A 62-year-old woman presented with a 4-month history of persistent and progressively worsening left upper chest pain despite analgesia. One month prior, she had noticed a hoarseness of voice. Physical examination at the time revealed cyanosis, clubbing of hands and feet, and pulmonary regurgitation murmur but no right ventricular failure. There was prominent bulging of her rib cage over the left upper precordium. Cardiac catheterization at 12 years of age had confirmed Eisenmenger syndrome (with mean pulmonary arterial pressure of 86 mm Hg) due to an atrial septal defect and significant shunt reversal. In recent years, she had progressive dilation of her pulmonary arterial trunk despite phosphodiesterase inhibitor treatment for 6 years.

A transthoracic echocardiogram revealed an aneurysmal pulmonary arterial trunk with severely elevated right ventricular systolic pressure at 84 mm Hg and right ventricular hypertrophy typical of Eisenmenger syndrome (Figure 1). Computed tomography pulmonary angiogram revealed a giant main pulmonary arterial aneurysm with maximal dimension of 7.2 cm but no radiographic evidence of dissec-

Figure 1. Transthoracic echocardiography shows the giant pulmonary aneurysm with pulmonary hypertension. A. Parasternal short-axis view showing the giant pulmonary artery aneurysm. B through C. Severely dilated and hypertrophied right ventricle in apical 4-chamber view with elevated right ventricular systolic pressure of 84 mm Hg measured on Doppler.
tion, leakage, or rupture (Figure 2A–2C). Polysplenia was also noted consistent with Heterotaxy syndrome.

Laryngoscopy performed to assess her hoarse voice revealed paralysis of the left vocal cord as shown in online-only Data Supplement Movie I, consistent with left-sided recurrent laryngeal nerve palsy. In the light of unrelenting pain with high risk of sudden death from impending aneurysmal rupture or dissection, she proceeded to pulmonary artery surgery.

The aneurysm was repaired with a 24-mm Gore-Tex tube inclusion graft sewn in at the sinotubular junction of the pulmonary artery trunk and the bifurcation of the pulmonary artery distally, using cardiopulmonary bypass and cardioplegic arrest. Postoperatively, she had no hemodynamic difficulties with pulmonary arterial pressure initially managed by nebulized prostacyclin and subsequently returned to sildenafil treatment. Transthoracic echocardiography images and thoracic computed tomography images after surgery had revealed marked reduction in the size of the pulmonary arterial trunk to near-normal caliber (Figure 2D–2F). Online-only Data Supplement Movies II and III, respectively, show the pre- and postoperative 3-dimensional computed tomography reconstruction of the pulmonary artery. At follow-up, there were no cardiac complications; her precordium pain settled and she had successful medialization laryngoplasty of her paralytic vocal cord. Unfortunately, 9 months postoperatively, she succumbed to pneumonia.

**Discussion**

Pulmonary arterial trunk aneurysm has been a very rare entity. Deterling et al found 8 cases of proximal pulmonary aneurysms in 109,571 autopsies. Pulmonary arterial trunk aneurysm is usually an incidental finding but can also present with various symptoms ranging from dyspnea to (rarely) severe chest pain. Hoarseness of voice from left recurrent laryngeal nerve palsy due to mechanical injury by the dilated great vessels from different cardiovascular causes has been
reported as cardiovocal syndrome (Ortner syndrome) for nearly a century. The left recurrent laryngeal nerve descends to a small space between the aortic arch and pulmonary artery, loops around the aortic arch posterior to the ligamentum arteriosum and ascends to supply the larynx. In cardiovocal syndrome, dilatation of the aorta or pulmonary artery compresses the left recurrent laryngeal nerve among the aorta, pulmonary artery, and ligamentum arteriosum. To the best of our knowledge, cardiovocal syndrome resulting from a giant pulmonary arterial trunk aneurysm due to Eisenmenger syndrome has not been reported.

Because of the high risk of impending aneurysmal rupture and increasing chest pain, our patient elected to undergo isolated aneurysmorrhaphy for her giant main pulmonary artery aneurysm despite her potential high perioperative risk. Although surgical correction for main pulmonary artery aneurysms in the setting of other cardiac anomalies has been performed since the 1970s, isolated surgery for pulmonary artery aneurysm due to Eisenmenger syndrome, to our knowledge, has not been previously reported. Rare case reports indicate that pulmonary artery dissection in young patients with Eisenmenger syndrome had been treated successfully with heart-lung transplantation, but this would not serve as an option for our patient. Successful surgical correction had been reported in a pulmonary artery dissection case with probable Eisenmenger syndrome. Our case highlights that in the modern era of advanced anesthesia and sophisticated surgical management, pulmonary artery surgery is an option and should be considered in patients with Eisenmenger syndrome with associated life-threatening pulmonary artery aneurysm.

Disclosures

None.

References

Hoarseness and Chest Pain in Eisenmenger Syndrome With Pulmonary Artery Aneurysm
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