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CASE REPORT

Multidetector Computed Tomography in the Diagnosis of Pulmonary Sling: Case Report

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ABSTRACT

Pulmonary artery sling is a rare developmental vascular abnormality. In this abnormality, the left pulmonary artery arises from the right pulmonary artery, courses between the trachea and oesophagus, and forms a sling around the distal trachea. This arrangement gives rise to upper respiratory obstruction. This case report, of a 3-month-old boy, highlights the use of multidetector computed tomography as an imaging modality to diagnose pulmonary artery sling and to enable rapid diagnosis of airway obstruction in infants.

Key Words: *Heart defects, congenital; Infant; Pulmonary artery/abnormalities; Tomography, X-ray computed/methods*

INTRODUCTION

Pulmonary artery sling is a rare developmental vascular abnormality in which the left pulmonary artery arises from the right pulmonary artery and courses between the trachea posteriorly and oesophagus anteriorly. This arrangement creates a sling around the distal trachea and the proximal main bronchus. Examples of associated abnormalities are complete tracheal rings, tracheal anomalies, and cardiac anomalies.[1] Computed tomography (CT) and magnetic resonance imaging (MRI) have been used for the diagnosis of both tracheobronchial tree anomalies and vascular rings. MRI is currently regarded as the standard imaging method when evaluating paediatric airway obstruction.[2] However, long acquisition times and motion artefacts pose problems. The development of spiral CT has provided another imaging modality for evaluation of paediatric airway obstruction.[3] This case report demonstrates the valuable role of multidetector spiral CT in diagnosing pulmonary sling, as well as the ability of CT to demonstrate other known associations of the condition.

CASE REPORT

A 3-month-old boy had been born at 36 weeks of gestation. At 3 weeks of age, he developed an episode of respiratory distress and a diagnosis of bronchiolitis was made. At age 2 months, his respiratory distress worsened and required intubation and assisted ventilation in a hospital. During endotracheal intubation, the paediatrician was unable to insert a small (size 2.5) endotracheal tube 1 cm beyond the vocal cord. The patient was then referred to the University of Malaya Medical Centre, Kuala Lumpur, Malaysia, in November 2002 for further management.

Physical examination performed on arrival revealed a good-size baby. No stridor was heard. The peripheries were cold and no cardiac murmurs were heard, but there was a general reduction in air entry in both lungs. A plain chest X-ray revealed upper-lobe consolidation in the right lung. Contrast-enhanced spiral CT using a LightSpeed 16-slice CT scanner (GE Medical Systems, Milwaukee, WI, United States) with 10 mL of iopromide contrast medium (Ultravist 300; Schering AG, Berlin, Germany) was performed. Scanning was started after the injection of 5 mL of contrast medium. Scanning settings were as follows: pitch, 1.375; thickness, 0.625 mm; and interval with no gap. Retro-reconstruction of the raw data — using a 0.625-mm slice interval and a 1.25-mm slice thickness — demonstrated a pulmonary sling and collapse consolidation of the left upper lobe and consolidation of the right upper lobe. In addition, an anomalous right upper lobe bronchus was detected (Figures 1-3). Echocardiography showed levocardia, atrial situs solitus, and a dilated main pulmonary artery, as well as the right pulmonary artery and the left

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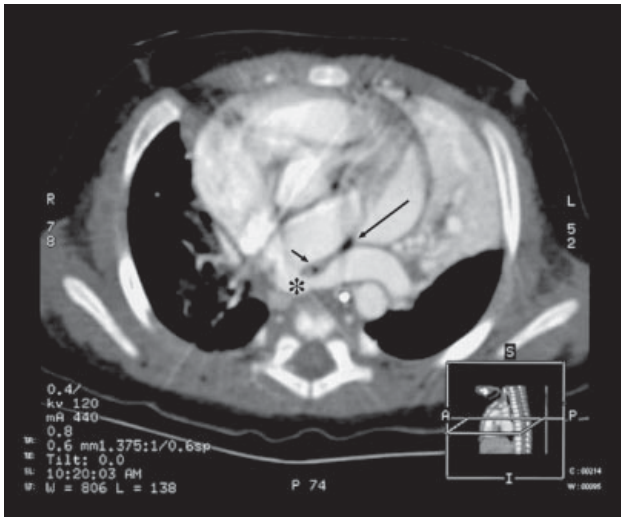

Figure 1. Axial computed tomogram of the thorax following intravenous contrast enhancement showing the aberrant left pulmonary artery (asterisk) arising from the right pulmonary artery, coursing behind the trachea, and causing compression and appreciable narrowing of the distal trachea (short arrow); and left upper lobe collapse consolidation with a patent left upper-lobe bronchus (long arrow).

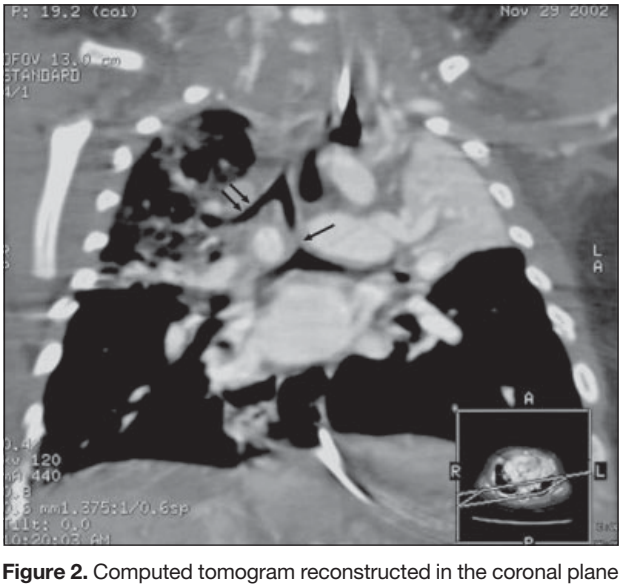

Figure 2. Computed tomogram reconstructed in the coronal plane showing an aberrant right upper lobe bronchus (double arrow), near-total occlusion of the distal trachea (single arrow), and left upper-lobe collapse consolidation.

pulmonary artery arising from the distal right pulmonary artery (Figure 4). Bronchoscopy revealed tracheomalacia, narrowing of the distal trachea, and a narrowed origin of the right main bronchus due to compression. The origin of the aberrant right upper lobe bronchus was not visible.

The patient underwent surgery to release the pulmonary artery sling; tracheobronchoplasty and right upper lobectomy were also performed. Findings at surgery confirmed

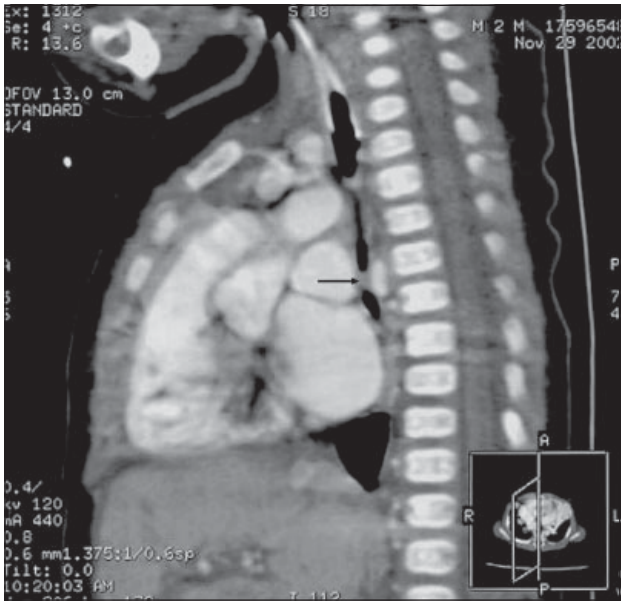

Figure 3. Computed tomogram reconstructed in the sagittal plane showing total occlusion of the distal trachea by the anomalous vessel posteriorly (arrow).

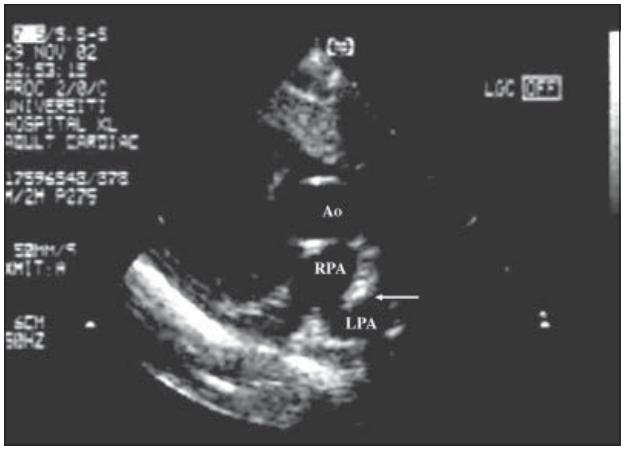

Figure 4. Two-dimensional echocardiogram in the parasternal short axis view showing anomalous origin of the left pulmonary artery (LPA) from the distal right pulmonary artery (RPA), the aorta (Ao), and the trachea (arrow). [*Image courtesy of Dr. Zarin Ikmal Zan. Mohd Zain, Department of Paediatrics, University of Malaya Medical Centre.*]

the presence of a left pulmonary artery arising from the distal right pulmonary artery, slinging around the trachea, and causing compression of the distal trachea, as well as a 2-cm stenotic tracheal segment proximal to the sling. An aberrant right upper lobe bronchus with a non-aerated non-functional right upper lobe was also present. There were no cardiac or great-vessel abnormalities.

Postoperatively, the patient developed a right pneumothorax, which was complicated by right lung injury caused by the intercostal drain. A repair was attempted, but the patient died despite attempts at resuscitation.

DISCUSSION

The pulmonary sling is a rare developmental vascular abnormality thought to be due to the embryonic presence of a common pulmonary mass between the developing right and left pulmonary arteries. Failure of the developing left lung to make contact with the pulmonary artery results in a persistent connection with the right pulmonary artery.[1] The anomalous left pulmonary artery originates from the right pulmonary artery and supplies the left lung. It courses over the right main bronchus, posterior to the trachea or carina and anterior to the oesophagus, and reaches the hilum of the left lung. The arrangement causes compression to the lower trachea and right main bronchus, which produces upper-airway obstructive symptoms. Compression caused by the sling can produce obstructive emphysema and atelectasis of the lungs.

There are also associated abnormalities in the arterial supply to 1 or both lungs. Occasionally, the anomalous pulmonary artery supplies the left upper lobe while maintaining normal pulmonary arterial supply to the left lower lobe. In addition, partial anomalous supply of the right upper lobe of the lung from this anomalous artery has been described.[4]

Besides other arterial abnormalities, associated abnormality of the trachea at the level of the aberrant vessel, such as complete tracheal rings, may be found.[1] Other tracheal anomalies that may occur include an anomalous origin of the right upper lobe bronchus from the trachea (i.e., bronchus suis)[5] and tracheomalacia. Hypoplasia and stenosis of tracheal segments may also occur, and these potentiate airway obstruction.

Congenital heart defects are found in 50% of patients with pulmonary sling.[4] The most common are patent ductus arteriosus (25%), atrial septal defect (20%), left superior vena cava (20%), and ventricular septal defect (10%).[6] Abnormalities in the other systems are also described. These include imperforate anus, Hirschsprung's disease, biliary atresia, and genitourinary defects. Abnormalities of the ovaries, vertebrae, thyroid gland, and pulmonary parenchyma have also been reported.[4]

Clinical symptoms include respiratory distress manifested by stridor, recurrent pulmonary infections, wheezing, and cyanosis. These symptoms typically occur within the first month of life. Although dysphagia is rare, obstructive apnoea may occur. Patients with pulmonary sling tend to be more symptomatic and present earlier than those with a vascular ring due to a double aortic arch. When tracheal obstruction exists, the infant may have constant retractions and tachypnoea.[4]

Findings from the chest X-ray include deviation of the lower trachea to the left, which may appear compressed on its right side. Hyperinflation of the right lung may be seen because of impingement or compression of the right main bronchus. The left lung may appear hyperinflated because of obstruction at the level of the carina and the left main bronchus. In patients with severe obstruction, atelectasis of a single lung or single lobe is uncommon. A lateral chest X-ray may demonstrate a rounded density anterior to the oesophagus, which is actually the left pulmonary artery viewed end-on. A barium swallow examination is performed if the infant is not critically ill. On the lateral projection, the barium-filled oesophagus will show an anterior indentation; the frontal projection will show displacement of the oesophagus with an oblique impression immediately posterior to the carina.

Echocardiography will identify the pulmonary trunk and its branches, in addition to other associated cardiac defects. In the coronal plane scanned through the suprasternal notch, there will be an absence of the normal bifurcation of the main pulmonary trunk into the right and left pulmonary arteries,[4] and only the right pulmonary artery is seen to arise from the pulmonary trunk. Furthermore, the left pulmonary artery can be seen to arise from the right pulmonary artery on the echocardiogram.

Bronchoscopy is generally not recommended.[4] If it is performed, tracheal compression is noted and accompanying tracheomalacia or tracheal stenosis is commonly seen. Some physicians recommend bronchoscopic evaluation of selected surgical candidates, because surgical reconstruction of the trachea or bronchi may be necessary if the airway compromise is severe.

Pulmonary artery angiography is now seldom performed because of the widespread use of non-invasive imaging modalities, such as CT and MRI, which can provide excellent morphological abnormalities in 3-dimensional planes before surgical correction.

MRI is now the standard imaging modality when evaluating paediatric airway obstruction.[2,3] It provides

excellent definition of mediastinal, vascular, and airway anatomy in multiple planes without the need for contrast administration.[7] However, the disadvantages of MRI include a long scanning time, the need for sedation, and respiratory artefacts that degrade the image quality. Overall, MRI is a relatively costly examination.

Multidetector spiral CT provides fast volume scanning and enables gapless data acquisition within 1 breath-hold. This approach can avoid data misregistration and reduce motion artefacts. Multidetector spiral CT also provides overlapping images which can be reconstructed retrospectively without additional radiation exposure,[8] thereby providing high image quality and best 3-dimensional resolution. The major disadvantage of this examination method is the radiation dose. In the case described in this report, however, multidetector spiral CT provided the most useful information and enabled rapid diagnosis in a critically ill infant.

In conclusion, we illustrate a case of pulmonary sling diagnosed by contrast-enhanced multidetector spiral CT. In view of its fast scanning time and excellent anatomical resolution, we recommend this imaging modality for the evaluation of congenital vascular abnormality in critically ill infants presenting with airway obstruction.

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