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

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Keywords

Marfan syndrome; pulmonary artery; pulmonary hypertension; valve repair

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 (CXR) performed for cough and wheezing. The MPA was 4.9cm on computed tomographic angiogram (CTA); mean pulmonary artery pressure was 33 mmHg on right heart catheterization. Transthoracic echocardiogram (TTE) showed a right ventricular systolic pressure (RVSP) of 83 mmHg and 2+ pulmonary regurgitation (PR). Mitral regurgitation was mild; neither aortic regurgitation nor aortic root dilatation was present. Over the next 4 years, the MPA progressed to 7.9cm despite initiating sildenafil therapy for idiopathic pulmonary arterial hypertension. Operative repair was deferred due to poor control of PH. Switching to amlodipine and ambrisentan improved her symptoms. RVSP fell to 30 mmHg, and the MPA stabilized in size for 6 years, but cardiac magnetic resonance in 2014 showed progression to 8.4cm with a pulmonary valve regurgitant fraction of 7-14% (Figures 1A and B). On CTA, the right (RPA) and left PA (LPA) were aneurysmal, 4.0 and 3.3cm, respectively; as seen previously, there was gross deformity of the left chest wall (Figures 2A and B). TTE revealed dilatation of the pulmonary sinotubular junction (STJ) with cusp

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malcoaptation and 2+ PR. The patient was referred for surgical intervention. A grossly enlarged MPA was visible on CXR (Figure 2C).

Intraoperative transesophageal echocardiogram (TEE) showed a PAA (Figure 3A and B). PAA graft replacement was performed on cardiopulmonary bypass (CPB) with the heart beating (Figures 4A and B). Remodeling the pulmonary STJ with a ring of 28mm Dacron graft augmented cusp coaptation. A 24mm knitted Dacron graft was used to replace the LPA and MPA, (Figure 4C) and a 34mm knitted graft –the diameter of the vessel in the hilum– for the RPA (Figure 4D). Post-CPB TEE showed minimal PR. Histologic section revealed cystic medial degeneration and elastin loss as might be expected in the Marfan aorta (Figure 4E and F). Postoperatively, inhaled epoprostenol was initiated for PH, and the patient was extubated. Due to respiratory compromise, she was reintubated on postoperative day (POD) #5, and bronchoscopy with bronchioloalveolar lavage revealed *citrobacter* infection, which was treated with antibiotics. She was re-extubated POD #9. Inhaled epoprostenol was weaned, and amlodipine and ambrisentan were restarted. Fluoroscopy documented decreased left diaphragmatic excursion without paralysis. She improved with aggressive pulmonary toilet and was discharged home on POD #20. Postoperative CTA demonstrated no technical faults (Figures 2D and E). Early on after discharge, she struggled with fatigue and respiratory insufficiency requiring 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 TTE showed mild (1+) PR; RVSP was 30mmHg. She was last seen 5 months postoperatively doing well with no limitations.

Discussion

PAAAs are infrequently encountered with only 8 cases found in a series of 109,571 post-mortem examinations.¹ Many cases of PAA are associated with congenital heart disease; however, other important causes exist including connective tissue disorders, PH, vasculitides, and iatrogenesis.^{1, 2}

Patients may present with respiratory complaints or symptoms related to associated processes, e.g., TR, 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 al suggest that up to 19% of PAAs without PH dissect,² this figure is doubtful as 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 >5.5cm, rapid growth, mass effect, rupture, dissection, and concomitant operation for associated conditions.^{2, 3} Aneurysmectomy 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 non-invasive evaluation of PA pressure. However, progression of disease necessitating operation occurs often.

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References

1. Deterling RA Jr, Clagett OT. Aneurysm of the pulmonary artery; review of the literature and report of a case. *Am Heart J.* 1947; 34:471–499. [PubMed: 20266464]
2. Kreibich M, Siepe M, Kroll J, Hohn R, Grohmann J, Beyersdorf F. Aneurysms of the pulmonary artery. *Circulation.* 2015; 131:310–316. [PubMed: 25601950]
3. Finch EL, Mitchell RS, Guthaner DF, Fowles RF, Miller DC. Pulmonary artery surgical aneurysmorrhaphy: Where do we go from here? *Am Heart J.* 1983; 106:614–618. [PubMed: 6881048]
4. Inayama Y, Nakatani Y, Kitamura H. Pulmonary artery dissection in patients without underlying pulmonary hypertension. *Histopathology.* 2001; 38:435–442. [PubMed: 11422480]

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.

Figure 2.

(A) 3D VR reconstruction from CTA demonstrating 8.4cm aneurysm of the MPA axial CTA image; (C) Preoperative CXR; (D) Postoperative AP projection 3D VR CTA; (E) Postoperative lateral projection 3D VR CTA reconstruction.

Figure 3.

(A) Right ventricular inflow-outflow view demonstrating a gross enlarged pulmonary artery;
(B) Pulmonary artery aneurysm measuring 8.8cm.

Figure 4.

(A) Intraoperative picture of pulmonary artery aneurysm, (B) Intraoperative photograph demonstrating resected pulmonary artery aneurysm, (C) MPA and LPA reconstruction with Dacron graft, (D) Completed surgical reconstruction, (E) Hematoxylin and eosin stain showing cystic medial degeneration, (F) Elastin – Verhoeff's Van Gieson (EVG) stain showing loss of elastin.