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CASE REPORT

Interruption of Pulmonary Artery

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ABSTRACT

Congenital interruption of the pulmonary artery is a rare condition. We present a case of isolated interruption of the right pulmonary artery in a young girl and a case of interruption of the left pulmonary artery associated with Tetralogy of Fallot in an adult male. The radiological and clinical features of this condition are also discussed.

Key Words: *Congenital abnormalities; Pulmonary artery; Radiography, thoracic*

中文摘要

肺動脈中斷

郭啟欣、羅尚銘、曾子勤、范子和、關鼎樂

先天性肺動脈中斷的病例很罕見。本文分別報告兩個有關病例，其中一名年幼女童的右肺動脈出現單純中斷，另一名成年男子為法洛氏四合症合併左肺動脈中斷。本文並就這種病的放射學及臨床特徵作出了討論。

INTRODUCTION

Interruption of the pulmonary artery is a rare congenital disorder. The clinical symptoms are non-specific and radiological investigations are the key to diagnosis. We present two cases of interruption of a pulmonary artery. The first was a baby girl with an isolated interruption of right pulmonary artery, who presented with recurrent chest infections. The second was an adult male with known uncorrected Tetralogy of Fallot, who presented with hemoptysis and was found to have an interrupted left pulmonary artery. In addition, the radiological and clinical features of having an interrupted pulmonary artery are reviewed.

CASE REPORTS

Case 1

A 19-month-old girl was investigated for recurrent chest infections. She had experienced 3 episodes of pneumonia since her full-term normal spontaneous delivery in 2007.

Review of her previous chest radiographs (Figure 1) showed the volume in the right lung was persistently small and there was over-inflation of the left lung, which herniated to the contralateral side. The right hilum was small and indistinct while the left hilar vessels were prominent. Increased opacity was consistently noted in the right para-tracheal region and right upper lobe collapse was initially suspected by clinicians.

The patient was therefore admitted for computed tomography (CT) of the thorax with injection of intravenous contrast. CT showed volume loss of the right lung. Images in the soft tissue window (Figure 2) showed ab-

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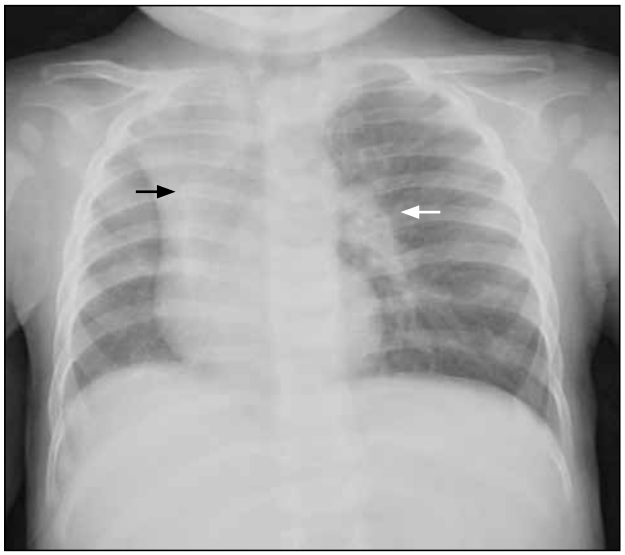

Figure 1. Plain chest radiograph of the first patient. There is volume loss of the right lung with herniation of the left lung to the contra-lateral side (black arrow). The left hilar vessels are prominent (white arrow) while the right hilum is indistinct.

sence of the mediastinal portion of the right pulmonary artery. The inter-lobar portion of the right pulmonary artery was small, and the left pulmonary artery and its branches were prominent. An enlarged right bronchial artery was noted. The aortic arch was on the left side. In addition, images in the lung window (Figure 2) revealed increased reticular markings at the periphery of right lung. Mosaic attenuation was seen in the left lung. The bronchial branching pattern was normal. It was concluded that the patient had congenital interruption of right pulmonary artery, and the right para-tracheal opacity noted on the plain radiograph was attributed to the thymus.

An echocardiogram revealed no associated congenital heart disease. Conservative management was adopted and regular follow-up was arranged.

Case 2

A 37-year-old man with known uncorrected Tetralogy of Fallot since childhood presented with haemoptysis in 1999.

Review of his chest radiographs (Figure 3) showed a grossly enlarged heart (probably related to Tetralogy of Fallot), a right-sided aortic arch, and an oligaemic left lung. The right pulmonary artery and pulmonary trunk were enlarged, but the left hilum was indistinct. Rib notchings were noted at the left 4th to 6th ribs.

Subsequently, a contrast CT thorax was performed as investigation for the haemoptysis, and the images in the soft tissue window (Figure 4) revealed interruption of left pulmonary artery. The right pulmonary artery and its branches were prominent while the inter-lobar portion of the left pulmonary artery was small. An enlarged left bronchial artery was seen, and prominent left intercostal arteries (with associated serrated left pleural thickening) were noted. There was a right-sided aortic arch and descending thoracic aorta. The heart was enlarged with pericardial effusion and pleural effusion, all probably related to Tetralogy of Fallot with heart failure. Lung window images showed increased reticular markings at the periphery of left lung.

The patient was then assessed by cardiothoracic surgeons and considered unsuitable for corrective surgery of Tetralogy of Fallot. He eventually developed heart failure and passed away in 2000.

DISCUSSION

Congenital interruption of a pulmonary artery is a

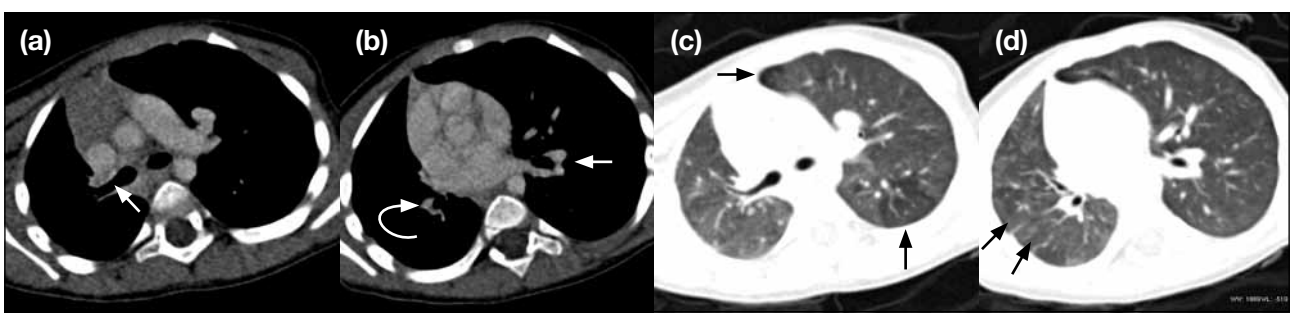

Figure 2. Contrast computed tomographic (CT) thorax of the first patient — at the soft tissue window: (a) there is absence of the mediastinal portion of the right pulmonary artery; a prominent right bronchial artery is illustrated (white arrow), and (b) there is volume loss of the right lung with herniation of the left lung to the contralateral side; branches of the left pulmonary artery are prominent (straight arrow) compared to the inter-lobar portion of the right pulmonary artery (curved arrow). CT thorax of the first patient — at the lung window: (c) normal bronchial branching pattern is demonstrated; mosaic attenuations (black arrows) are noted in normal left lung, and (d) increased reticular pattern is noted at periphery of right lung (black arrows).

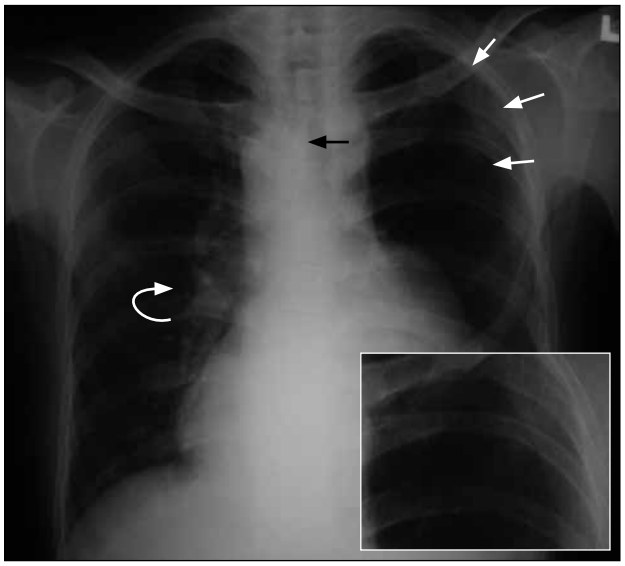

Figure 3. Plain chest radiograph of the second patient. There is cardiomegaly and a right-sided aortic arch (black arrow). The left lung is oligemic, and the left hilum is indistinct, while the right hilar vessels (curved white arrow) and pulmonary trunk are prominent. Rib notchings are present at the left 4th to 6th ribs (straight white arrows) [inset].

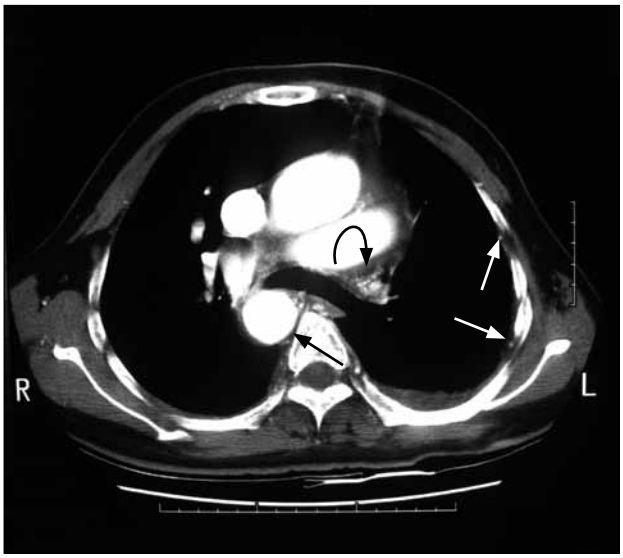

Figure 4. Contrast computed tomographic thorax of the second patient — at the soft tissue window. The left pulmonary artery is absent. There is an enlarged left bronchial artery (curved black arrow). Prominent left intercostal arteries (straight white arrows) are evident with associated serrated pleural thickening. There is also a right-sided aortic arch (straight black arrow) and a left pleural effusion.

rare disorder. Its prevalence was estimated to be 1 in 200,000.[1,2] The interrupted pulmonary artery is more common on the right side (63%),[1] and is usually located on the side opposite to the aortic arch.[1,3] Other congenital heart diseases such as Tetralogy of Fallot are associated with interruption of left pulmonary artery.[3] This was illustrated in our second patient.

Various radiological signs of an interrupted pulmonary artery can be detected on the plain chest radiograph. On the affected side, the hemi-thorax usually shows evidence of volume loss and the ipsilateral hilum is small and indistinct, because the corresponding pulmonary artery is interrupted. In well-established cases, rib notching may be visible due to collaterals from intercostal arteries. The unaffected hemi-thorax is often subject to compensatory hyperinflation of the lung, which may herniate into the affected side. The hilum of the unaffected side is enlarged, because it receives the entire output from right ventricle. Most of these radiological features are well demonstrated in the chest radiographs of our patients.

These features are better appreciated in CT scans, which clearly show the abnormal vascular anatomy with intravenous contrast. The interruption of the pulmonary artery can be revealed and collaterals supplying the affected lung are often visualised. The latter may be derived from bronchial, intercostal, internal mammary, subclavian or innominate arteries.[3] Secondary signs of collateral intercostal arteries such as rib notching and serrated pleural thickening may be identified. The CT scan in the lung window is useful to confirm a normal bronchial branching pattern. Increased sub-pleural interstitial lung marking is another secondary sign of collateral intercostal arteries. Mosaic attenuation may be seen in both lungs, and is better revealed by high-resolution CT. The mosaic pattern in the affected lung may be related to hypoxic vasoconstriction and that in the unaffected lung to over-perfusion. Most of these radiological features were visualised in the CT images of our patients.

Magnetic resonance imaging (MRI) has been used to evaluate the vascular anatomy of interrupted pulmonary arteries.[1-4] It avoids ionising radiation and the use of iodinated intravenous contrast.

Echocardiography is helpful in detecting and assessing the severity of congenital heart diseases, which are associated with interruption of a pulmonary artery. MRI also has a role in this respect.

Major radiological differential diagnoses of isolated interruption of the pulmonary artery include Swyer-James syndrome and hypogenetic lung syndrome. Swyer-

James syndrome is related to childhood viral infection causing bronchiolitis and obliteration of small airways. While the affected lung is hyperlucent, the volume loss is less severe and there is evidence of air-trapping.[2,3] On the other hand, presence of scimitar sign, which represents an anomalous vein draining from right lung to below diaphragm, helps differentiate hypogenetic lung syndrome from interruption of pulmonary artery.[3]

Clinically, some patients with isolated interruption of a pulmonary artery remain asymptomatic for long periods and are incidentally detected to have abnormal chest radiographs.[1,2,5] Others present with shortness of breath, recurrent pneumonia, haemoptysis, or other symptoms of pulmonary hypertension or heart failure.[1,2,5] Such symptoms may be unmasked by pregnancy and high altitude,[1,5] owing to the presence of a hyper-dynamic circulation. In patients with unilateral pulmonary artery agenesis, haemoptysis can result from an excessive collateral circulation.[1] Although isolated interruption of the pulmonary artery may have an indolent course, the development of pulmonary hypertension and haemoptysis have a bearing on long-term survival. Overall mortality is reported as 7% per year after diagnosis.[1]

There is still no consensus regarding the treatment of isolated interruption of the pulmonary artery. In asymptomatic patients, some clinicians adopt conservative management with close follow-up. Others opt for early revascularisation of the interrupted pulmonary artery to restore a physiological pulmonary circulation,[1,5] which may lead to regression of pulmonary hypertension.[1,5] If massive haemoptysis ensues, selective embolisation of systemic collaterals may be an option.[1,6]

In conclusion, interruption of pulmonary artery is a rare condition. Its clinical presentation is non-specific and it is sometimes asymptomatic. Recognition of the relevant radiological features is the keystone of diagnosis.

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