

## ***CASE FOR DIAGNOSIS***

# **A patient with haemoptysis and a smaller right lung**

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### **Case history**

A 41-year-old businessman was admitted to National Taiwan University Hospital for evaluation of a 2-week history of haemoptysis productive of 50–100 mL of fresh blood daily. The patient stated that haemoptysis had first occurred 10 yrs earlier, had lasted for about 1 month and had spontaneously subsided. Both episodes of haemoptysis were preceded by flu-like symptoms. There was no past history of epistaxis, exertional dyspnoea, or loss of body weight.

Physical examination revealed a well-developed man in no distress. His blood pressure was 112/70 mmHg, pulse 96 beats·min<sup>-1</sup> and respiratory rate 22 breaths·min<sup>-1</sup>. There was a slightly decreased chest wall excursion, and breath sounds in the right hemithorax. No audible bruits were heard over the chest. Heart sounds were regular, without murmurs. No clubbing, cyanosis, telangiectasia or oedema were noted. Arterial blood gases in room air showed: a pH of 7.41; arterial carbon dioxide tension (PaCO<sub>2</sub>) of 4.9 kPa (36.5 mmHg); and arterial oxygen tension (PaO<sub>2</sub>) of 13.1 kPa (98.3 mmHg). The sputum was negative for acid-fast bacilli, fungi and malignant cells. An electrocardiogram was normal. Echocardiography showed normal cardiac size and good left ventricular contractility, with no evidence of congenital cardiac lesions or pulmonary hypertension. On bronchoscopy, no endobronchial lesion or definite site of bleeding was found.

Imaging studies, including a posteroanterior (PA) chest radiograph (fig. 1), and computed tomography (CT) scan at the level of the aorticopulmonary window (fig. 2) are shown.

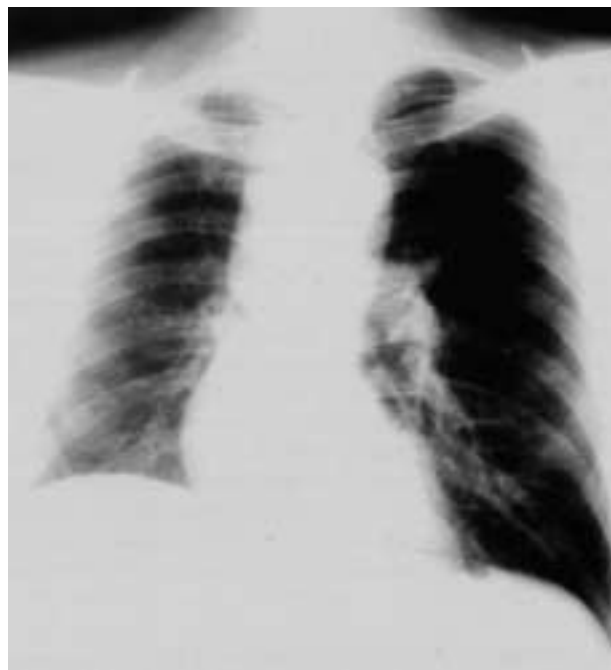


Fig. 1. – Posteroanterior chest radiograph.

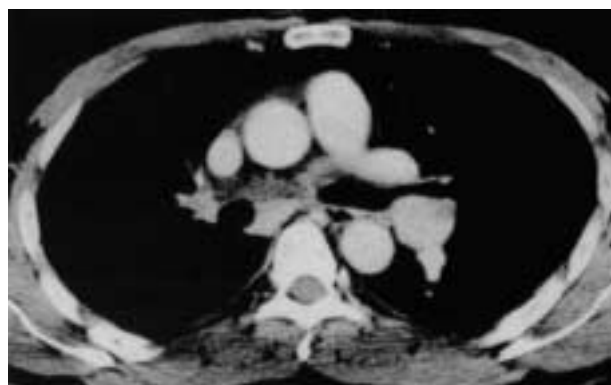


Fig. 2. – Computed tomography (CT) scan at the level of the aorticopulmonary window.

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**BEFORE TURNING THE PAGE:  
INTERPRET THE ROENTGENOGRAM AND THE CT SCAN AND  
SUGGEST THE DIAGNOSIS.**

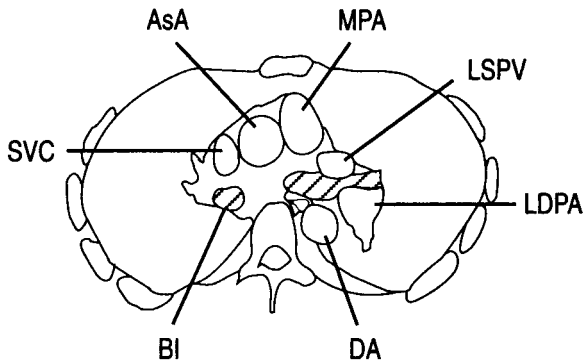


Fig. 3. – Diagrammatic illustration of figure 2. SVC: superior vena cava; AsA: ascending aorta; DA: descending aorta; MPA: main trunk of pulmonary artery; LSPV: left superior pulmonary vein; LDPA: left descending pulmonary artery; BI: bronchus intermedius.

### Interpretation of the chest radiograph and CT scan

The PA chest radiograph reveals loss of volume of the right lung and ipsilateral shift of the heart and mediastinum. Right hilar vascular shadows are not definitely seen. The left lung is relatively hyperinflated and the left pulmonary artery markedly dilated.

The CT scan at the level of the aorticopulmonic window (fig. 2) shows absence of the right branch of the pulmonary artery, which should normally run immediately behind the ascending aorta and superior vena cava, and anterior to the bronchus intermedius (see the illustrative diagram in figure 3). Note the diameter of left descending pulmonary artery (LDPA), which is larger than that of the adjacent descending aorta (DA). The left superior pulmonary vein (LSPV) is also enlarged.

### DIAGNOSIS: “Unilateral absence of the pulmonary artery (UAPA)”

Pulmonary angiography confirmed the absence of the right branch of the pulmonary artery (fig. 4a). A thoracic aortogram (fig. 4b) revealed prominent and engorged

right bronchial arteries (arrows), with some aneurysmal dilatations (arrowhead). No other abnormalities of the heart chambers or great vessels were noted.

### Discussion

Unilateral absence of the pulmonary artery (UAPA) is an unusual cause of haemoptysis. It may be seen as an isolated condition or in association with other congenital heart defects. Absence of the left pulmonary artery is more commonly seen in patients with tetralogy of Fallot or truncus arteriosus, whilst absence of the right pulmonary artery is more often an isolated finding [1]. The embryological basis for UAPA is malformation of the sixth aortic arch on the affected side [1, 2]. These patients have a normal pulmonary trunk but unilateral absence of the branch pulmonary artery. Some authors prefer to use the term “proximal interruption of a pulmonary artery” [3, 4], since the more distal parenchymal pulmonary vessels are generally intact and patent, though diminished in calibre [4].

The involved lung is usually smaller than the opposite lung and is supplied by bronchial collateral vessels that can be derived from any of the systemic arteries in the chest [1, 5]. Most of the collateral vessels hypertrophy following birth, and produce numerous vascular channels in the submucosa of the bronchial walls [4]. Bronchial blood flow to the affected lung may be increased 15–25 fold and can account for up to one third of the left ventricular output [1, 6]. The incidence of pulmonary hypertension in the isolated cases is 19% [1].

The clinical presentation of these patients is variable: about 30% of patients remain asymptomatic throughout their lives [2]. There may be mild dyspnoea on exertion [5], and up to 40% of patients suffer from recurrent pulmonary infections [7]. In their review of 98 patients, POOL *et al.* [1] found haemoptysis in 10%, but others have documented an increased frequency of haemoptysis as the presenting symptom, especially in adults [7]. Haemoptysis is most often inconsequential and results from hypertrophied collateral vessels [1, 3, 7]. Venules

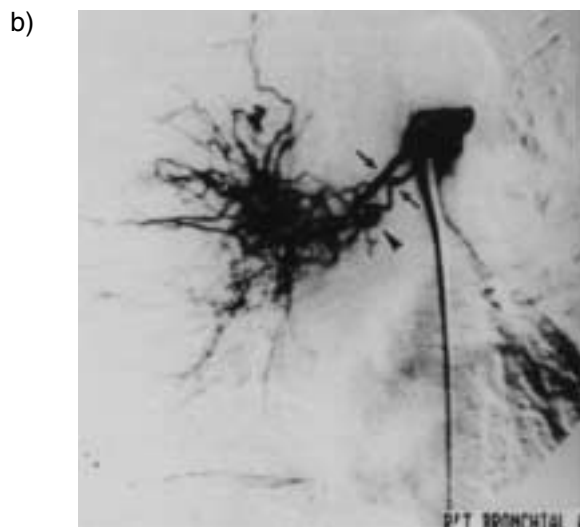
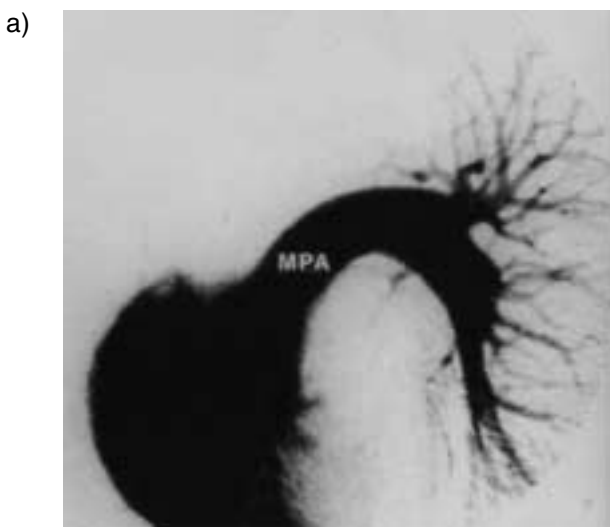


Fig. 4. – a) Pulmonary angiography showing the absence of the right branch of the pulmonary artery. b) Thoracic aortogram revealing the prominent right bronchial arteries (arrows), with some aneurysmal dilatations (arrowhead). MPA: main trunk of pulmonary artery; R'T BRONCHIAL A: right bronchial artery.

ruptured because of the higher circulation pressures in the affected lung can cause haemoptysis of a potentially lethal magnitude [8, 9]. Bronchiectasis has been reported in some patients, but its pathogenesis and association with recurrent infections remain unclear [5]. No evidence of bronchiectasis was found on chest CT of the case presented here.

Physical findings can include an asymmetrical chest with decreased breath sounds in the affected side, mediastinal shift and an occasional systolic murmur across the pulmonary outflow tract because of turbulent blood flow into the remaining pulmonary artery [1]. Cardiac examination and the electrocardiogram are usually within normal ranges when there is no associated congenital heart disease and pulmonary hypertension. Some patients may have digital clubbing.

The chest radiograph can be very valuable, typically revealing an ipsilateral small, contracted lung, and a contralateral enlarged, hyperinflated, well-vascularized lung that frequently herniates across the midline [8]. There is usually ipsilateral tracheal deviation, diaphragmatic elevation, narrowed intercostal spaces, and small hilum. A reticular pattern from the collateral circulation to the involved lung can, at times, be seen on plain chest film [4, 8]. Scintigraphic studies will demonstrate complete absence of perfusion to one whole lung, with diminished ventilation but no delay in the wash-out phase of the ventilation scan [5]. Bronchoscopy and bronchograms generally reveal normal bronchial structure, although it is not unusual to find crowding or irregular constrictions on the affected side [1]. Routine pulmonary function studies are generally within the normal range [1], but a mild restrictive pattern may be found [5]. Conventional CT with contrast should be performed to assess the pulmonary artery. High-resolution CT has also been recommended to assess the presence of bronchiectasis [5]. Digital subtraction angiography (DSA) and pulmonary angiography are usually definitive in establishing the diagnosis. DSA is probably the procedure of choice, being easier to perform and safer than the latter [5]. Magnetic resonance imaging (MRI) has recently been used for accurate anatomical depiction of UAPA. It may be the preferable noninvasive technique for defining the anatomy of the main and branch pulmonary artery and also helpful in the identification of accompanying congenital abnormalities.

Diagnosis of UAPA is based on a detailed history and physical examination, coupled with a reasonable index of suspicion. Other conditions mimicking UAPA on plain chest radiograph are: collapse of a lobe or of one lung, chronic tuberculous scarring, unilateral emphysema, hypoplasia of one lung, Swyer-James syndrome or hyperlucent lung, and complete obstruction of a main pulmonary artery secondary to thromboembolism, neoplasm [10], or fibrosing mediastinitis [11].

Therapeutic approach should be based on symptomatology and severity of disability. Attempts have been

made to revascularize the affected lung in infants with isolated UAPA [10]. In older patients, revascularization is not encouraged or even feasible because the intrapulmonary arteries have been found to be severely narrowed or even completely obstructed by fibrosis [8]. Embolization and ligation of the numerous collateral vessels have been attempted unsuccessfully to control bleeding [8]. In view of the fact that the affected lung in UAPA does not contribute to ventilation, it has been recommended that if the patient is fit, pneumonectomy should be performed when massive haemoptysis occurs [9]. Aggressive treatment is indicated when haemoptysis, haematemesis, recurrent infection or pulmonary hypertension become intractable. Since this patient's haemoptysis was infrequent and of minor severity, no invasive therapeutic intervention was performed.

**Keywords:** Haemoptysis, pulmonary artery, unilateral absence

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