



Case report

Two rare cases of horseshoe lung with scimitar syndrome in Vietnam

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ABSTRACT

Horseshoe lung is a rare, congenital, pulmonary anomaly in which the caudal and basal segments of the left and right lungs are joined together. Most cases of horseshoe lung are associated with scimitar syndrome. Horseshoe lung can be diagnosed using pulmonary angiography, showing that the isthmus of the pulmonary parenchyma crosses the midline into the contralateral side. The isthmus parenchyma is typically supplied by the hypoplastic pulmonary artery. Clinical symptoms, therapeutic methods, and prognosis depend on the incidence of pulmonary hypertension, heart failure, recurrent pneumonia, and other combinations of congenital malformations. In this article, we describe two cases of horseshoe lung associated with scimitar syndrome and pulmonary malformation.

1. Introduction

Horseshoe lung is a rare congenital malformation characterized by the midline connection between the parenchyma from both lungs, behind the heart, and anteriorly to the esophagus, thoracic aorta, and

spinal column [1]. This disease was first described by Spencer in 1962 [2]. Most cases of horseshoe lung are associated with unilateral lung hypoplasia and a partial, anomalous, pulmonary venous return to the systemic venous system [1]. Other associations have also been described, including cardiovascular anomalies [3]. A baby with a

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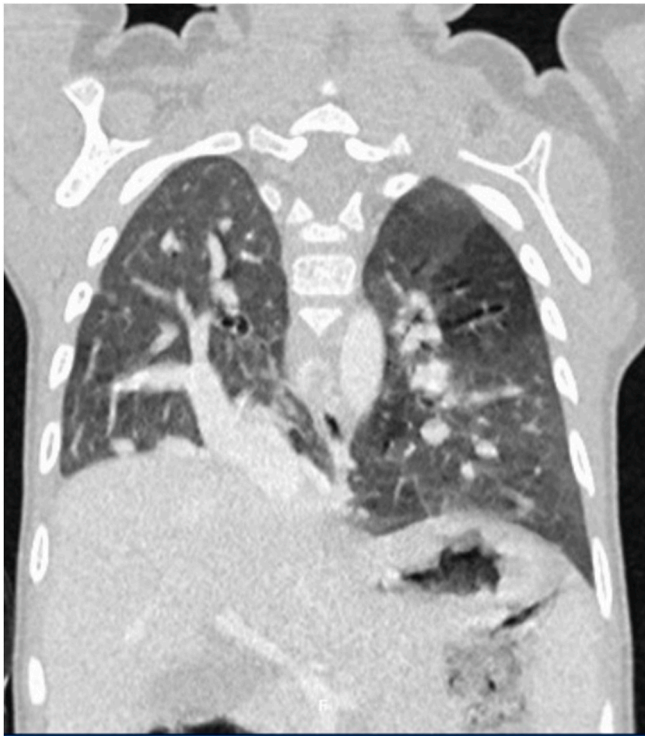


Fig. 1. Coronal chest CT image derived from multi-planar reconstruction with contrast enhancement showed that the right lung displayed reduced volume, suggestive of pulmonary hypoplasia.



Fig. 3. Coronal chest CT image derived from multi-planar reconstruction with contrast enhancement showed the abnormal right pulmonary vein (arrow) draining into the right atrium (arrowhead).



Fig. 2. Axial chest CT image showed that the heart was enlarged and displaced to the right. An isthmus behind the heart and anterior to the esophagus and thorax aorta connected the right and left lungs. A pleural line was observed in the parenchymal isthmus (white arrow). Opacities (arrowhead) and consolidation (black arrow) were observed in the bottom of the right lung.

horseshoe lung typically presents with severe cardio-respiratory distress [4]. When a horseshoe lung is discovered, clinicians should be aware of the associated congenital abnormalities. In the present report, we describe two cases of horseshoe lung accompanied by scimitar syndrome and pulmonary malformation.

2. Case report

2.1. Case 1

A 2-month-old female baby presented to the hospital with dyspnea and slow weight gain. The patient presented with respiratory distress since birth but without cyanosis. The patient was born by Cesarean delivery at 35 weeks of gestation due to the identification of pulmonary hypertension on a prenatal ultrasound. The birth weight was 3000 g, and the bodyweight at admission was 3500 g. A physical examination revealed tachypnea, measured at 62 breaths per min, intercostal retractions, and a systolic murmur of grade 3/6 over the left sternal border. Blood tests, including complete blood cell counts and liver and kidney function tests, were normal. Chest radiographic examination revealed that the heart was displaced on the right side, and the right lung had a reduced volume. Echocardiography showed right pulmonary venous drainage into the right atrium, an atrial septal defect (ASD) of 3 mm in diameter, a left-to-right shunt, and right ventricular dilatation. Pulmonary artery pressure was 40 mmHg on echocardiography. Chest multislice computed tomography (MSCT) with intravenous contrast enhancement revealed the decreased volume of the right lung, and the mediastinum was displaced to the right, which was suggestive of a hypoplastic right lung (Fig. 1). The heart was enlarged, and the cardiothoracic ratio was 0.63 (Fig. 2). An isthmus was identified, connecting the right and left lung, behind the heart and anterior to the esophagus, thorax aorta, and spine, indicating the presence of a pleural line in the parenchymal isthmus (Fig. 2). Opacities and consolidation were observed at the bottom of the right lung (Fig. 2). The isthmus parenchyma featured bronchi originating from the right lung and was supplied by small branches from the right pulmonary artery. Chest MSCT in the venous phase showed that the abnormal right pulmonary vein drained into the right atrium (Fig. 3). A diagnosis of horseshoe lung associated with scimitar syndrome was established, and the patient was treated with antibiotic therapy. The patient then underwent treatment

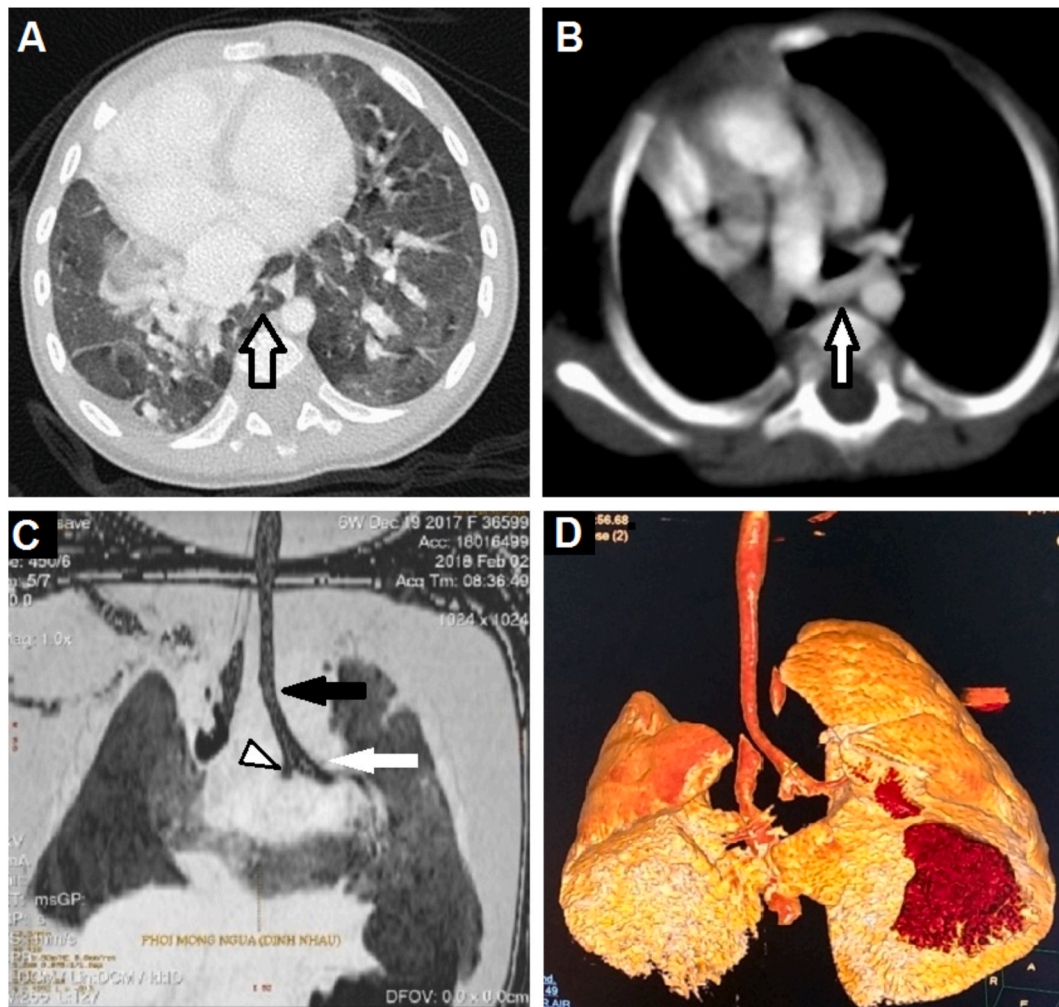


Fig. 4. Axial chest CT image demonstrating the fusion of the posterior segments of the right and left lungs (A, arrow). Trapped air in both lung and consolidation of the right lung was observed (A). The left pulmonary artery originates and encircles posterior to the left main bronchus (B, arrow). Coronal chest CT image derived from multi-planar reconstruction revealed that stenosis was observed in the lower section of the trachea (C, black arrow) and the left main bronchus (C, white arrow). The right main bronchus featured stenosis of greater than 90% of the luminal diameter, and only the main stem bronchus was clearly detected (C, arrowhead). Chest CT image with 3D-volumetry showing stenosis of both main bronchi and reduced volume of the right lung (D).

for atrial septal defects with an Amplatzer septal occluder at the age of 9 months. The patient was followed for 7 years and had one episode of pneumonia.

2.2. Case 2

A 2-month-old female patient was admitted to the hospital with wheezing and respiratory distress, lasting for more than 1 month. Physical examination showed respiratory failure with intercostal retractions. The patient had low blood pH levels (pH 7.12). Chest X-ray revealed opacification of the right lung. Echocardiography showed that an ASD of 2 mm in diameter, the left pulmonary vein returned to the left atrium, and the left pulmonary artery encircled the left main bronchus; the right pulmonary vein was difficult to approach. Chest MSCT demonstrated the fusion of the posterior segments of the right and left lungs, the presence of trapped air in both lung, and the consolidation of the right lung (Fig. 4). The left pulmonary artery originated from the posterior aspect of the right pulmonary artery and encircled posterior to the left main bronchus (Fig. 4). The right pulmonary vein returned to the superior vena cava. Narrowing of the lower segment of the trachea (to a 3-mm diameter, representing an approximately 50% reduction in the luminal diameter) and the left main bronchus (the stenosis site featured a 2-mm diameter, representing an approximately 70% reduction in

luminal diameter) and severe stenosis of the right main bronchus (stenosis representing a greater than 90% reduction in luminal diameter) were also detected (Fig. 4). The right lung was decreased the volume (Fig. 4). Bronchoscopy showed the narrowing of the distal trachea, and the tube was not able to pass through the trachea at the narrow point (Fig. 5). The final diagnosis was horseshoe lung associated with scimitar syndrome, tracheal stenosis, and pulmonary artery sling. This patient presented with recurrent pneumonia and was treated with antibiotic therapy. Surgery was not performed at this time because the tracheal stenosis was very severe.

3. Discussion

Horseshoe lung is a rare congenital anomaly characterized by the fusion of the right and left lungs by a parenchymal isthmus located between the heart and the thorax aorta [5]. The etiology of horseshoe lung remains unclear [6]. Two possibilities exist that may explain the development of horseshoe lung. First, horseshoe lung may be the result of a fusion process that occurs in the parenchyma between the lungs [1]. Second, the nonseparation of the splanchnic mesoderm may result in a connection between the lungs and the pleural cavities [1]. Most horseshoe lung cases are associated with unilateral lung hypoplasia, which occurs more frequently in the right lung than the left lung, accounting

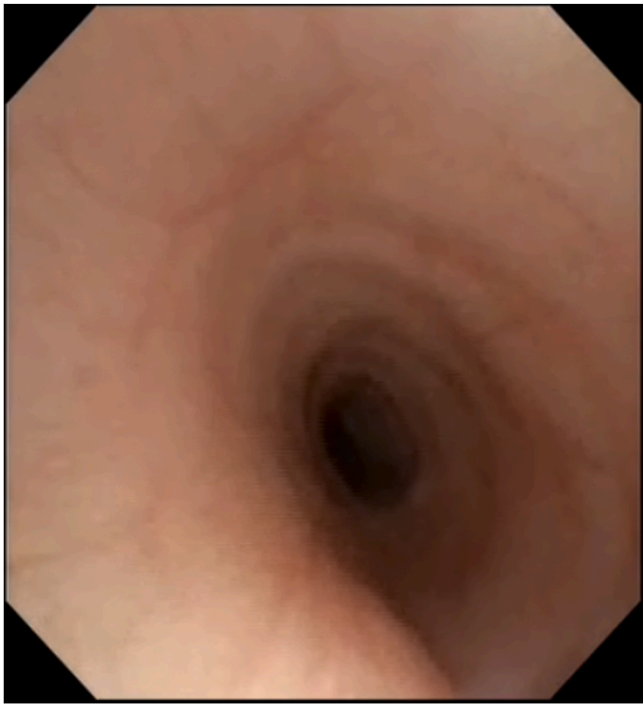


Fig. 5. The narrowing of the distal trachea, which the bronchoscope could not pass through.

for approximately 80% of cases [3,7]. As previously reported by Bando et al. [3], only 10 cases of horseshoe lung have been associated with left lung hypoplasia. Figa et al. [5] classified horseshoe lung into three patterns: (1) lung fusion without the intervening pleura; (2) the presence of two pleural layers between the crossover lung tissue; and (3) the presence of four pleural layers between the crossover lung tissue, and the isthmus lung tissue is surrounded by its own visceral and parietal pleural envelopes. Horseshoe lung has a high incidence of association with scimitar syndrome, with co-occurrence identified in 80%–85% of cases [8]. Scimitar syndrome typically occurs in patients with right lung hypoplasia [3]. A statistic by Bando et al. [3] indicated that no patients with horseshoe lung associated with left lung hypoplasia presented with scimitar syndrome; however, most of these patients present with pulmonary hypertension. Scimitar syndrome is characterized by anomalous pulmonary venous drainage of the hypoplastic right lung into the systemic venous circulation [9]. Other cardiovascular abnormalities have been reported in horseshoe lung, such as atrial septal defects, interventricular septal defects, persistent ductus arteriosus, and tetralogy of Fallot [10]. Horseshoe lung may be associated with bronchopulmonary malformations, such as the absence of a pulmonary artery, pulmonary sling, or esophagobronchial fistula [11,12]. Patients with horseshoe lung may be asymptomatic; however, patients may also present with respiratory symptoms, including respiratory distress, pneumonia, recurrent pulmonary infection, or symptoms of pulmonary hypertension [3,13], although these symptoms are nonspecific. Most patients are diagnosed below the age of 1 year [14]. For the diagnosis of horseshoe lung, CT represents a useful imaging technique [8]. The fusion of both lungs via a parenchymal isthmus can be easily detected on CT images [6]. The pleural line may appear as a thin fibrous band in the parenchymal isthmus [14]. Angiography and bronchography revealed that the bronchial and arterial branches that supply the isthmus arise from the hypoplastic lung [15]. The hypoplastic lung is reduced in volume, and the mediastinum is typically displaced to that side [14]. CT imaging is useful for detecting other abnormalities [15]. One of the differential diagnoses for horseshoe lung is mediastinal lung herniation [3]. Lung herniation can result from inflammatory changes, such as necrotizing

pneumonia, resulting in compensatory hypertrophy of the other lung [16]. Furthermore, lung herniation may occur following thorax surgery [17]. The differentiation between mediastinal lung herniation and horseshoe lung can be very difficult. However, the absence of a pleural line at the isthmus parenchyma and the presence of abnormal pulmonary venous drainage can suggest horseshoe lung [18]. Treatment depends on the clinical symptoms and the identification of any other malformations. Symptomatic treatment or surgery may be applied [3]. The prognosis of horseshoe lung depends on the co-occurrence of other malformations and pulmonary hypertension [11]. Therefore, the early and accurate diagnosis of horseshoe lung is very important.

The two cases we described here presented with symptoms before the age of 1 year and were both associated with scimitar syndrome. The diagnosis was easily determined by thorax MSCT. The first patient had a cardiovascular abnormality in the form of an atrial septal defect and pulmonary hypertension. The second patient had both cardiovascular and bronchopulmonary malformations, presenting with atrial septal defect, severe tracheal and bronchi stenosis, and pulmonary artery sling. Due to trapped air in the right lung, lung hypoplasia was more difficult to evaluate in the second patient than the first patient. Both patients received symptomatic treatment and did not undergo surgery.

4. Conclusion

Horseshoe lung is a rare congenital anomaly commonly associated with scimitar syndrome. This disease tends to be associated with pulmonary hypertension. MSCT is a useful and non-invasive medical imaging technique for the diagnosis of horseshoe lung. Horseshoe lung may be asymptomatic or may present with the symptoms of pneumonia. Treatment and prognosis depend on the degree and occurrence of pulmonary hypoplasia, pulmonary hypertension, and recurrent pneumonia, and the presence of cardiovascular malformations.

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Author contribution

Le TV and Nguyen MD contributed to this article as co-first authors. All authors have read the manuscript and agree to the content.

Declaration of competing interest

Authors do not have any conflict of interests.

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