

## Surgical Correction of Cor Triatriatum Associated with Pulmonary Artery Thrombosis in an Adult

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We herein present a case of a successful correction of cor triatriatum associated with thrombotic pulmonary hypertension diagnosed in an adult female patient. We confirmed diagnosis using transthoracic and transesophageal echocardiography in addition to cardiac computed tomography and magnetic resonance imaging. Surgical repair comprised excision of the fibromuscular membranous septum in the left atrium, patch closure of an atrial septal defect, and reconstruction of the pulmonary arteries with a vascular graft. Cor triatriatum complicated pulmonary thrombotic hypertension with atrial septal defect is amenable to surgical correction with satisfactory results.

Key words: 1. Pulmonary arteries  
2. Heart septal defects, atrial  
3. Hypertension, pulmonary  
4. Thrombosis  
5. Cor triatriatum

### CASE REPORT

A 44-year-old woman was admitted to our hospital for assessment of an over 10-year history of fatigue, severe dyspnea on exertion, functional impairment class III (NYHA), and intermittent hemoptysis. When the patient was admitted to the hospital, she could not walk even 4 or 5 meters and had been misdiagnosed with Eisenmenger syndrome by various clinics. The patient also had clubbing of her fingers and toes. During her physical examination, she displayed hypotension (91/60 mmHg) with a regular beat of 70 beats/min. Oxygen saturation on room air was approximately 76% and PaO<sub>2</sub> 41.8 mmHg based on an arterial blood gas analysis (ABGA). Cardiac auscultation revealed a grade III systolic ejection

murmur at the left upper parasternal border and a slightly accentuated second heart sound. An electrocardiogram showed a sinus rhythm with a right bundle-branch block, right axis deviation, right atrial enlargement, and an S1Q3T3 pattern (an S wave in lead I, a Q wave, and an inverted T wave in lead III). Chest radiography displayed significant cardiomegaly, especially a right-sided heart enlargement and pulmonary venous congestion. A transthoracic two-dimensional echocardiography (TTE) revealed an abnormal linear echo density membrane, which divided the proximal superior common pulmonary venous chamber and the distal true left atrial cavity (Fig. 1). It also showed a moderate degree of tricuspid insufficiency with severe pulmonary hypertension, in which the right ventricle systolic pressure (RVSP) was estimated at 122

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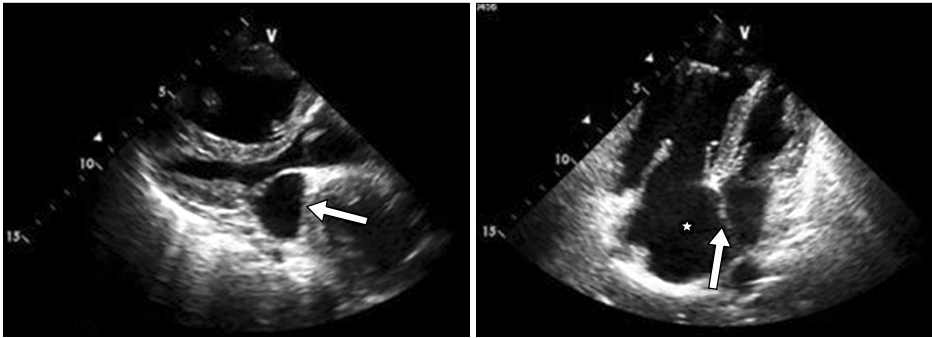
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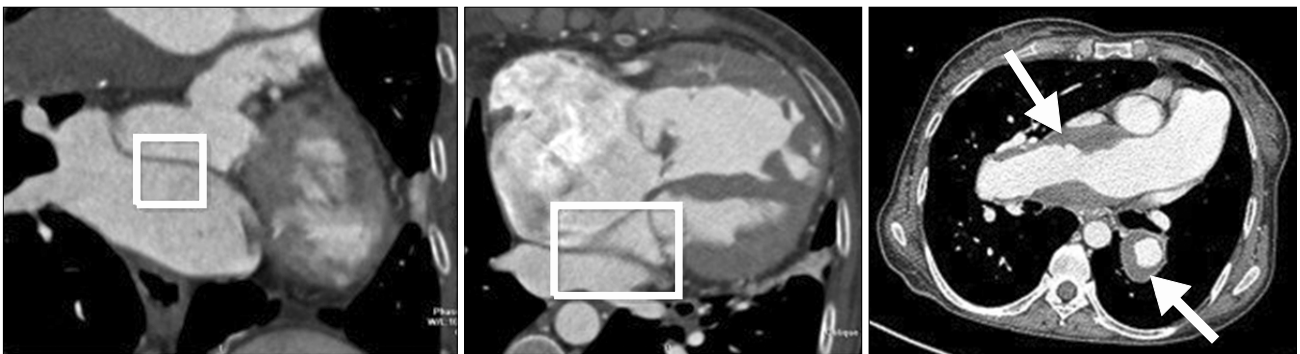
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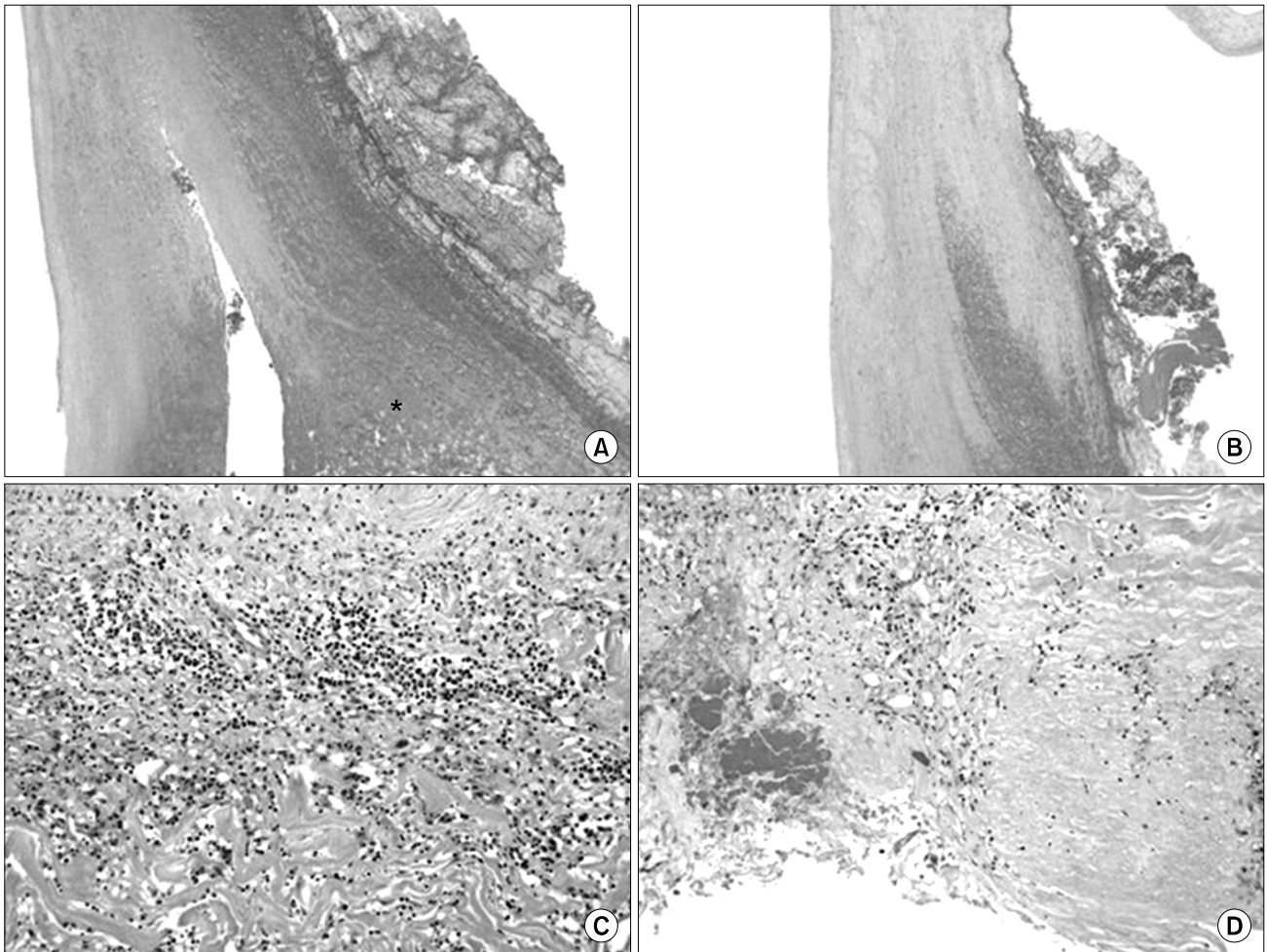
**Fig. 1.** A cross-sectional echocardiography shows the dividing membrane in left atrium (arrows) and atrial septal defect (asterisk).



**Fig. 2.** Computed tomography shows divided left atrium (square line) and huge thrombus (arrow) in pulmonary artery.

mmHg. This condition was verified by transesophageal echocardiography, which revealed cor triatriatum with a large atrial septal defect (ASD) sized at 35 mm, which was communicating between the right atrial chamber and both proximal and distal chambers of the left side. Prior to surgery, cardiac computed tomography (CT) and magnetic resonance imaging scans were performed. The procedures confirmed the diagnosis of cor triatriatum, ASD, and severe pulmonary hypertension with pulmonary artery aneurysm containing a large intraluminal thrombus, which caused nearly total obstruction of both pulmonary artery passages (Fig. 2). Extremely dilated pulmonary arteries and left-side shifting of interventricular septum were visible, but otherwise the left ventricular function was normal. The ratio of pulmonary blood flow to systemic blood flow (QP/QS) was 1.33. No further cardiovascular anomaly was depicted. The patient elected to undergo a surgical operation consisting of a median sternotomy. After a cardiopulmonary bypass (CPB) was initiated, cardioplegia was then induced under deep hypothermia by infusing a cold blood cardioplegic solution into the aortic root. A

large ASD was discovered, and the anomalous membrane in the left atrium was examined through the ASD. The membrane was successfully resected and the atrial septal defect was then closed with a bovine pericardial patch. We subsequently continued to perform a thrombectomy in the main pulmonary artery (MPA) and in the right and left pulmonary arteries (RPA & LPA), which were nearly obstructed by numerous thrombi located even in the distal branches of both pulmonary arteries. We also noticed severe dilatation sized at 5 cm in diameter from the distal MPA to both the RPA and LPA, which were very firm and stiff on palpation. After resection of the extremely dilated MPA with both pulmonary arteries, a complete thrombectomy was performed, even in the distal branches of both pulmonary arteries by suction. A 24 mm vascular intergard interposition graft was then placed in an end-to-end fashion between the main pulmonary artery and the right pulmonary artery. The left pulmonary artery was anastomosed successively to the 18 mm vascular interposition graft in an end-to-side fashion. At the microscopic level, diffuse formations of fibrinoid thrombi were found in



**Fig. 3.** Histological findings of the resected pulmonary artery. Microscopically, diffuse formations of fibrinoid thrombi (asterisk) were found in the arterial wall showing fibrosclerotic degeneration (A, B). There were also infiltration of chronic inflammatory cells, medial fibrosis, and intimal thickening in the cross-section of arterial walls, suggesting arteriosclerosis due to longstanding pulmonary hypertension (C, D). Hematoxylin-Eosin stain, A, B:  $\times 200$ , C, D:  $\times 400$ .

the arterial wall showing fibrosclerotic degeneration. An infiltration of chronic inflammatory cells, medial fibrosis, and intimal thickening in the cross-section of the arterial wall were also visible, which suggested the presence of arteriosclerosis due to longstanding pulmonary hypertension (Fig. 3). The pulmonary artery thrombectomy and reconstruction were performed in under 35 minutes of total circulatory arrest at  $15.0^{\circ}\text{C}$ . The cardiopulmonary bypass time was 290 minutes and aortic cross-clamping time was 202 minutes. Mechanical ventilation was applied postoperatively for 5 days. The patient was transferred from the ICU to the general ward 13 days after the operation.

Pulmonary hypertension rapidly regresses after surgery. Using a Swan-Ganz catheter to estimate systolic pulmonary arterial pressure indicated  $50\sim 55$  mmHg within 3 days after surgery, which decreased remarkably when compared to the 122 mmHg revealed by preoperative echocardiography. A postoperative follow-up echocardiogram and CT revealed satisfactory results by displaying no blood flow disturbance in the left atrium, successful resection of the obstructing membrane, and no remnants of thrombus in the pulmonary arteries (Fig. 4). The patient's oxygen saturation in room air was 98%,  $\text{PaO}_2$  90 mmHg, and PH 7.44 checked using ABGA 10 days after surgery. Her exercise tolerance significantly im-

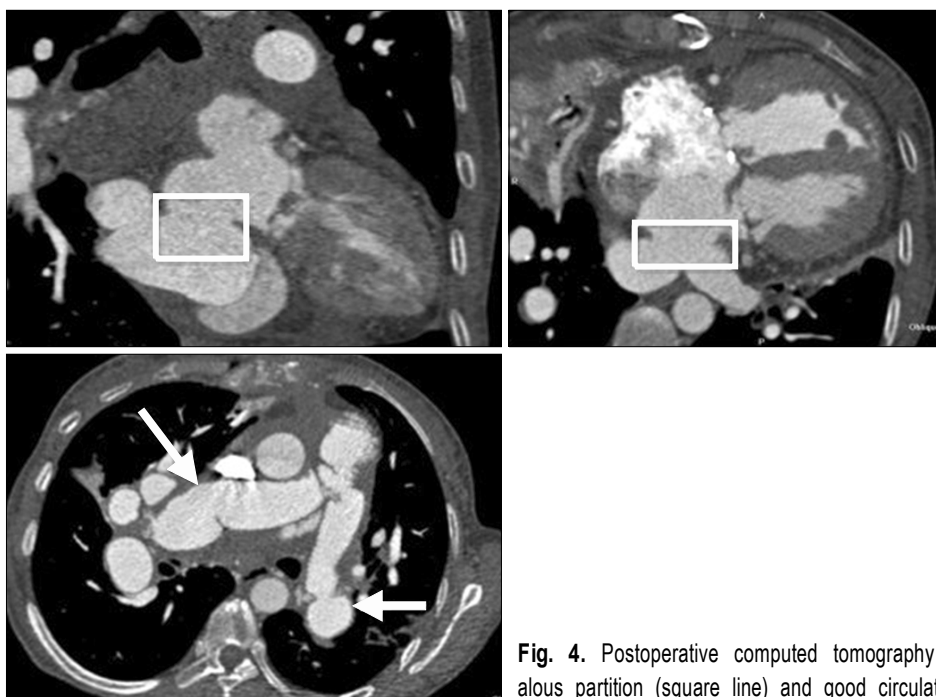


Fig. 4. Postoperative computed tomography shows successful resection of anomalous partition (square line) and good circulation of pulmonary artery (arrows).

proved from 20~30 meters of walking distance to 300 meters for 6 minutes by approximately 3 weeks after surgery. She was prescribed Tracleer<sup>®</sup> (bosentan) to manage pulmonary hypertension and within the 6 months after the operation she was in good condition during periodic visits to an outpatient clinic.

## DISCUSSION

Cor triatriatum is a rare congenital anomaly characterized by an abnormal separation of the left atrium into a proximal pulmonary venous chamber and a distal true chamber [1]. Approximately 75% of patients with such malformations die in infancy, because symptoms usually develop early, therefore surgical operation is requisite during the first year of a patient's life. When older patients present with chronic symptoms, immediate surgical correction is also necessary [2]. The embryogenesis of cor triatriatum remains controversial and is usually explained by either entrapment theory, as proposed by Van Praagh and Corsini [3], or by a failed incorporation of the common pulmonary veins into the left atrium [4]. Though it is being diagnosed with increased frequency in adulthood, cor triatriatum remains a rare form of congenital heart

disease. In a review by Niwayama of 36 patients diagnosed with the disease, ten were older than 12 years of age [5]. Our 44-year-old patient represents one of the oldest reported living patients. In this report, we present a 44-year-old woman with cor triatriatum, a large atrial septal defect (ASD) sized at 35 mm accompanied by chronic pulmonary artery thrombosis. In about 75% of patients there is an atrial septal defect between the right atrium and the proximal venous accessory chamber or the true distal atrium. However, cor triatriatum with severe pulmonary hypertension complicated by chronic pulmonary artery thrombosis is very rare. Pulmonary hypertension with an atrial septal defect might not have been influenced by pulmonary venous obstruction because there was a large atrial septal defect (ASD) of 35 mm, which looked like a common atrium and communicated between the right atrial chamber and both the proximal and distal chambers on the left. Proximal pulmonary artery thrombosis has been reported in patients with ASD and pulmonary artery hypertension (PAH) [6]. In the present case, the thrombi were considered to have formed in situ as there were no symptoms suggesting systemic embolization of such large thrombi. The aggravation of symptoms could have been caused by the increased obstruction of blood flow by the thrombi and minute

embolizations from the large thrombi to the peripheral pulmonary circulation. We could easily identify thrombi and minute embolizations into the distal pulmonary arteries by a pre-operative computed tomography (CT) and in the operative field as well. Endothelial dysfunction induced by PAH, abnormal coagulation factors, and platelet dysfunction have been determined to be the causes of thrombosis in secondary PAH [7]. The relatively sluggish blood flow in the dilated pulmonary arteries and polycythemia might have contributed to the formation of such large thrombi in this patient. The natural history of large in situ thrombi in the pulmonary arteries is unknown, as only a few case reports are available. In patients with PAH and pulmonary embolism due to such large thrombi, acute worsening of symptoms with right heart failure may lead to death in weeks to months, or occasionally, sudden death may occur. In summary cor triatriatum is a rare disease associated with chronic thrombotic pulmonary hypertension that can be treated successfully if the diagnosis is done well and sufficient information is available preoperatively.

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