Giant Pulmonary Artery Aneurysm in a Patient With Marfan Syndrome and Pulmonary Hypertension

Peter Chiu, MD; Mallory Irons, BA; Matt van de Rijn, MD, PhD; David. H. Liang, MD, PhD; D. Craig Miller, MD

Aneurysmal dilatation of the pulmonary artery is rare.1 Management considerations, including indications for surgery, are not well defined.2,3 We present a case of giant pulmonary artery aneurysm (PAA) in a patient with pulmonary hypertension (PH) and connective tissue disorder.

Case Presentation
This is a 58-year-old woman with Marfan syndrome, FBN1 mutation c.6423delG, who, in 2004, was found to have a dilated main pulmonary artery (MPA) on chest x-ray performed for cough and wheezing. The MPA was 4.9 cm on computed tomographic angiogram; mean pulmonary artery (PA) pressure was 33 mm Hg on right heart catheterization. Transthoracic echocardiogram showed a right ventricular systolic pressure of 83 mm Hg and 2+ pulmonary regurgitation. Mitral regurgitation was mild; neither aortic regurgitation nor aortic root dilatation was present. Over the next 4 years, the MPA progressed to 7.9 cm despite initiating silde- nafil therapy for idiopathic pulmonary arterial hypertension. Operative repair was deferred because of the poor control of PH. Switching to amiodipine and ambrisentan improved her symptoms. Right ventricular systolic pressure fell to 30 mm Hg, and the MPA stabilized in size for 6 years, but cardiac magnetic resonance in 2014 showed progression to 8.4 cm with a pulmonary valve regurgitant fraction of 7% to 14% (Figure 1A and 1B). On computed tomographic angiography, the right PA and left PA were aneurysmal, 4.0 and 3.3 cm, respectively; as seen previously, there was gross deformity of the left chest wall (Figure 2A and 2B). Transthoracic echocardiogram revealed dilatation of the pulmonary sinotubular junction with cusp malcoaptation and 2+ pulmonary regurgitation. The patient was referred for surgical intervention. A grossly enlarged MPA was visible on chest x-ray (Figure 2C).

Intraoperative transesophageal echocardiogram showed a PAA (Figure 3A and 3B). PAA graft replacement was performed on cardiopulmonary bypass with the heart beating (Figure 4A and 4B). Remodeling the pulmonary sinotubular junction with a ring of 28-mm Dacron graft augmented cusp coaptation. A 24-mm knitted Dacron graft was used to replace the left PA and MPA (Figure 4C), and a 34-mm knitted graft – the diameter of the vessel in the hilum – for the right PA (Figure 4D). Post–cardiopulmonary bypass transesophageal echocardiogram showed minimal pulmonary regurgitation. Histological section revealed cystic medial degeneration and elastin loss as might be expected in the Marfan aorta (Figure 4E and 4F). Postoperatively, inhaled epoprostenol was initiated for PH, and the patient was extubated. Because of respiratory compromise, she was reintubated on postoperative day 5, and bronchoscopy with bronchoalveolar lavage revealed Citrobacter infection, which was treated with antibiotics. She was reextubated on postoperative day 9. Inhaled epoprostenol was weaned, and amiodipine and ambrisentan were restarted. Fluoroscopy documented decreased left diaphragmatic excursion without paralysis. She improved with aggressive pulmonary toilet and was discharged home on postoperative day 20. Postoperative computed tomographic angiography demonstrated no technical faults (Figure 2D and 2E). Early on after discharge, she struggled with fatigue and respiratory insufficiency that required readmission; nocturnal bilevel positive airway pressure and supplemental oxygen were instituted. Two months postoperatively, her condition improved; bilevel positive airway pressure and supplemental oxygen were discontinued. Follow-up transthoracic echocardiogram showed mild (1+) pulmonary regurgitation; right ventricular systolic pressure was 30 mm Hg. She was last seen 5 months postoperatively doing well with no limitations.

Discussion
PAAs are infrequently encountered with only 8 cases found in a series of 109,571 postmortem examinations.1 Many cases of PAA are associated with congenital heart disease; however, other important causes exist including connective tissue disorders, PH, vasculitides, and iatrogenic causes.1,2

Patients may present with respiratory concerns or symptoms related to associated processes, eg, tricuspid regurgitation, right ventricular dysfunction, etc; however, many patients remain asymptomatic.2 Dissection is a feared complication that may occur in the absence of PH.4 Kreibich et al3 suggest that up to 19% of PAAs without PH dissect; this figure is doubtful because only 10 of 52 documented PA dissections in their reference occurred in the absence of PH or iatrogenesis,4 and the incidence of PAA without PH is unclear.
Guidelines for operative repair include size >5.5 cm, rapid growth, mass effect, rupture, dissection, and concomitant operation for associated conditions.\textsuperscript{2,3} Aneuysmectomy with pulmonary valve preservation is preferred, but valve replacement may be necessary. Adequate preoperative control of PH should be mandatory. Patients managed conservatively should undergo serial imaging with noninvasive evaluation of PA pressure. However, progression of disease necessitating operation occurs often.

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**References**

**Figure 1.**
A, MR angiogram, right ventricular 3-chamber view demonstrating regurgitant jet. B, MR angiogram, right ventricular outflow tract view demonstrating an aneurysmal pulmonary artery. MR indicates magnetic resonance.

**Figure 2.**
A, Three-dimensional VR reconstruction from CTA demonstrating an 8.4-cm aneurysm of the MPA. B, Axial CTA image. C, Preoperative CXR. D, Postoperative anteroposterior projection 3D VR CTA. E, Postoperative lateral projection 3D VR CTA reconstruction. CTA indicates computed tomographic angiography; CXR, chest x-ray; MPA, main pulmonary artery; and VR, volume rendering.
Figure 3. **A**, Right ventricular inflow-outflow view demonstrating a gross enlarged pulmonary artery. **B**, Pulmonary artery aneurysm measuring 8.8 cm.
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