A 36-Year-Old Woman with a History of Dextrocardia and Dyspnea

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Case Report

A 36-year-old woman with a history of dextrocardia presented to the clinic for evaluation of several months of exertional dyspnea. She denied any cough, wheezing, fever, chills or rigors, as well as any orthopnea, leg edema or hemoptysis. Past medical history was significant for tachycardia, orthostatic hypotension, dextrocardia diagnosed 15 years ago, hyperlipidemia and chronic headaches. She had a history of endometrial ablation in May 2007. She had had an uneventful pregnancy 10 years ago. Current medications included nadolol 20 mg twice a day. Family history was not significant for any cardiac or pulmonary disease. She had worked at a childcare center in the past but was now a student. She denied any smoking history and drank 1–2 glasses of wine per week. Review of systems was positive for postnasal drip, nasal congestion and headaches. On examination, vitals were normal. Chest exam showed decreased breath sounds on the right side. There was limited movement of the diaphragm on the right side with respiration. Heart sounds were shifted to the right side. There was no clubbing, cyanosis or edema. A 2D echocardiogram done 15 years ago was interpreted as dextrocardia with normal valves and normal cardiac function. A chest radiograph (fig. 1) done at an outside hospital was read as volume loss on the right side with dextrocardia. Pulmonary function tests including spirometry, lung volumes, and diffusing capacity were within normal limits.

What is your diagnosis?

Fig. 1. Chest radiograph showing right lung volume loss with mediastinal shift.

Diagnosis: Unilateral Absence of a Pulmonary Artery (UAPA) with Right Hypoplastic Lung

Fig. 2. Contrast-enhanced chest CT of the patient showing the mediastinal vessels.

Fig. 3. Chest CT showing hypoplastic right lung.

A contrast-enhanced CT of the chest (fig. 2–5) followed by a repeat 2D echocardiogram confirmed the absence of a right pulmonary artery along with a right hypoplastic lung. She did not have dextrocardia but had dextroversion due to volume loss in the right chest which mimicked dextrocardia.

Discussion

UAPA is a rare congenital disorder frequently associated with cardiac abnormalities. This entity was first reported in 1868 [1]. Isolated UAPA (without cardiovascular congenital abnormalities) constitutes about 30–50% of the cases [2]. Bouros et al. [3] found the prevalence of isolated UAPA to be 1 in 200,000 persons. There is no predilection for either sex. The median age of these patients was 14 years (range 0.1–58 years) with 12% of them being infants [2]. Absence of a right pulmonary artery as seen in the present patient is more common than absence of the left pulmonary artery [2]. Most patients are symptomatic and commonly present with recurrent pulmo-

nary infections, decreased exercise tolerance, hemoptysis or mild exertional dyspnea [4]. About a third of these patients present with recurrent pulmonary infections. Though less common, fatal cases of necrotizing pneumonia can occur and recurrent infections can also lead to bronchiectasis [3, 5, 6]. Bouros et al. [3] suggested that alveolar hypocapnia can cause bronchoconstriction, while impaired mucociliary clearance and the diminished delivery of appropriate inflammatory cells may contribute to the high incidence of infections in patients with UAPA [3]. Hemoptysis is the presenting symptom in 20% of the patients [2, 7]. This is due to development of excessive collateral circulation in bronchial, intercostal, subclavian, or subdiaphragmatic arteries [8]. Hemoptysis can be self-limiting for many years, but can lead to massive hemorrhage and death [9]. Pulmonary arterial hypertension (PAH) has been reported in 20–25% of patients with UAPA [1, 4]. This is usually unmasked by predisposing factors like high-altitude pulmonary edema (HAPE) and pregnancy [10–14]. The etiology of PAH in these patients is still unclear.

Fig. 4. 3D reconstruction of the chest CT of the patient showing absence of a right pulmonary artery.

Fig. 5. 3D reconstruction of the chest CT of a normal person compared to that of the patient.

Although a chest radiograph usually suggests the diagnosis of UAPA, a contrast-enhanced CT scan or a 2D echocardiogram is needed to confirm the diagnosis. The chest radiograph may show the absence of a hilar shadow, a shrunk affected lung, and a shift of the mediastinal structures to the affected side. In addition, the absence of the left or right pulmonary artery, ipsilateral grossly diminished pulmonary vascular markings, a small hemithorax and intercostal bone space, ipsilateral cardiac and mediastinal displacement, ipsilateral hemidiaphragm elevation, and contralateral lung hyperinflation may also be seen [8]. The echocardiogram also helps exclude other associated cardiac abnormalities and PAH. The most common congenital cardiac malformations associated with UAPA are coarctation of the aorta, either isolated or in combination with a ventricular septal defect; subvalvular aortic stenosis; transposition of the great arteries, either isolated or in combination with ventricular septal defect or pulmonary stenosis; Taussig-Bing malformation and coarctation; congenitally corrected transposition and pulmonary stenosis, and scimitar syndrome [2].

Common causes of mortality (7%) in isolated UAPA are from massive pulmonary hemorrhage, right heart failure, respiratory failure, PAH or HAPE [2, 9, 15]. PAH

may have devastating effects, especially when it develops during pregnancy. PAH can be improved by revascularization of the hidden pulmonary artery, although this has only been described in younger patients [16]. When revascularization is proposed, cardiac catheterization is necessary, including pulmonary venous wedge angiography to discover hilar arteries. If revascularization is contraindicated, patients with PAH may benefit from vasodilator treatment used to treat primary PAH. Pneumnectomy or lobectomy is needed for massive hemoptysis, necrotizing bronchopneumonia and bronchiectasis [2].

The present patient had exertional dyspnea without any hemoptysis or evidence of lower respiratory tract infections. Further evaluation included a normal 2D echocardiogram which was followed by a right- and left-heart catheterization. This showed pulmonary artery pressures of 16/2 (mean 8) mm Hg, left-ventricular ejection fraction of 60% and normal coronaries. A cardiopulmonary exercise test at our facility showed ventilatory limitation. She had a normal heart rate reserve but was limited by dyspnea, which occurred at a reduced maximum VO_2 of 1.6 l/min (85% predicted), maximum work of 120 W (78% predicted) and maximum heart rate of 140 (80% predicted). However, her ventilation at maximum exercise was 90% of the calculated maximum voluntary ventilation.

The likely explanation of this ventilatory limitation is a paradoxical worsening of the dead space ventilation with exercise. This has been reported previously to be a cause of dyspnea in patients with isolated UAPA with normal pulmonary artery pressures [17]. Currently, her symptoms have improved with pulmonary rehabilitation.

Key Words

Absence of pulmonary artery • Hypoplastic lung • Dextroversion

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