Radiological Features of Isolated Unilateral Absence of the Pulmonary Artery

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ABSTRACT

We report a case of isolated unilateral absence of the pulmonary artery, a rare congenital anomaly, in a woman with exertional dyspnoea. The chest radiograph appearances of this anomaly are often mistaken for other conditions, such as Swyer-James-MacLeod’s syndrome or thromboembolic disease. The interpretation of chest radiograph findings in order to differentiate isolated unilateral absence of the pulmonary artery from other conditions is discussed, along with the appropriate use of ancillary imaging techniques.

Key Words: Bronchiectasis, Computed tomography, thorax, Isolated unilateral absence of pulmonary artery, Magnetic resonance angiography

INTRODUCTION

Isolated unilateral absence of the pulmonary artery (IUAPA) is a rare congenital anomaly, which is usually discovered incidentally on an adult chest radiograph. Most patients have minimal symptoms, and are often misdiagnosed with other conditions, such as Swyer-James-MacLeod’s syndrome (SJMS) or pulmonary thromboembolic disease. Misdiagnosis arises due to the confusing chest radiograph appearances of IUAPA, in which the affected lung is hypoluent and contracted, while the contralateral lung is hyperlucent. The latter is thus often erroneously regarded as the abnormal lung. The current case illustrates the process of reaching the correct diagnosis, with the aid of ancillary modalities such as magnetic resonance (MR) angiography and computed tomography (CT) of the chest. This is the first reported case of a patient with IUAPA and ipsilateral bronchiectasis.

CASE REPORT

A 35-year-old Chinese housewife, a non-smoker, presented to Queen Mary Hospital with increasing exertional dyspnoea. The patient had a 5-year history of rheumatoid arthritis, which had been satisfactorily controlled using nonsteroidal anti-inflammatory agents alone, without major pulmonary complications. The patient reported an initially gradual onset of exertional dyspnoea, which had progressed rapidly in the preceding 2 months. Associated symptoms included a mild, central, dull aching chest pain, with cough but no haemoptysis.

Clinical examination of the cardiovascular, respiratory and abdominal systems revealed no abnormalities. Electrocardiographic assessment demonstrated a normal sinus rhythm pattern. Routine biochemical and arterial blood gas analyses were unremarkable. Fibreoptic bronchoscopy indicated no endobronchial lesions. Bronchoalveolar lavage and post-bronchoscopy sputum analysis yielded no respiratory pathogens (including mycobacterium), or malignancy. Chest radiograph, however, revealed a contracted right hemithorax, with reduced lung volume and diminished lung markings, a small right hilum, elevated right hemidiaphragm, right mediastinal shift, and a hyperlucent left lung (Figure 1).

To exclude the diagnosis of a pulmonary thromboembolic event, the patient underwent further evaluation with contrast-enhanced CT scanning (General Electric (GE) LightSpeed MultiSlice CT Scanner, Milwaukee, Wisconsin, USA) of the thorax, with dynamic bolus
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injection of contrast (150 ml Omnipaque 240 mg/mL, 4 mL/s). The right hemithorax was noted to be smaller and hypolucnt compared with the left. The right pulmonary artery was entirely absent and the right pulmonary vasculature was considerably smaller in calibre compared with the contralateral side (Figures 2 and 3). No emboli were detected in the central segmental, and subsegmental pulmonary arteries in either lung. There was also no obvious cardiac anomaly. Of interest, there was generalised cylindrical bronchiectasis, with bronchial wall thickening in all lobes of the right lung, which was confirmed with a high resolution CT (HRCT) study (Figure 3).

As the left lung was hypolucnt, an expiratory HRCT scan was obtained to exclude air trapping and therefore the diagnosis of SJMS. Radionuclide perfusion ventilation scanning demonstrated gross ventilation/perfusion mismatch, with almost complete absence of perfusion of the right lung (Figure 4). A contrast-enhanced MR angiogram was performed with a 1.5 tesla system (GE Signa, Milwaukee, Wisconsin, USA), using a bolus injection of 40 ml of gadolinium-DTPA. Volume rendering software (GE Advantage...
Windows, 3D Analysis Package, Milwaukee, Wisconsin, USA) was used to generate a 3-dimensional model of the heart and great vessels. Disarticulation of the ascending aorta confirmed the absence of the right pulmonary artery. Multiple collateral systemic supplies were demonstrated in the right hemithorax, including hypertrophic right superior intercostal, right bronchial artery and right internal mammary arteries (arrows) providing collateral circulation to the right pulmonary vasculature.

Figure 5. Magnetic resonance angiogram of the thorax illustrates the absence of the right pulmonary artery, and the hypertrophied right bronchial, superior intercostal and internal mammary arteries (arrows) providing collateral circulation to the right pulmonary vasculature.

with right pulmonary artery agenesis and a normally sited left aortic arch usually survive into adulthood with minimal symptoms. Therefore, diagnosis in such cases is exceptionally difficult, as other conditions including SJMS, compensatory emphysema, and pulmonary thromboembolic disease may mimic the radiographic appearance of IUAPA.

The typical radiographic appearances of IUAPA are an ipsilateral contracted lung with ipsilateral mediastinal and tracheal shift, elevation of the ipsilateral hemidiaphragm, diminished ipsilateral vascular markings, absent air trapping on expiratory film, and hyperinflation of the contralateral lung. One third of patients are asymptomatic, while approximately 50% experience recurrent chest infections. Haemoptysis is thought to be caused by the rupture of hypertrophic collaterals arising from large systemic-pulmonary vascular communications involving the bronchial, intercostal and internal mammary arterial collaterals, and occurs in up to 10% of cases. Pulmonary hypertension develops in only a small proportion of patients.

Bronchiectasis has not previously been reported in association with this condition. The presence of bronchiectasis in this patient is interesting given that the bronchial tree in the contralateral lung was entirely normal. We hypothesise that this reflects the impact of IUAPA on the mucosal defence of the lung. The patient is rendered prone to recurrent infections by hypoperfusion and V/Q mismatch secondary to the indirect collateral supply, and hence bronchiectasis develops as a result. Ipsilateral emphysema has been reported with IUAPA, although the underlying pathogenesis is not fully understood. Other potential explanatory hypotheses for bronchial complications, including the effects of protease-antiprotease imbalance, and relative arterial hypoperfusion, remain subject to debate.

The differential diagnosis for conditions associated with radiological findings of unilateral hyperlucent lung includes SJMS, compensatory or obstructive emphysema, and pulmonary embolism. The key factor in arriving at the correct diagnosis is to decide which of the lungs is abnormal. In these other conditions, the hyperlucent lung is the site of abnormality. In IUAPA, the reverse is true, as compensatory hyperinflation in the contralateral normal lung gives one the false impression of unilateral hyperlucency. The hyperlucent lung in SJMS also demonstrates air trapping on expiratory scans or chest radiographs, a unique

DISCUSSION

Congenital unilateral absence of the pulmonary artery (UAPA) is usually associated with other cardiac anomalies and thus, diagnosed in early childhood. Presentation in later life occurs in patients without cardiac anomalies, and is therefore aptly termed IUAPA. This disorder is rare and the incidence is estimated to be less than 0.3% in the general population.

Left pulmonary artery agenesis is frequently associated with several life-threatening congenital cardiac abnormalities such as Tetralogy of Fallot. Early diagnosis and surgical intervention are required for this condition in the childhood period. In contradistinction, patients
feature of SJMS which is helpful in the differential diagnosis.

Although angiography remains the traditional gold standard for diagnosing IUAPA, it is rarely performed now unless embolisation is suggested by massive haemoptysis. While both CT and MRI can accurately depict the absent pulmonary artery, MR angiographic techniques can clearly delineate the collateral systemic vascular supply non-invasively. MR angiography has been shown to have comparable accuracy to conventional and modern digital subtraction angiography. 3-dimensional MR angiography in the current case not only provided the definitive diagnosis, but also demonstrated the hypertrophied right lower intercostal and internal mammary arteries, and the normal patent left pulmonary artery. V/Q scanning (Figure 4) is useful in distinguishing SJMS from IUAPA. However, in the context of a suspected pulmonary thromboembolic event, correct image interpretation may be difficult without additional clinical history and other imaging findings. A further limitation of V/Q scanning is its inability to demonstrate pulmonary vascular anatomy and collateral arterial supply.

CONCLUSION
The diagnosis of IUAPA should be considered when an adult chest radiograph shows a unilateral hypolucuent contracted lung, with small or absent ipsilateral hilum, and a contralateral hyperlucent lung. Confirmation of the diagnosis can then be achieved with subsequent CT or MR imaging.

REFERENCES