapping (Figure 2, Video 2). No ventricular septal defect or left ventricular outflow tract obstruction was found. There was also no aortic cusp prolapse or aortic regurgitation.

Under mild sedation, electrocardiographically gated multislice computed tomography (MSCT) was performed using a 64-channel, multidetector computed tomographic scanner. MSCT confirmed a giant subaortic LVD (15 × 10 × 20 mm). The LVD was under the aortic valve, adjacent to the noncoronary cusp. The neck of the diverticulum measured 12.5 mm in length and 5 × 3 mm in width (Figure 3). There was no communication between the diverticulum and the right ventricle. The diverticulum did not compress on normal coronary arteries.

In our case, because of the lack of cardiac symptoms and the absence of thrombus, conservative treatment for the LVD was chosen. With a risk for aortic regurgitation from its subaortic location, we decided to schedule follow-up every 6 to 12 months with an endocarditis prevention strategy. We used TTE to serially examine the aortic valve, diverticulum size, and cardiac function. Because of the presence of synchronized LVD contraction, the patient was not placed on anticoagulant therapy. At the 3-year follow-up visit at our hospital, the patient was asymptomatic, and electrocardiography showed normal sinus rhythm with no evidence of ischemia. Neither changing ST-T nor arrhythmias were revealed on Holter electrocardiography. The most recent TTE showed trivial aortic regurgitation, good biventricular function, and stable diverticulum size. Close clinical follow-up is planned with routine TTE plus MSCT, cardiac magnetic resonance imaging, and exercise stress testing as indicated.

VIDEO HIGHLIGHTS

Video 1: Apical four-chamber view showing the LVD with three layers of left ventricular myocardium and synchronized contraction.

Video 2: Apical "five-chamber" view using two-dimensional echocardiography and color Doppler showing an LVD communicating with the LVOT through a narrow channel. An aliasing and bidirectional flow in and out of the outpouching's cavity is demonstrated, with no aortic stenosis or regurgitation. *AV*, Aortic valve; *D*, diverticulum; *LV*, left ventricle; *LVOT*, left ventricular outflow tract; *RV*, right ventricle.

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DISCUSSION

Congenital LVD is an uncommon cardiac malformation that was first reported in 1816 (in Germany) and then by O'Bryan in 1837 (in England).[1-3] LVD prevalence has been reported to be 0.42% among adult patients undergoing ventriculography after being diagnosed. The majority of cardiac diverticula arise from the apex of the LV;

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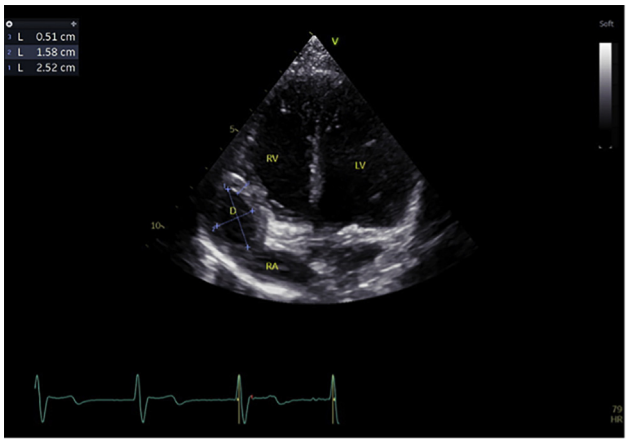

Figure 1 Apical four-chamber view showing an LVD, 16 × 25 mm, with 5.1-mm wall thickness, anterior and lateral to the right ventricle. *D*, Diverticulum; *LV*, left ventricle; *RA*, right atrium; *RV*, right ventricle.

LVD from another location or right ventricular origin is also seen, but rarely.[4]

A series of four subaortic LVD cases was reported in China in 2006,[5] but not all involved the three cardiac layers of myocardium required for the diagnosis of a muscular diverticulum. The differential diagnosis of a pouch overlying the free wall of the LV includes a ventricular diverticulum, aneurysm, or a pseudoaneurysm. The wall of the diverticulum is formed by endocardium, myocardium, and pericardium and contracts synchronously. On the other hand, a myocardial aneurysm is a fibrous saccular lesion that contracts paradoxically.[6] The term *fibrous diverticulum* has been used interchangeably with *aneurysm*, but the diagnosis of a true diverticulum should be synonymous with that of a "muscular diverticulum." A pseudoaneurysm, or false aneurysm, is a hematoma that results from an arterial tear, usually from trauma or surgery. The presentations of a diverticulum, aneurysm, and pseudoaneurysm are distinct, with the diagnosis based on morphologic features. The correct diagnosis is important for directing the search for associated defects, treatment decisions, and prognosis.

A ventricular diverticulum is a congenital anomaly that is often associated with midline thoracic abdominal defects or other congenital cardiac malformations, including ventricular septal defect with or without pulmonary stenosis, atrial septal defect, dextrocardia, endocardial cushion defect, tricuspid atresia, and anomalous pulmonary or systemic venous return (about 70% of cases).[7,8] Patients are usually asymptomatic and discovered incidentally. However, previous subaortic diverticular cases have been reported with aortic regurgitation as a complication. Apart from valvular regurgitation, the other major complications are thrombosis, embolism, rupture, congestive heart failure, and ventricular arrhythmias.[9]

There are many diagnostic techniques to detect LVD, including echocardiography, MSCT, and magnetic resonance imaging. TTE is a useful and noninvasive diagnostic tool to detect congenital cardiac diverticula. It allows an accurate assessment of morphology, location, and possible thrombosis in the diverticulum and demonstrates other congenital cardiac abnormalities. When TTE is technically limited, transesophageal echocardiography may help define the components of the outpouching more clearly and can aid in the diagnosis of an inferior ventricular diverticulum.[10] Magnetic resonance imaging and MSCT

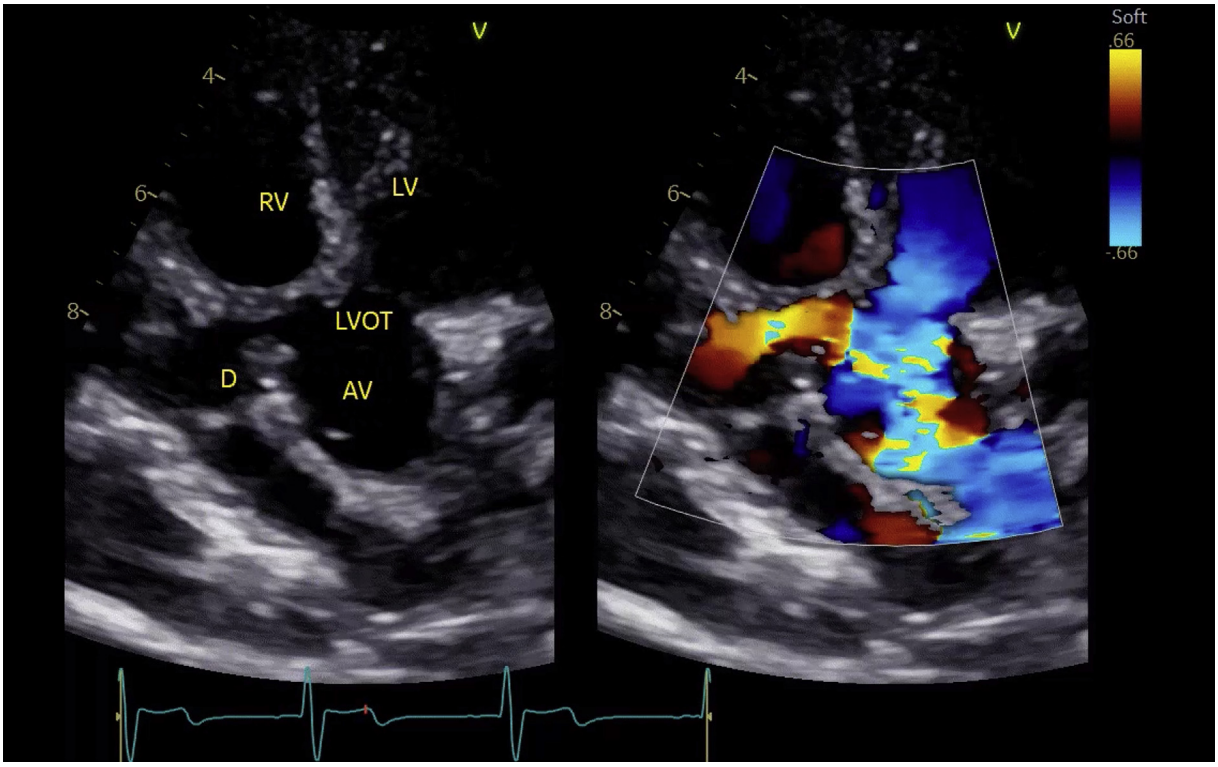

Figure 2 Apical "five-chamber" view using two-dimensional echocardiography and color Doppler showing an LVD communicating with the left ventricular outflow tract (LVOT) through a narrow channel. An aliasing flow at the outpouching's cavity is demonstrated, with no aortic stenosis or regurgitation. *AV*, Aortic valve; *D*, diverticulum; *LV*, left ventricle; *RV*, right ventricle.

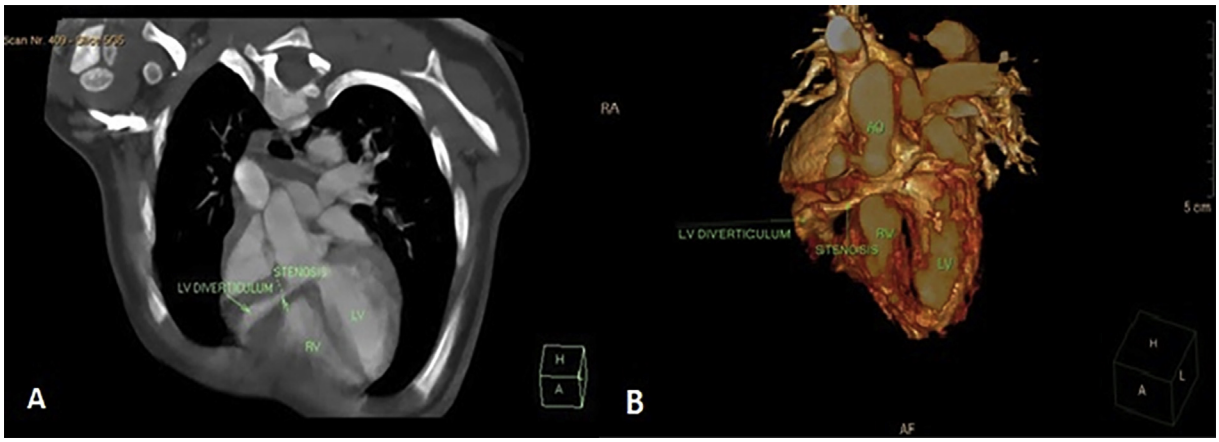

Figure 3 **(A)** Computed tomographic images of the LVD with a stenotic neck. **(B)** Three-dimensional reconstruction of MSCT showing that the LVD was under the aortic valve, adjacent to the noncoronary cusp. *Ao*, Ascending aorta; *LV*, left ventricle; *RV*, right ventricle.

may also allow accurate detection, but they are not as readily available as echocardiography. Left ventricular angiography remains the gold standard for the diagnosis of left ventricular aneurysm and diverticulum. It has an essential role in excluding coronary artery disease, the possible presence of contractility anomalies, the size, and the possible communication between the left and right ventricles.[11] Nevertheless, it is an invasive tool that makes it less acceptable than TTE.[10]

The treatment of LVD remains controversial, and little is known on the management of subaortic LVD because of its rarity. In 2016, an unfortunate case of sudden death associated with subaortic LVD was reported.[12] The patient developed chest pain on the scheduled day of surgery and died of cardiogenic shock. In that case, the diverticulum was shown on MSCT to compress the proximal right coronary artery. Many authors recommend surgery for LVD, even if patients are asymptomatic, to prevent serious and potentially lethal complications. In one large series, spontaneous rupture of the LVD occurred in 15 of 411 patients (an incidence of 3.7%). Other experts have suggested a nonsurgical strategy with careful follow-up. In a small series of 16 patients, uneventful outcomes were demonstrated in 94% of patients over 127 months (mean, 61 months), with an event rate of approximately 1.2% per year. No cardiac death was seen during follow-up.[13] A separate case report showed that LVD did not increase in size over 13 years, suggesting that the detection may be benign.[14] Therefore, in selected cases, LVD repair may be safely postponed, after complete workup, until there is an indication for surgery of associated congenital cardiac anomalies or other complications.[11]

CONCLUSION

Subaortic muscular LVD is a rare congenital cardiac malformation. Careful assessment for symptoms of ischemia and arrhythmias is essential. The diagnosis of a diverticulum should be distinguished from that of aneurysm or pseudoaneurysm. Echocardiography is a useful tool for diagnosis and follow-up, but additional tests may be indicated for long-term management.

SUPPLEMENTARY DATA

Supplementary data to this article can be found online at https://doi.org/10.1016/j.case.2021.06.001.

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