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Case Report

# An adult case of giant congenital left atrial wall aneurysm

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#### A R T I C L E I N F O

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## ABSTRACT

Congenital left atrial wall aneurysm is a rare disorder that occurs in a wide range of age groups from infancy to adulthood. Here, we present a case of a congenital left atrial wall aneurysm that was detected in a 19-year-old man who was surgically treated. Although the patient was asymptomatic without any pre-existing conditions, chest radiography performed as part of a routine health examination detected abnormalities in the heart. Contrast-enhanced computed tomography revealed a giant aneurysm measuring 72 mm  $\times$  56 mm that extended from the posteroinferior wall of the left atrium to the posterior surface of the left ventricle. Transthoracic echocardiography revealed mild mitral regurgitation. The patient was diagnosed as having a congenital left atrial wall aneurysm associated with mild mitral regurgitation. The aneurysm was resected through median sternotomy under cardiopulmonary bypass with cardioplegic arrest. During surgery, no structural abnormalities were noted in the mitral valve. After surgery, the patient was discharged without complications. Neither recurrence of the aneurysm nor exacerbation of mitral regurgitation was observed at 1 year postoperatively.

**Learning objective:** A congenital left atrial wall aneurysm is a rare disorder. Rupture of the aneurysm is rare. However, when they are left untreated, there are concerns regarding arrhythmias, heart failure, and systemic embolism. Thus, surgical treatment is recommended. Aneurysms are resected under cardiopulmonary bypass. In cases of aneurysms complicated by moderate or severer mitral regurgitation, mitral valve repair is necessary. The prognosis following surgical treatment is favorable.

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#### Introduction

A congenital left atrial wall aneurysm is a rare disorder of unknown cause, with only about 20 cases currently reported [1,2]. They often arise from the posteroinferior wall of the left atrium and extend toward the apex of the left ventricle. Although they exhibit symptoms of heart failure, such as palpitations and respiratory distress, few cases are detected while being asymptomatic. Since aneurysms arise near the annulus of the mitral valve, attention should be paid to concomitant mitral regurgitation (MR). Rupture of the aneurysm is rare. However, when they are left untreated, there are concerns regarding the incidence of arrhythmias, heart failure, and systemic embolism. Thus, surgical treatment is recommended. Surgical resection of aneurysms is the standard treatment, and simultaneous mitral valve repair is necessary in some cases.

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Here, we present a case of a giant left atrial wall aneurysm incidentally detected in adulthood, in which surgical resection achieved favorable outcomes.

#### **Case report**

A previously healthy 19-year-old man visited our hospital for further examination because chest radiography, part of a routine health checkup, revealed abnormalities in the heart. The patient presented with no subjective symptoms and had no family history of heart disease.

At the initial visit, the patient's blood pressure was 135/75 mmHg; heart rate, 83 beats/min; and respiratory rate, 12 breaths/min. As for the body constitution, the patient's height and weight were 174 cm and 75 kg, respectively. On physical examination, a holosystolic murmur (Levine II/VI) was heard at the cardiac apex. No other abnormal findings were noted. The hematologic tests showed a brain natriuretic peptide level of 16 pg/mL, which was within the normal range. There were no other remarkable findings. Plain chest radiography showed a cardiothoracic ratio of 51 % and protrusion of the left third arch. Electrocardiography showed a sinus rhythm without ST changes. Transthoracic

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echocardiography showed left ventricular dilatation (end-diastolic dimension, 62 mm; end-systolic dimension, 46 mm) and left ventricular ejection fraction of 49 %. There was an echo-free space that extended from the posterior wall of the left atrium near the posterior annulus of the mitral valve and was as large as the left ventricular cavity. There was no prolapse of the mitral leaflets. MR was mild with a central regurgitation (Fig. 1). The patient had no other concomitant congenital heart disease. Contrast-enhanced computed tomography (CT) revealed a left atrial wall an eurysm measuring 72 mm  $\times$  56 mm that protruded from the posteroinferior wall of the left atrium toward the apex. The left circumflex artery ran through the atrioventricular groove, while the coronary sinus (CS) ran on the surface of the left atrial wall aneurysm (Fig. 2). Based on these findings, the patient was diagnosed as having a giant left atrial wall aneurysm. As the patient presented with left ventricular dilatation, mildly reduced left ventricular ejection fraction, and mild MR. the patient was determined to be indicated for surgery to prevent the exacerbation of heart failure. Prior to surgery, the patient provided informed consent for the publication of this report.

Surgery was performed through median sternotomy (Fig. 3). Under cardiopulmonary bypass with cardioplegic arrest, the aneurysmal wall was resected along with the left atrial appendage. The redundant free wall of the left atrium was also resected, and the incision was resutured to reduce the left atrial volume. The CS was visualized as running in the left atrial wall. Thus, the CS was detached with a 2-mm margin from the surrounding atrial wall and preserved. There was a rim of approximately 20 mm between the orifice of the aneurysmal sac and the posterior annulus of the mitral valve, and the mitral annulus was not deformed. There were no abnormalities in the leaflets or subvalvular apparatus of the mitral valve, and no thrombus was found in the aneurysm. The cardiopulmonary bypass time was 110 min, and the cardioplegic arrest time was 72 min. The resected aneurysmal wall indicated that the aneurysm was a true aneurysm with the normal 3layered structure. Although the histopathological examination showed that the wall was mostly composed of fibrous tissue, there were also hypertrophied cardiomyocytes in some parts. No particular complications were observed after surgery, and the patient was discharged on postoperative day 16. At 1 year after surgery, no cardiac events, such as heart failure and arrhythmia, were observed. Contrast-enhanced CT showed no recurrence of the left atrial wall aneurysm (Fig. 3). Echocardiography showed the left ventricle with an end-diastolic dimension of 59 mm and an end-systolic dimension of 44 mm, which indicated that left ventricular dilatation was improving. The left ventricular ejection fraction improved to 51 %. MR was trivial to mild, showing improvement (Fig. 3).





#### Discussion

Congenital left atrial wall aneurysm is a rare disorder with only about 20 cases reported [1,2]; however, when combined with congenital left atrial appendage aneurysm, about 100 cases of congenital left atrial aneurysm have been reported [3]. Left atrial aneurysms can be classified into congenital and acquired aneurysms according to their etiology. Congenital aneurysm is rare, as described above [1–3]. Acquired aneurysms are often reported as being secondary changes due to rheumatic heart disease, tuberculosis, and syphilitic myocarditis. As our patient had no particular pre-existing conditions that could have caused the aneurysm, it was assumed to be congenital.

Congenital left atrial aneurysms are classified into the following four categories based on their etiology and sites of origin: (1) intrapericardial aneurysms of the left atrial wall, (2) intrapericardial aneurysms of the left atrial appendage, (3) herniation of the left atrial appendage through a congenital pericardial defect, and (4) multi-saccular aneurysms of the left atrial wall [1,4]. In our patient, the aneurysm was a solitary aneurysm arising from the posteroinferior wall of the left atrium, and no pericardial defect was observed. Thus, the aneurysm was classified as an intrapericardial aneurysm of the left atrial wall.

Aneurysms of the left atrial appendage and the left atrial wall present with varying clinical features [1–3]. The incidence of left atrial appendage aneurysms is higher [3], and intra-aneurysmal thrombi are also more frequently detected in patients with left atrial appendage aneurysms. MR is more common in those with left atrial wall aneurysms, whereas supraventricular arrhythmia is observed in patients with both types of aneurysms. Common clinical manifestations are systemic embolisms associated with intra-aneurysmal thrombi and symptoms of heart failure for aneurysms of the left atrial appendage and wall, respectively. Heart failure can be caused by a variety of conditions, such as MR, left ventricular diastolic dysfunction, and anomalous pulmonary venous drainage owing to compression caused by an aneurysm [1,2]. In our case, although arrhythmia was not observed, the patient presented with dilatation of the left atrium and ventricle, mildly reduced left ventricular

ejection fraction, and mild MR, which were typical clinical features of left atrial wall aneurysms.

Surgical treatment is effective, and the prognosis is considered favorable [1–3]. Postoperative recurrence has been reported only in one case [5], and there have been no reported cases of rupture. Although there are no clearly defined indications for surgery, it should be actively considered for patients with comorbidities, such as heart failure, cerebral embolism, and atrial fibrillation. Even in asymptomatic patients, surgery is considered desirable to prevent these comorbidities that worsen the prognosis [1]. Although our patient was asymptomatic, the patient was determined to be indicated for surgery to prevent the exacerbation of heart failure for the following reasons: the extremely large diameter of the aneurysm, a mildly reduced left ventricular ejection fraction of 49 %, and concomitant mild MR.

The standard surgical procedure for left atrial wall aneurysms appears to be the resection of aneurysms through median sternotomy under cardiopulmonary bypass with cardioplegic arrest. As most left atrial wall aneurysms arise from the posteroinferior wall of the left atrium, they need to be manipulated at sites deep in the pericardial sac. Thus, the use of cardiopulmonary bypass is optimal for maintaining hemodynamics and obtaining secure suturing. Moreover, because left atrial wall aneurysms are often complicated by MR, it is necessary to confirm the presence or absence of valve deformation and abnormalities in the valve leaflets or subvalvular apparatus. Mitral valve repair should be actively performed in cases with structural abnormalities of the mitral valve or moderate or severe regurgitation [1,6,7]. In our case, because our patient had a left atrial wall aneurysm associated with mild MR, we performed median sternotomy under cardiopulmonary bypass with cardioplegic arrest. After confirming the absence of abnormal findings in the mitral valve, we resected the aneurysm and the left atrial appendage and resutured the incision to reduce the left atrial volume. At 1 year after surgery, no recurrence of the aneurysm was observed, and MR was improving.

In conclusion, we report the successful surgical resection of a rare congenital left atrial wall aneurysm. One year after surgery, there were no cardiac events such as arrhythmia or embolism, and cardiac

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function and MR showed a trend toward improvement. Surgical aneurysm resection should be considered in patients with cardiac dysfunction or MR.

#### **Declaration of competing interest**

The authors declare that there is no conflict of interest.

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### Patient consent statement

Informed consent was obtained from the patient for publication of the case.

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