

➤ **Case Report** ◀

Pulmonary Stenosis Caused by Ductus Arteriosus Aneurysm: A Case Report

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A 76-year-old woman with a 2-week history of dyspnea on exertion was admitted to our hospital. A computed tomography scan showed a 70-mm diameter aortic arch aneurysm containing a large thrombus that was compressing the pulmonary artery. Echocardiography showed severe pulmonary stenosis and no shunt flow. Operative findings revealed an aneurysmal thrombus protruding into the lumen of the pulmonary artery through a foramen. A ductus arteriosus aneurysm was diagnosed. After the thrombus removal, arch replacement and ductus closure with a prosthetic patch were performed. Histological examination showed that the thrombus had no vascular components. The patient's symptoms were relieved, and she was discharged.

Keywords: ductus arteriosus aneurysm, pulmonary stenosis, heart failure

Introduction

A ductus arteriosus aneurysm is a congenital cardiac abnormality with an 8.8% reported incidence among full-term neonates.¹ Ductus arteriosus aneurysms diagnosed in the neonatal period are often asymptomatic and have a benign natural course, resulting in spontaneous regression and disappearance in most cases.^{1,2} Rarely, a ductus arteriosus aneurysm may present with symptoms requiring medical attention. Reported symptoms include thromboembolism, infection, compression of an adjacent structure, and spontaneous rupture.^{1–6} However, the full clinical picture remains unclear. Here, we report a rare case of a large ductus arteriosus aneurysm complicated by

pulmonary stenosis and resultant heart failure in an adult patient.

Case Report

A 76-year-old woman diagnosed with an aortic arch aneurysm was admitted to our hospital for worsening dyspnea on exertion. The patient presented without hoarseness or symptoms of airway obstruction. The dyspnea had started 2 weeks before admission and gradually worsened until provoked by light exercise. The patient was referred to our department for treatment. Upon admission, the physical examination showed a blood pressure of 95/70 mmHg, heart rate of 100 beats/min, and oxygen saturation of 95% as measured by pulse oximetry. After light exercise, her oxygen saturation decreased to 90%. Laboratory tests showed increased liver transaminase (64 U/L and 100 U/L of aspartate aminotransferase and alanine aminotransferase, respectively) and brain natriuretic peptide concentrations (1967 pg/mL). A computed tomography (CT) scan showed that a large 70-mm diameter saccular aortic arch aneurysm had substantially compressed the periphery of the pulmonary artery and that a large thrombus had protruded into the main trunk of the pulmonary artery. No other CT findings of chronic pulmonary thromboembolism, including abrupt narrowing, obstruction, or calcified thrombi, were found in the pulmonary artery branches (Fig. 1). Echocardiography showed preserved left ventricular ejection fraction of 61.5%; decreased left ventricular dimension of 26.6 and 18.2 mm in diastole and systole, respectively; profound pulmonary hypertension characterized by a right ventricular systolic pressure of 95.9 mmHg; moderate to severe tricuspid regurgitation; and pulmonary stenosis indicated by a peak velocity of 3.84 m/s and a peak pressure gradient of 59.2 mmHg. Echocardiography did not show intracardiac or extracardiac left-to-right shunt flow. The patient was preoperatively diagnosed with a large saccular aortic arch aneurysm that caused the pulmonary stenosis. An urgent aortic repair was scheduled on day 6 of hospitalization.

During anesthetic induction, the patient's systolic blood

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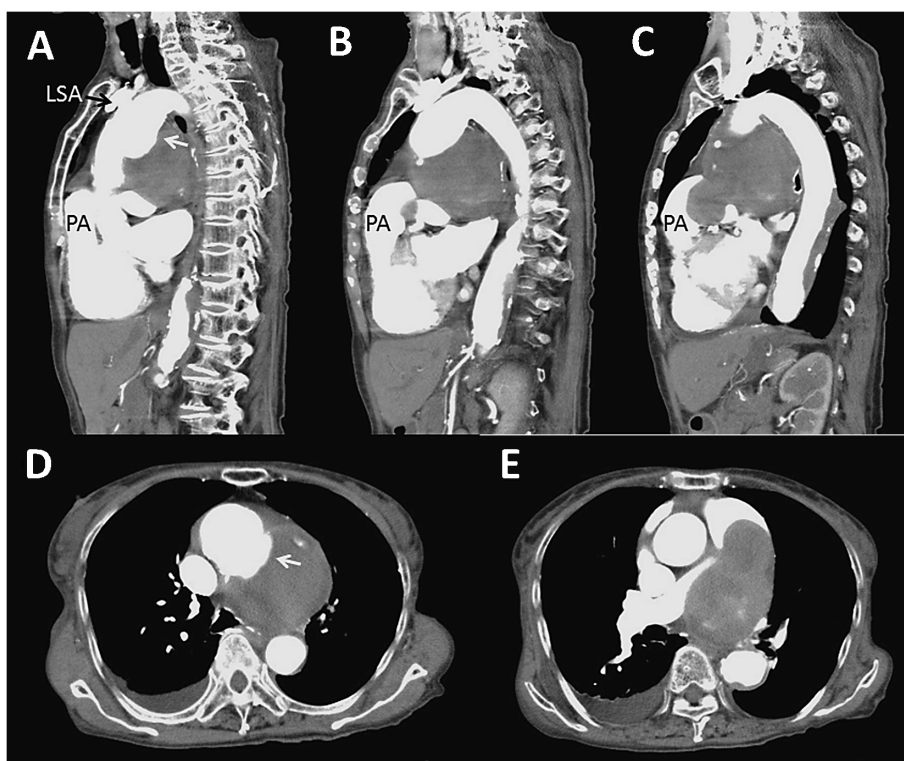


Fig. 1 Preoperative computed tomography images. The sagittal images showed the aneurysm entrance at the lesser curvature of the aortic arch (**A**, white arrow) and a protruding thrombus narrowing the main trunks of the pulmonary artery (**B**, **C**). The axial images showed the aneurysm entrance (**D**, white arrow) and a large thrombus compressing the main trunk of the pulmonary artery (**E**). LSA: left subclavian artery; PA: pulmonary artery

pressure was temporally decreased to 40 mmHg. During the surgery, transesophageal echocardiography showed no intracardiac or extracardiac left-to-right shunt flow. A median sternotomy was performed, and a serous pericardial effusion was found in the pericardial space. The ascending aorta was intact on inspection. A cardiopulmonary bypass was established with an ascending aorta cannulation, bicaval venous cannulations, and placement of a left ventricular vent via the right superior pulmonary vein. The patient's body temperature was cooled down to a tympanic temperature of 26°C, and the cardiopulmonary bypass was arrested. The ascending aorta was incised, and a selective cerebral perfusion via the three supra-arch branches was initiated; the cerebral circulatory arrest time was 5 min. Aortic arch opening revealed a thrombus filling the entrance of the saccular aneurysm at the lesser curvature of the aortic arch. First, the aortic wall was trimmed at the distal portion of the thoracic aorta over the entrance of the saccular aneurysm, and a four-branched 26-mm vascular prosthesis (Japan Lifeline Co., Ltd., Tokyo, Japan) was then anastomosed to the distal thoracic aorta. After the distal anastomosis, the lower body perfusion was resumed via a side branch of the prosthesis; lower body ischemia

time was 59 min. After removing the thrombus from the aneurysm, a 3-cm diameter foramen was detected at the bottom of the aneurysmal sac in contact with the lumen in the main trunk of the pulmonary artery (**Fig. 2B**). No obvious pus or tumor was present inside or surrounding the aneurysmal cavity. She was diagnosed with ductus arteriosus aneurysm. The pulmonary stenosis was considered to have been caused by not only the external compression but also the thrombus protrusion into the pulmonary artery. The foramen in the anterior wall of the pulmonary artery was closed using a vascular patch (**Fig. 2C**) and proximal anastomosis was performed. Following tricuspid annuloplasty, the vascular prosthesis was declamped; the myocardial ischemia time was 170 min. The three supra-arch vessels were reconstructed. The duration for cardiopulmonary bypass, selective cerebral perfusion, and surgery were 274, 199, and 440 min, respectively.

The postoperative CT scan showed disappearance of the pulmonary artery stenosis and successful placement of the three-branched vascular prosthesis (**Fig. 3**). Echocardiography confirmed a 33-mmHg decrease in right ventricular systolic pressure and reduction in trivial tricuspid regurgitation. The preoperative dyspnea experienced by

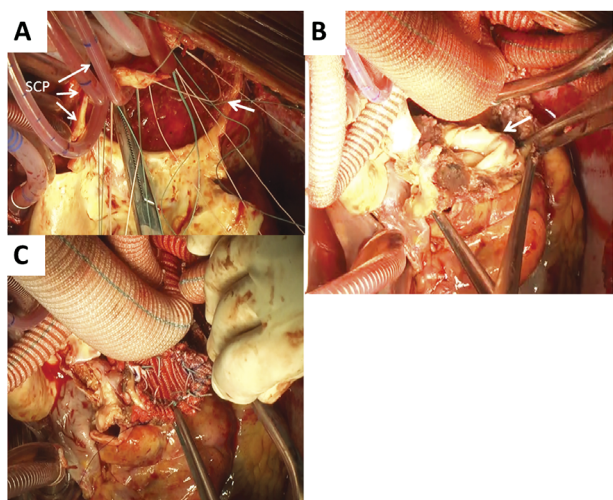


Fig. 2 Intraoperative image. **(A)** Aortic opening revealed a thrombus filling the entrance (white arrow) of the saccular aneurysm at the lesser curvature of the aortic arch. **(B)** After the distal anastomosis and thrombus removal, a foramen with a diameter of approximately 3 cm (white arrow) was seen at the bottom of the aneurysmal sac in contact with the lumen of the pulmonary artery. **(C)** The foramen was closed with a vascular patch. SCP: selective cerebral perfusion

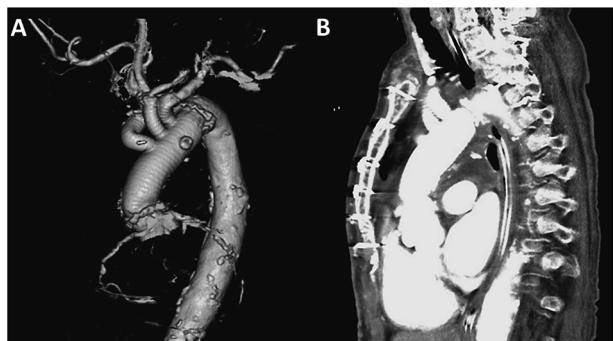


Fig. 3 A computed tomography image after the aortic arch replacement. **(A)** The three-dimensional computed tomography image demonstrated no aneurysm post aortic arch replacement. **(B)** The sagittal reconstruction showed resolution of the pulmonary stenosis.

the patient on exertion was relieved. The pathological examination showed that the thrombus protruding into the pulmonary artery was composed of thrombotic materials alone without any vascular wall components. Postoperatively, the patient experienced a parietal lobe stroke. However, she gradually recovered after rehabilitation and was discharged on postoperative day 61.

Discussion

A ductus arteriosus aneurysm is a rare congenital cardiac abnormality and genetic factors may play a role in its de-

velopment. Underlying genetic abnormalities including trisomy 21, trisomy 13, Smith–Lemli–Opitz syndrome, type IV Ehlers–Danlos syndrome, and Marfan syndrome are reportedly present in one-fourth of affected patients.⁷⁾ Previous studies have shown that the aneurysmal wall of the ductus arteriosus contains poorly functional elastic fibers and media degeneration characterized by mucoid deposition in contrast to a healthy aorta or pulmonary artery.^{8,9)} These findings suggest that the intrinsic vulnerability of the ductus arteriosus may be related to the aneurysm formation in the neonatal or infantile period. In contrast, the etiologies of ductus arteriosus aneurysms in adults remain unclear. The aforementioned pathological fragility of the ductus arteriosus aneurysmal wall in infants may contribute to the formation of a larger aneurysm in adults. It is believed that the ductus closure process begins from one end of the pulmonary artery and progresses toward the other end of the aorta; however, the ductus closure may have been incomplete, leading to an aortic diverticulum or patent ductus arteriosus.²⁾ A ductus arteriosus aneurysm may also result from delayed closure of the aortic end of the ductus arteriosus with exposure of the ductal wall to systemic arterial pressure.¹⁰⁾ Furthermore, hypertension and atherosclerotic changes in the aortic wall may be related to the ductus arteriosus dilatation and the distal aortic arch in adults.⁹⁾ Although unrelated to the present case, a ductus arteriosus aneurysm in adults may also develop following infective endarteritis, surgical closure, or transcatheter coil occlusion of patent ductus arteriosus.²⁾

The most common age at diagnosis of a ductus arteriosus aneurysm is <2 months, and most cases are incidentally identified.¹⁾ However, a favorable prognosis is reported in neonatal patients with ductus arteriosus aneurysms. Jan et al.¹⁾ reported that all 48 neonates with a ductus arteriosus aneurysm incidentally detected by echocardiography showed spontaneous regression and complete disappearance of the aneurysm by 1 month after birth. Symptomatic ductus arteriosus aneurysms are uncommon, especially in adults, but can lead to serious cardiovascular adverse events requiring medical treatment. Obstruction or narrowing of the adjacent vasculature is a rare complication of ductus arteriosus aneurysms. McArdle et al.⁶⁾ reported a case involving a neonate with a thrombosed ductus arteriosus aneurysm measuring 7.8 × 8.2 mm, inducing a mass effect with resultant pulmonary stenosis. In the present case, our initial diagnosis was severe pulmonary stenosis caused by a giant saccular aortic arch aneurysm because the preoperative CT scan did not show the patent ductus arteriosus. The intraoperative findings revealed a 3.0-cm thrombosed fistula between the pulmonary artery and an aortic arch aneurysm, leading to the definitive diagnosis of a ductus arteriosus aneurysm. The thrombosed ductus arteriosus resulted in the absence of the left-to-right shunt,

rendering it difficult for an accurate preoperative diagnosis to be made. We propose that the thrombosed ductus arteriosus aneurysm had gradually increased in size until it reached a hazardous point at which it caused a profound pulmonary stenosis, resulting in heart failure. The thoracic aortic aneurysm rupture and its impact on the pulmonary embolism present an alternative explanation for the heart failure. Comparatively, this condition may present as more fulminant. However, the gradually developing dyspnea and no extracardiac shunt flow support the diagnosis that the ductus arteriosus aneurysm may have led to the development of pulmonary stenosis.

Various procedures have been reported in the treatment of ductus arteriosus aneurysms. Endovascular closure of the ductus with a stent graft is an established treatment option for patients with ductus arteriosus aneurysms.²⁾ Roques et al.¹¹⁾ reported that stent-grafting in the proximal descending thoracic aorta successfully closed the inflow of the ductus arteriosus aneurysm in an adult patient, resulting in the improvement of pulmonary hypertension. Compared with these transcatheter closure techniques, open surgical closure often requires cardiopulmonary bypass and is performed through a median sternal or anterior thoracic approach, carrying a high morbidity risk especially in elderly patients. We successfully performed open surgical resection of a ductus arteriosus aneurysm and concomitant aortic arch replacement based on the preoperative diagnosis of an aortic arch aneurysm. Even when a ductus arteriosus aneurysm is preoperatively suspected, open surgical treatment aiming to achieve a complete thrombus removal would be an appropriate approach to eliminate the future risk of rupture and relieve the pulmonary stenosis. Our patient showed remarkable hemodynamic improvement as observed by postoperative echocardiography. We propose that transcatheter treatment would not have been feasible in the present case because of the patient's worsening heart failure.

Conclusion

In conclusion, we experienced a very rare case of severe pulmonary stenosis caused by a large unruptured ductus arteriosus aneurysm. The patient successfully underwent an open surgical resection of the aneurysm, concomitant aortic arch replacement, and pulmonary artery reconstruction with a prosthetic patch without the development of a circulatory collapse or critical thromboembolism.

Disclosure Statement

None of the authors declare any conflicts of interest.

Author Contributions

Study conception: YT, NK

Data collection: YT

Analysis: YT, NK

Investigation: YT, NK

Writing: YT, NK

Funding acquisition: no

Critical review and revision: all authors

Final approval of the article: all authors

Accountability for all aspects of the work: all authors

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