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Case Report

A Case of Surgical Treatment for a Large Pulmonary Artery Aneurysm with a Quadricuspid Pulmonary Valve

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A 65-year-old man was referred to our hospital for the annual assessment of the diameter and dilation of a pulmonary artery (PA) aneurysm. He had a small ventricular septal defect (VSD) that had closed naturally. Echocardiography revealed a dilated main PA, mild pulmonary regurgitation and no VSD. Computed tomography confirmed the dilation of the main PA (66.7×47.8 mm), right PA (37.1×32.9 mm), and left PA (36.7×34.0 mm). The patient underwent pulmonary artery replacement using a prosthetic vascular graft. A quadricuspid pulmonary valve was identified intraoperatively. Early surgical intervention could help to prevent rupture and dissection of PA aneurysms.

Key words: pulmonary artery aneurysm, quadricuspid pulmonary valve, pulmonary valve regurgitation and stenosis, congenital heart disease, pulmonary artery graft replacement

Pulmonary artery aneurysm (PAA) is a rare condition that arises from various etiologies, including congenital heart disease, connective tissue disorders, autoimmune disorders, vasculitis, iatrogenic causes, trauma, infection, inflammation, malignancy and idiopathic factors [1,2]. Rupture of a PAA is frequently fatal. In previous report, one-third of cases of PAA rupture resulted in death [2]. Additionally, coronary artery compression and pulmonary artery (PA) dissection are life-threatening complications. Given these risks, early intervention is essential [1,2].

We report the case of a patient who developed a large PAA that was successfully treated, preventing severe complications.

Case Report

The patient was a 65-year-old man. At the age of 60, a chest X-ray revealed enlargement of the left first cardiac silhouette border, and a computed tomography (CT) scan showed an enlarged PA. His referral physician initiated annual monitoring of the PA diameter with CT scans. The patient was referred to our department at age 65 for further evaluation and assessment of surgical indications due to the aneurysm's large diameter and PA dilation. He had a history of a small ventricular septal defect (VSD) that had closed spontaneously. His medical history also included hypertension managed with an angiotensin II receptor blocker and a calcium channel blocker, diabetes treated with metformin and dapagliflozin, prostatectomy and asymptomatic cerebellar hemorrhage.

Echocardiography revealed a dilated main PA with a diameter of 60 mm, mild stenosis of the PA valve (1.8 m/s), mild regurgitation of the PA valve, no VSD, and a 65% left ventricular ejection fraction (EF). CT showed the dilated main PA (66.7×47.8 mm), dilated

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right PA (37.1×32.9 mm), and dilated left PA (36.7×34.0 mm) without findings of coronary artery stenosis, PA inflammation, infection, or tumor (Fig. 1). Cardiac angiography revealed a main PA pressure of 19/11/ (12) mmHg in systole/diastole (mean), a right PA pressure of 24/10/ (15) mmHg, a left PA pressure of 22/8/ (11) mmHg, a right ventricular (RV) pressure of 37/11 mmHg, and a mean right atrial (RA) pressure of 11 mmHg, all of which were within normal limits, no VSD (pulmonary blood flow/systemic blood flow = 1.00) and no coronary stenosis. Cardiac magnetic resonance imaging (MRI) showed an RV end diastolic volume index of 60.08 ml/m^2, an RV end systolic volume index of 37.36 ml/m^2, both of which were within normal limits, and an RV EF of 37.8%, which was low.

Considering the risk of rupture, dissection and/or coronary compression due to the large PAA, the decision was made to resect the PAA and replace it using a prosthetic vessel graft. A median sternotomy was performed under general anesthesia. Cardiopulmonary bypass was established between the ascending aorta, superior vena cava (SVC), and inferior vena cava. The patient's body temperature was cooled to 34°C. The main and bilateral PA diameters were enlarged (Fig. 2A). The dilated main PA was transected above the PA valve, revealing quadricuspid pulmonary valve (QPV) with mildly thickened but smoothly opening leaflets and adequate coaptation (Fig. 3). As the valve showed no significant stenosis or regurgitation, valve intervention was deemed unnecessary. The left PA was transected 10 mm proximal to its first branch, and a 26 mm prosthetic graft was selected to match the proximal anastomosis. The graft was anastomosed to the thin-walled left PA using a felt strip. Then the main PA was anastomosed to the graft using 4-0 polypropylene. The right PA was transected at the level between the SVC and the ascending aorta and anastomosed to the graft. The proximal end of the graft anastomosed to the right PA was subsequently anastomosed to the main PA graft in end-to-side fashion (Fig. 2B). The cardiopulmonary bypass was weaned uneventfully. The total bypass time was 136 min.

The patient was extubated the day after surgery and his length of stay in the intensive care unit was three days. The drain was removed on postoperative day 4 and oral intake was resumed the same day. CT and

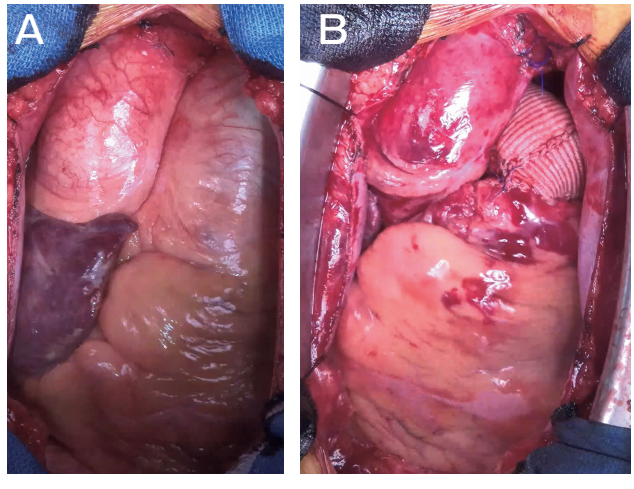

Fig. 2 A, PAA before operation; B, Reconstructed PA with the prosthetic graft.

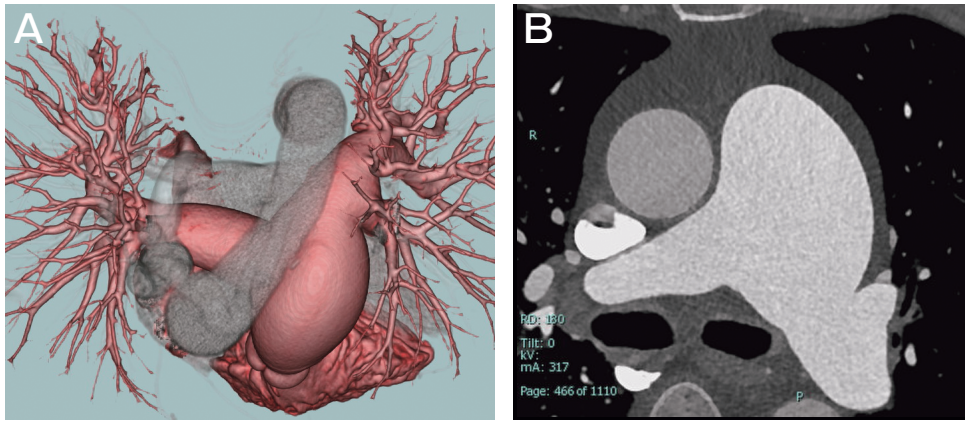

Fig. 1 A, Three-dimensional computed tomography (CT) showing pulmonary artery aneurysm (PAA); B, CT showed a dilated main pulmonary artery (PA) (66.7 × 47.8 mm), right PA (37.1 × 32.9 mm), and left PA (36.7 × 34.0 mm).

echocardiography were performed on postoperative day 8. Postoperative echocardiography showed trivial regurgitation and no stenosis in the pulmonary valve, which was consistent with preoperative findings. CT showed no evidence of residual aneurysm or pseudoaneurysm (Fig. 4). The histopathological findings of the PAA wall included mild mucinous change in the tunica media without fibrotic tears or inflammation. The patient was discharged on postoperative day 19 uneventfully.

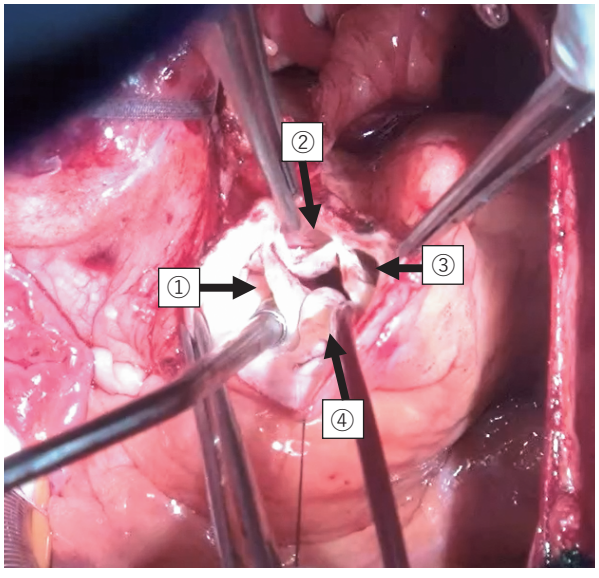

Fig. 3 The PA was transected above the PA valve. Quadricuspid pulmonary valve was observed.

Discussion

PAA is a rare vascular anomaly. Deterling and Clagett report only 8 cases in 109,571 consecutive postmortem examinations [3]. More than half of PAA cases are associated with congenital heart disease, including VSD, atrial septal defect, persistent ductus arteriosus, hypoplastic or absent pulmonary valve, and hypoplastic aortic valve [2].

In our case, the patient had congenital VSD and QPV with mild regurgitation, which might have contributed to the formation of the aneurysm. Histopathological results support our hypothesis about the

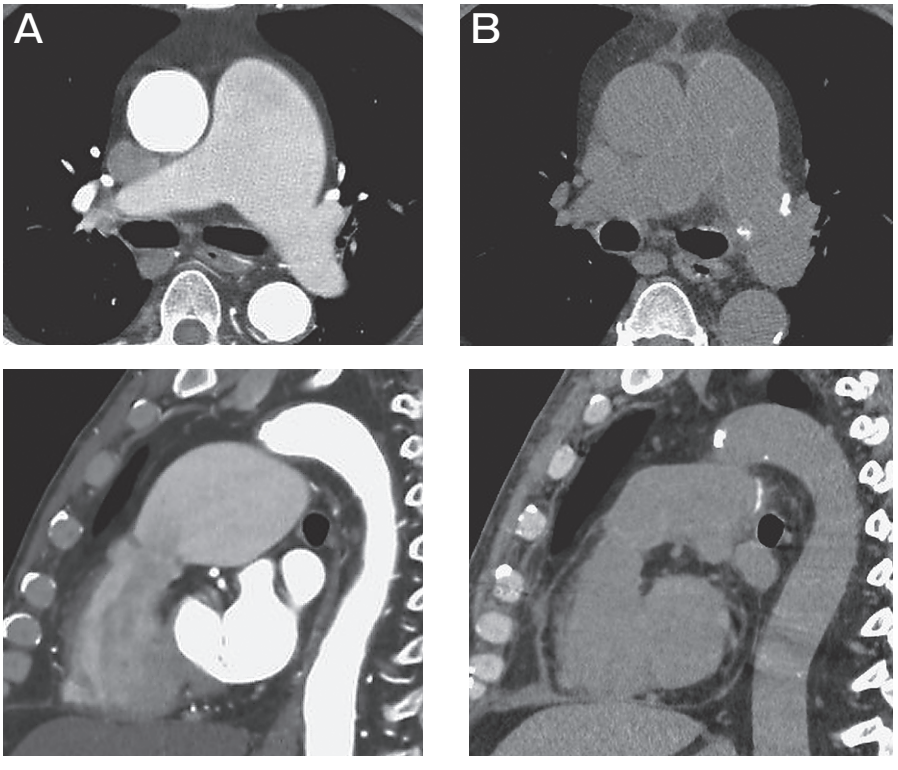

Fig. 4 Postoperative CT shows replacement of the PA with a prosthetic graft, in comparison with the native PA observed on preoperative contrast-enhanced CT. **A**, Preoperative CT with contrast; **B**, Postoperative CT.

etiology of the PAA.

Other etiologies of PAA include connective tissue disorders (*e.g.*, Ehlers-Danlos Syndrome, Marfan Syndrome, cystic medial necrosis), vasculitis (*e.g.*, Behçet Syndrome, Hughes-Stovin Syndrome), infections (*e.g.*, syphilis, tuberculosis, pyogenic bacteria), malignancy, and iatrogenic causes (*e.g.*, cardiac surgery, catheter placement, angiography) [1, 2].

Jashari *et al.* report that QPV was found in only 0.2% of 3,861 donor hearts [4]. Other case reports of PAA associated with QPV are similarly rare [5-7]. About two-thirds of QPV cases are hypoplastic and cause pulmonary valve regurgitation, which generates PAA [6].

PAA is often asymptomatic and detected incidentally, even in cases with diameters exceeding 70 mm [2]. When symptoms do occur, they are usually nonspecific and may include chest pain, dyspnea, hoarseness, palpitations, cough, or hemoptysis.

Even though our patient showed no symptoms, severe pulmonary valvular stenosis or regurgitation, the risk of lethal complications such as PAA rupture, dissection or coronary stenosis should be considered a possible surgical indication. In the present case, the decision was made to perform aneurysmectomy and PA replacement with an artificial graft, effectively eliminating this risk.

There are currently no specific guidelines for PAA treatment. Kreibich *et al.* propose surgical indications based on an absolute PAA diameter of >5.5 cm, aneurysm growth of >0.5 cm in 6 months, adjacent structure compression, thrombus formation in the aneurysm sack, the appearance of clinical symptoms, valvular pathologies, shunt flow, PA hypertension and/or signs of rupture/dissection [2]. Aneurysmectomy with PA replacement using a conduit including a Gore-Tex tube graft, Dacron tube, homograft and xenograft, is the standard surgical approach [2]. PA valve repair or replacement may be necessary for a malfunctioning PA valve [2].

Based on these criteria, we elected to perform aneurysmectomy and PA replacement using a prosthetic graft. Although PA valve intervention was not required at the time of the surgery, postoperative follow-up for QPV and potential pseudoaneurysm formation at the anastomosis site was deemed necessary.

The present case highlights the importance of early surgical decision-making in asymptomatic but high-risk patients with a large PAA. Although the timing of intervention should be individualized, factors such as aneurysm size, growth rate, pathology, and complications such as PA valve stenosis or regurgitation should be carefully considered. We recommend early referral to specialized cardiovascular centers when any of these risk factors are present, in order to prevent irreversible or potentially fatal complications.

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