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CASE REPORT

Aortic valve replacement and tricuspid valve annuloplasty via a left thoracotomy in an adult with left pulmonary agenesis

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Abstract
We report a case of a 66-year-old man who was diagnosed with severe aortic regurgitation, moderate tricuspid regurgitation and chronic atrial fibrillation. Preoperative computed tomography showed left lung agenesis. We performed aortic valve replacement, tricuspid valve annuloplasty and right pulmonary vein isolation via a left thoracotomy. This approach provided an adequate field of view.

Keywords Lung agenesis · Left thoracotomy

Introduction

Lung agenesis is a very rare congenital condition. It is usually diagnosed soon after birth because of other anomalies that involve the cardiovascular and skeletal systems. In some cases, the pathological changes are compatible with long-term survival, and the diagnosis of lung agenesis may be delayed until adulthood [1]. Cases with lung agenesis that undergo cardiovascular surgery are very rare. In these cases, the surgical approach is a problem because of a severe shift of the mediastinum. We report a case of successful surgery via a left thoracotomy for aortic regurgitation and tricuspid regurgitation.

Case

A 66-year-old Japanese man was referred to our hospital because of exertional dyspnea (New York Heart Association NYHA class II). His past medical history included hypertension and chronic atrial fibrillation. Agenesis of lung had not been diagnosed. Physical findings at the time of admission included an irregular heart rate of 109 beats/min, a blood pressure of 109/53 mmHg bilaterally and a class II diastolic heart murmur at the apex. Hematological examination revealed the absence of anemia, a normal platelet count, and normal liver and kidney function. The cardio-thoracic ratio could not be determined on the chest X-ray, because of left lung agenesis and a shift in the mediastinum. An electrocardiogram showed atrial fibrillation. Chest computed tomography (CT) revealed the absence of the left lung parenchyma, the left bronchus and the left pulmonary vascular structures. Therefore, the mediastinum was severely shifted to the left, and there was a compensatory hyperplasia of the right lung (Fig. 1). Transthoracic echocardiography revealed severe aortic valve regurgitation due to right coronary cusp prolapse and moderate tricuspid valve regurgitation with annular dilatation. The left ventricular end-diastolic diameter was 57 mm, the end-systolic diameter was 36 mm and the ejection fraction was 66%. Coronary angiography showed normal coronary arteries. Aortic valve replacement, tricuspid valve annuloplasty, right pulmonary vein isolation and left atrial appendectomy were planned.

With the patient in the half-right-side lying position, a left anterior thoracotomy (the incision length was about 17 cm) was performed through the fourth intercostal space (Fig. 2). At the same time, the right femoral artery was exposed. Cardiopulmonary bypass (CPB) was established with arterial inflow to the right femoral artery and venous drainage from the right femoral vein and superior vena cava. A left ventricular vent tube was inserted from the right upper pulmonary vein. Right pulmonary vein

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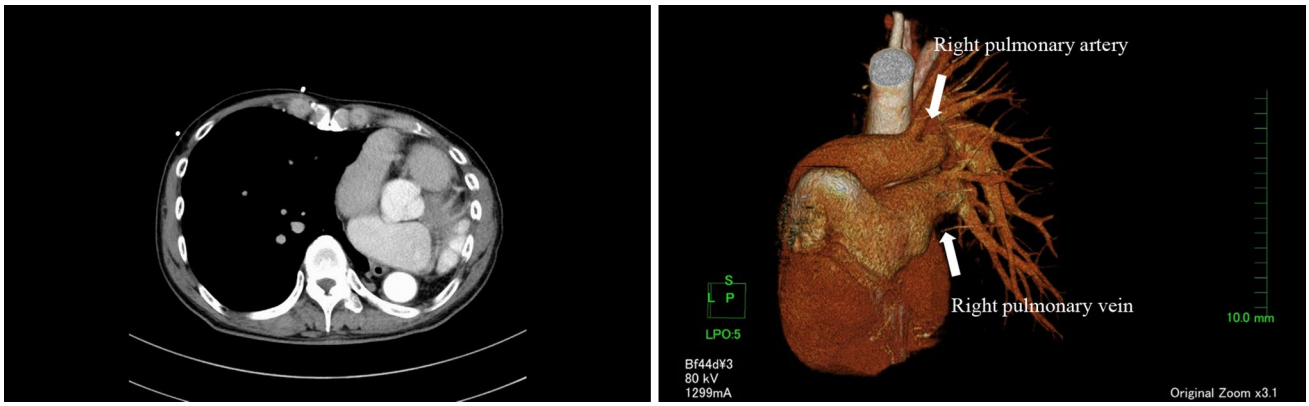

Fig. 1 Enhanced chest CT revealed the absence of the left lung parenchyma, the left bronchus and the left pulmonary vascular structures, and there was a severe shift of the mediastinum

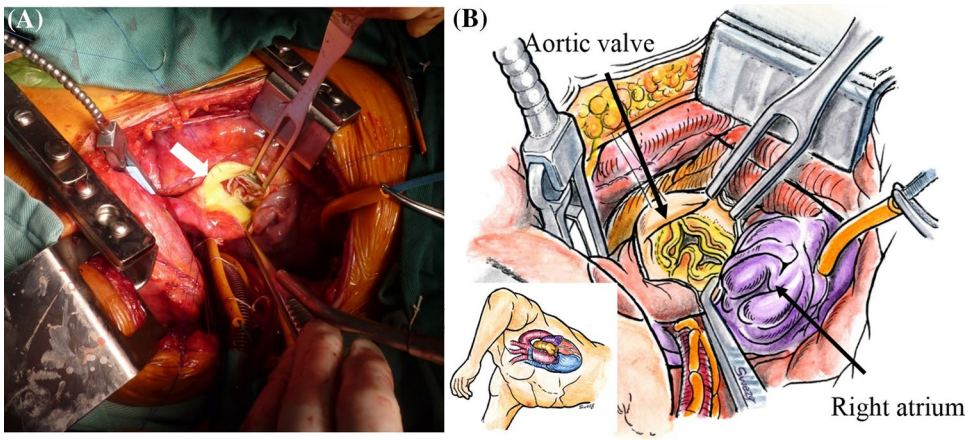

Fig. 2 **a** Exposure of the heart and great vessels via a left thoracotomy. The white arrow is the aortic valve. **b** A schematic figure of the operative findings

isolation was performed with the heart beating normally using the AtriCure bipolar radiofrequency system (AtriCure, Inc, Cincinnati, OH, USA). The ascending aorta was clamped and selective anterograde cold cardioplegia was administered. After cardiac arrest, the left atrial appendix was resected and closed. Then, the right atrium was opened. The annulus of the tricuspid valve was dilated. Therefore, tricuspid valve annuloplasty was performed with the Contour 3D Annuloplasty Ring (Medtronic, Minneapolis, MN, USA). An aortotomy was performed and the incision was extended to the middle of the noncoronary sinus of Valsalva. Prolapse of the right coronary cusp was seen. The aortic valve was excised. A Magna Ease 23 mm valve (Edwards Lifesciences, Irvine, CA, USA) was implanted at the intra-annular position. The aortotomy was closed and the patient was weaned from CPB. The operation time, CPB time and aortic clamp time were 429, 184 and 132 min, respectively. The patient suffered no perioperative complications and was discharged on postoperative day 14. The postoperative echocardiogram confirmed the absence of aortic regurgitation and tricuspid regurgitation.

Discussion

Pulmonary agenesis is a very rare congenital anomaly that is associated with cardiovascular and skeletal malformations. It is defined as the absence of lung vessels, bronchus and parenchyma. The exact incidence of this anomaly remains unknown, but it was found in approximately one in 10,000 to 20,000 autopsies [2]. It is usually diagnosed soon after birth because of other anomalies that involve the cardiovascular and skeletal systems. The presence of other anomalies has a significant influence on prognosis. The mortality of right pulmonary agenesis is usually worse than that of left agenesis, because of coexisting cardiac abnormalities, and distortion of the great vessels and trachea due to a shift in the mediastinum [3].

The clinical features vary individually, and some cases may be asymptomatic until adulthood [4]. Our case also was asymptomatic until adulthood, and the diagnosis was not made until preoperative CT was performed.

Pulmonary agenesis can be diagnosed by chest X-ray or CT. There are severe shifts of the heart and other midline structures secondary to hyperplasia of the healthy lung. This shift becomes important in patients with pulmonary agenesis who need cardiac surgery. There are some reports of mitral valve surgery in patients with pulmonary agenesis [5, 6]. To our knowledge, there have been no published cases of aortic valve and tricuspid valve surgery within this population. A median sternotomy was chosen in these previous reports. This approach has several advantages in that CPB is easily established through a median sternotomy and provides an adequate field of view. However, there are several disadvantages such as an increased distance to the heart and the fact that the right lung crosses over into the left side of the chest. Thus, we chose to approach the heart via a left thoracotomy. This approach makes access easier because of the shorter distance to the heart and the lack of interference from the right lung. Aortic cannulation and bi-caval cannulation might be possible via left thoracotomy, but those cannulations were established at a femoral site to make the better field of view. In this case, a left thoracotomy was useful and safe.

Conclusion

A rare case of cardiac surgery with left pulmonary agenesis was presented. We performed aortic valve replacement, tricuspid valve annuloplasty and right pulmonary vein isolation via a left thoracotomy. This approach was useful and safe.

Compliance with ethical standards

Conflict of interest The authors have declared that no conflict of interest exists.

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