Isolated Unilateral Absence of a Pulmonary Artery — A Case Report

Yu-Ching Lin, Ming-Ren Chen, Chang-Hsien Yu and Albert Chen

Unilateral absence of a pulmonary artery (UAPA) is a rare pulmonary vascular anomaly that is usually associated with other cardiac anomalies. We report a 9-year-old female diagnosed with isolated unilateral absence of the left pulmonary artery and a right aortic arch but without other cardiac anomalies. The only symptoms the patient had were recurrent respiratory tract infections. The chest x-rays showed asymmetric lung fields, prompting subsequent studies. A series of imaging studies including chest x-rays, esophagography, echocardiography, chest computed tomography, lung ventilation-perfusion scintigraphy, angiocardiography, and 3-dimensional magnetic resonance angiography (3D-MRA) were done. The diagnosis was confirmed by the cardiac catheterization and pulmonary angiography, which disclosed absence of the left pulmonary artery. 3D-MRA conclusively demonstrated total absence of the left pulmonary artery without an occult left pulmonary artery or systemic-to-pulmonary collateral arteries. Therefore, surgical re-establishment of pulmonary continuity was impossible.

Key Words: Isolated • Unilateral absence of a pulmonary artery • Right aortic arch • Chest computed tomography • Ventilation-perfusion scintigraphy • Three-dimensional magnetic resonance angiography

INTRODUCTION

Unilateral absence of a pulmonary artery (UAPA) is an uncommon pulmonary vascular anomaly usually associated with other cardiac anomalies, such as tetralogy of Fallot, ventricular septal defect and patent ductus arteriosus.1,2 Isolated UAPA without other cardiac anomalies is quite rare. Patient with isolated UAPA usually have only minor symptoms or are asymptomatic. In 2002, Drs. Ten Harkel et al. reviewed the literature of UAPA from 1978 onward from the database of Pub-Med (National Library of USA).3 In Taiwan, isolated UAPA was reported by Wang et al. in 19974 and Lin YM et al. in 1999.5 Several investigators have found occult pulmonary artery branches extending into the parenchyma in some cases. The affected lung is also sometimes supplied by systemic-to-pulmonary collateral vessels. If an occult pulmonary artery can be located, circulation to the affected lung may be re-established surgically, improving the prognosis.

CASE REPORT

A 9-year-old female patient reported frequent upper airway infections for 1 year. She presented to our emergency room complaining of fever and cough for 4 days. Auscultation of the chest revealed coarse breath sounds but no rales or wheezing. She had a continuous grade 1/6 murmur at the right upper sternal border. A chest x-ray showed asymmetric lung fields with decreased volume and vascularity on the left, mild deviation of the trachea and mediastinum to the left, hyperinflation of the right lung, and a right perihilar infiltrate (Figure 1). Because of the unusual chest x-ray and history of frequent infections,
we suspected a vascular ring or other cardiac abnormalities. The electrocardiogram showed an incomplete right bundle branch block. There was no indentation on an esophagogram, essentially ruling out a vascular ring. On computed tomography (CT) of the chest, there was a right aortic arch with only a right pulmonary artery connected to the main pulmonary artery. No abnormal masses were seen. Echocardiography with Doppler studies showed absence of the left pulmonary artery, a dilated right ventricle, a dilated right pulmonary artery with possible distal stenosis, and a right aortic arch without coarctation.

On ventilation scanning, there was homogeneous ventilation of both lungs but an apparent decrease in the left lung volume. A perfusion scan showed normal perfusion of the right lung but nearly absent perfusion in the left. The findings were consistent with left UAPA and a hypoplastic left lung. On right heart catheterization, the right ventricular pressure was elevated at 62/9 mmHg (on irritable status), the main pulmonary artery pressure was 43/11 mmHg with a mean of 23 mmHg and right pulmonary artery pressure was 38/11 mmHg with a mean of 21 mm Hg. On the pullback tracing from the right to the main pulmonary artery and then to the right ventricle, there was no apparent pressure gradient. A pulmonary angiogram (Figure 2) showed only a right pulmonary artery connecting with the main pulmonary artery. The left pulmonary artery was completely absent. There was also distal right pulmonary artery stenosis with post-stenotic dilatation. The aortogram showed a right aortic arch without any apparent systemic-to-pulmonary collateral circulation to the left lung.

The patient was treated symptomatically for her respiratory symptoms, which subsided. She denied syncope, exertional dyspnea, hemoptysis, cyanosis, or chest pain. She was discharged and regularly followed in our clinic. One year later, 3-dimensional cardiac magnetic resonance angiography (MRA) in axial and coronal projections (Figures 3A and B) was performed in order to search for an occult pulmonary artery or systemic-to-pulmonary collateral circulation. It confirmed the presence of a right aortic arch, absent left pulmonary artery.

![Figure 1. Chest roentgenogram showing a small left hemithorax, hyperinflation of the right lung, deviation of the trachea and mediastinum to the left, and an engorged vascular shadow in the right hilum. The volume and vascularity of the left lung appear to be diminished.](image)

![Figure 2. Pulmonary angiography (left, PA view; right, lateral view) showing absence of the left pulmonary artery, a dilated main pulmonary artery, and a dilated right pulmonary artery with distal stenosis (arrows). Post-stenotic dilatation can be identified. Vascularity in the left lung is absent.](image)
artery, and marked dilatation of the right pulmonary artery with narrowing near the distal bifurcation. There were 2 superior and 2 inferior pulmonary veins bilaterally without evidence of aberrancy, although the venous return in the enlarged right superior pulmonary vein was much greater than normal. The predominant perfusion was to the right upper and middle lobes, with decreased volume in the left lung and the right lower lobe. As there was no occult left pulmonary artery or hilar artery, revascularization of the left lung was not possible.

**DISCUSSION**

Unilateral absence of a pulmonary artery (UAPA) is an uncommon congenital anomaly that occurs on the right more often than the left. About 80% of reported cases involving the left pulmonary artery have been associated with other congenital cardiovascular anomalies, such as tetralogy of Fallot, right aortic arch, ventricular septal defect, or patent ductus arteriosus. Typically, the absent artery is on the side opposite to the aortic arch, as was true in our case.

The prognosis depends on associated cardiovascular anomalies and the degree of pulmonary hypertension. The latter develops because of high flow through the remaining pulmonary artery and consequent remodeling of the pulmonary vascular bed. Chronic hypoxia caused by ventilation-perfusion mismatch, as well as chronic thrombosis or embolism secondary to polycythemia, may also contribute to the elevated pulmonary artery pressure. Coexisting contralateral peripheral pulmonary artery stenosis may exaggerate the degree of pulmonary hypertension and is a poor prognostic factor. Congestive heart failure may be the end result.

Patients with isolated UAPA are usually asymptomatic, which may delay the diagnosis. The patient may have an abnormal chest x-ray, recurrent respiratory infections, chronic cough, dyspnea, or, rarely, hemoptysis. In our case, the diagnosis was initially suspected because of an abnormal x-ray performed because of the patient recurrent respiratory infections. The slight leftward deviation of the trachea was due to hypoplasia of the left lung and overinflation of the right lung. The continuous murmur heard at the right sternal border was likely due to peripheral right pulmonary artery stenosis.

When there is normal or only mildly decreased ventilation to a lung with absent or markedly decreased perfusion seen on ventilation-perfusion scanning, the diagnosis is limited to pulmonary vascular problems, including severe pulmonary artery stenosis, peripheral pulmonary artery atresia, or acquired pulmonary artery occlusion. The only exception to this principle is when a large systemic collateral circulation supplies the affected lung, in which case an apparently normal ventilation-perfusion scan may be misleading. The ventilation-

![Figure 3. (A) MRA with maximal intensity projection in the coronal view shows marked dilatation of the right pulmonary artery with narrowing near the distal bifurcation and complete absence of the left pulmonary artery. There is no occult left pulmonary artery. No significant systemic-to-left lung collateral circulation can be identified. The vascularity of the right lung is markedly increased compared with the left lung. (B) MRA in the axial view showing absence of the left pulmonary artery and a dilated right pulmonary artery (arrow) connected to the main pulmonary artery (M).](image-url)
perfusion scan helped us focus the search on anomalies of the pulmonary vasculature even before the diagnosis was well established.

Several investigators have suggested that UAPA is caused by proximal interruption of the affected pulmonary artery,\textsuperscript{10,11} since occult pulmonary artery branches are often present. Pulmonary angiography is necessary for the diagnosis of UAPA, but it is not adequate for identifying an occult pulmonary artery. Chest CT and MRA are both useful noninvasive tools to look for such occult branches, especially when surgical re-establishment of pulmonary circulation is under consideration.\textsuperscript{12} Both imaging techniques allow congenital UAPA to be distinguished from acquired obstruction of the pulmonary artery. In addition to depicting static morphologic structures, MRA with contrast also provides real-time assessment of the hemodynamic status. In our opinion, it is superior to chest CT because it combines the benefits of MRI with partial catheterization angiography.

In our patient, the left pulmonary artery was completely absent. We could not identify an occult pulmonary artery or any collateral circulation. It is possible that there were tiny vessels too small to be identified by either angiocardiography or MRA. Other reports suggest that left pulmonary vein wedge angiography is a safe and useful imaging study for demonstrating an occult pulmonary artery and systemic collateral arteries. However, we believe a noninvasive study such as MRA is much safer. The drawback of MRA is that it requires expensive technology and is more time-consuming.

Treatment of isolated UAPA should be based on the symptoms and the pulmonary hemodynamic status. Asymptomatic patients should be followed without intervention unless lower respiratory infections intervene, requiring prompt treatment. Since a lung with an absent pulmonary artery is not well exposed to desaturated blood coming from the inferior vena cava that is rich in hepatic factors,\textsuperscript{13-15} it is at risk for development of arteriovenous malformation or fistula. In patients presenting with hemoptysis, demonstration of collateral circulation is vital so that appropriate treatment such as embolization of an arteriovenous malformation or ligation of systemic collateral vessel can be undertaken.\textsuperscript{16,17} In patients with intractable hemoptysis or necrotizing pneumonia, pneumonectomy may be necessary.\textsuperscript{18,19} Surgical anastomosis should be considered in cases of pulmonary hypertension with or without congestive heart failure if an occult pulmonary artery or adequate intrapulmonary arteries are present. Since our patient did not have an occult left pulmonary artery, reconstruction of normal left pulmonary circulation was not possible. Therefore, the only management at this point is conservative observation. Although currently asymptomatic, the patient’s long-term prognosis is poor.

REFERENCES

單獨的單側左肺動脈缺損 — 一病例報告

林育慶 陳銘仁 游昌憲 陳律霖
台北市 馬偕紀念醫院 小兒科

單側肺動脈缺損是一個罕見的肺血管異常，它通常合併其他心臟結構異常。我們報告一病例為九歲女姓診斷為單獨的左側肺動脈缺損併右主動脈弓，此外這病例並無合併其他心臟結構異常。一系列影像檢查包括：胸部X光、食道X光攝影、心臟超音波、胸部斷層掃描、通氣及灌流掃描、心導管血管攝影、三度空間核磁共振血管攝影。由一張左右肺不對稱的胸部X光，開啟一連串後續檢查。心導管及肺動脈血管攝影診斷為左側肺動脈缺損，核磁共振血管攝影是一有效的非侵襲性檢查，它確定地證明左肺動脈完全缺損且無隱藏的肺動脈。因此手術建立肺部血流連續是不可能的。

關鍵詞：單獨、單側肺動脈缺損、右動脈弓、胸部電腦斷層掃描、通氣及灌流造影術、三度空間核磁共振血管攝影。