Case report

Horseshoe lung associated with scimitar syndrome

Korcan Aysun Gonen,[1] Yakup Canitez,[2] Ozlem Mehtap Bostan,[2] Zeynep Yazici[3]

[1]Department of Radiology, Namik Kemal University, Tekirdag, Turkey
[2]Department of Pediatrics, Uludag University, Bursa, Turkey
[3]Department of Radiology, Uludag University, Bursa, Turkey

Correspondence to
Professor Zeynep Yazici, zyazici@uludag.edu.tr

Presented as a poster at the European Society of Paediatric Radiology 50th Annual Meeting, 3–7 June 2013, Budapest, Hungary.

Accepted 22 July 2019

SUMMARY

Horseshoe lung is a rare congenital anomaly and mostly accompanied by scimitar syndrome. Most aspects of this complex anomaly can be demonstrated via multidetector CT (MDCT). We present two baby girls who had horseshoe lung associated with right lung hypoplasia and scimitar vein. The chest roentgenograms showed displacement of the heart and mediastinum to the right with smaller right lung. Echocardiography revealed dextroposition, secundum atrial septal defect and bilateral slight peripheral pulmonary stenosis in the first case and dextroposition, severe pulmonary hypertension, secundum atrial septal defect and tricuspid regurgitation in the other one. On thoracic MDCT, the right lung and pulmonary artery were hypoplastic with cardiomediastinal shift to the right. There was an abnormal right pulmonary vein draining into the inferior vena cava on the lower zone of the right lung (scimitar vein). The posterobasal portions of the both lungs were fused through a midline isthmus behind the heart.

BACKGROUND

Horseshoe lung is a rare congenital bronchopulmonary anomaly in which the posterobasal portions of either lung are fused by a narrow isthmus of pulmonary parenchyma, posterior to the heart and anterior to the aorta and oesophagus.[1 2] The most common associated feature of the horseshoe lung is unilateral lung hypoplasia.[3] Horseshoe lung with right lung hypoplasia is more commonly seen in the scimitar syndrome.[4] The incidence of cardiovascular anomalies in this syndrome is 25%.[4] At the present time, comprehensive diagnosis for these rare complex anomalies is possible by MDCT without invasive examinations such as catheter angiography and bronchoscopy. Here, we present two patients of horseshoe lung associated with scimitar syndrome diagnosed by MDCT.

CASE PRESENTATION

Two infant patients, aged 3 and 4 months, with cardiac dextropositon and small right lung were examined with CT angiography. The examinations were performed on a 128-section CT scanner (Somatom Definition AS, Siemens, Erlangen, Germany). Non-ionic contrast medium (300 mg iodine/mL; 2 mL/kg) was injected manually in the first case and using a power injector in the second case. Three-dimensional (3D) volume-rendered (VR), maximum intensity projection (MIP) and minimum intensity projection (MinIP) images were obtained from the axial images using a separate workstation (Leonardo; Siemens Medical Solutions).

Case 1: the physical examination of a 3-month-old girl patient was unremarkable except heart murmur. On chest radiogram, her heart appeared to be displaced towards the right side and the right lung had reduced volume. Echocardiography revealed cardiac dextroposition, a small secundum atrial septal defect and mild peripheral pulmonary stenosis. Because of the presence of combination of pulmonary and cardiac pathology, thoracic CT angiography was performed. CT images at pulmonary window setting showed fusion of the posterobasal portions of either lung behind the heart, which was well demonstrated on a 3D VR image (figure 1A,B). There was a linear density between the isthmic portion and the left lung, representing an intervening pleural line (figure 1A). Due to air trapping, the horseshoe lung segment had decreased attenuation. A small branch arising from the right pulmonary artery supplied the horseshoe lung (figure 1C). The bronchi to the horseshoe lung arose from the right lower lobe bronchi (figure 1D). A venous structure indicating a scimitar vein traversed the right lower lung inferomedially towards diaphragm to join the IVC between the hepatic veins and the right atrium (figure 1E). The venous drainage of the horseshoe lung segment was also into the IVC (figure 1F). The right pulmonary artery was smaller than the left one.

Case 2: a heart murmur was detected in a 4-month-old girl presenting with respiratory distress. The chest radiogram of the patient revealed a small right lung and dextroposition of the cardiomediastinal structure. The echocardiographic findings were dextroposition of the heart, severe pulmonary hypertension, secundum atrial septal defect, severe tricuspid regurgitation and suspicion of right pulmonary sequestration. Thoracic CT angiography was performed. CT images demonstrated fusion of the posterobasal portions of both lungs via a parenchymal isthmus behind the heart with a pleural line between the isthmic portion and the left lung (figure 2A,B). The horseshoe lung segment had low attenuation consistent with air trapping (figure 2A). The right pulmonary artery was smaller compared with the left pulmonary artery (figure 2C). Enlarged two aberrant vessels in the right lower lung united at the base of the right lung immediately before connecting to the IVC, which were consistent with two scimitar veins (figure 2D,E). The arterial supply of the horseshoe lung arose from the right pulmonary artery (figure 2F). The bronchi to the horseshoe lung followed a similar pattern to the arterial supply and

To cite: Gonen KA, Canitez Y, Bostan OM, *et al*. *BMJ Case Rep* 2019;**12**:e204389. doi:10.1136/bcr-2014-204389

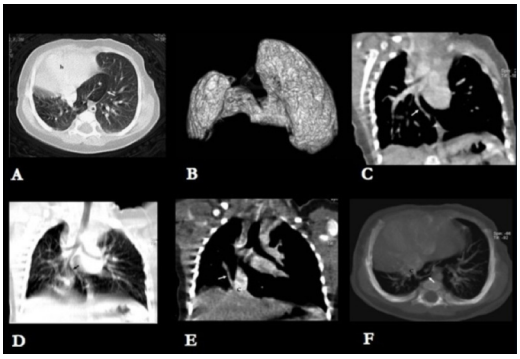

Figure 1 Case 1: a 3-month-old girl. Axial CT image (A) and three-dimensional volume-rendered CT image (B) demonstrate fusion of the posterobasal portions of the both lungs via a parenchymal isthmus behind the heart (h) and in front of the aorta (a) to form the horseshoe lung (asterisk). There is a pleural line (arrow (A)) between the two lungs. The heart (h) is displaced to the right because of the right lung hypoplasia. Oblique coronal slab MIP CT image (C) shows a small branch (arrow) from the right pulmonary artery supplying the horseshoe lung. Coronal slab MinIP CT image (D) shows the bronchi to the horseshoe lung (arrow) arising from the right lower lobe bronchi in a pattern similar to the arterial supply. Oblique coronal slab MIP CT image (E) reveals the abnormal pulmonary vein, the so-called scimitar vein (arrow) draining to the inferior vena cava (IVC) (c). Axial slab MIP CT image (F) shows that the pulmonary vein of the horseshoe lung (arrow) drains to the IVC (c) where the scimitar vein joins to the IVC. MinIP, minimum intensity projection; MIP, maximum intensity projection.

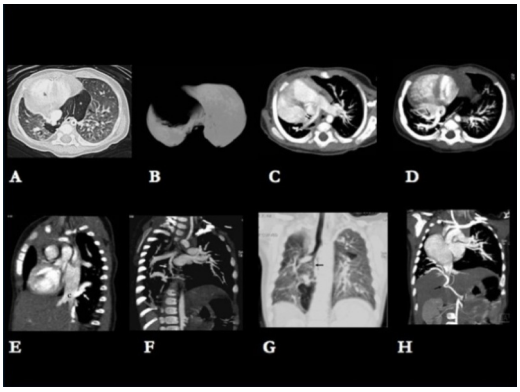

Figure 2 Case 2: a 4-month-old girl. Axial CT image (A) and three-dimensional volume-rendered CT image (B) demonstrate fusion of both lungs via a parenchymal isthmus behind the heart (h) and in front of the aorta (a) to form the horseshoe lung (asterisk). There is a pleural line (arrow (A)) between the two lungs. The heart (h) is displaced to the right because of the right lung hypoplasia. Axial slab MIP CT angiogram (C) shows the right pulmonary artery (double arrows) smaller compared with the left pulmonary artery (arrow). Axial (D) and oblique sagittal (E) slab MIP CT angiograms demonstrate two scimitar veins (arrows) uniting into a common trunk immediately before their confluence into the IVC (c). Oblique sagittal slab MIP CT angiogram (F) shows a small branch (arrow) from the right pulmonary artery supplying the horseshoe lung. Coronal slab MinIP CT image (G) shows the bronchi to the horseshoe lung (arrow) arising from the right lower lobe bronchi in a pattern similar to the arterial supply. Axial slab MIP CT angiogram (H) shows a systemic arterial supply (arrow) from the coeliac artery to the hypoplastic right lung. MinIP, minimum intensity projection; MIP, maximum intensity projection.

arose from the right lower lobe bronchi (figure 2G). An aberrant artery originating from the coeliac artery extended to the right lung (figure 2H).

INVESTIGATIONS

As discussed in the case presentation section.

TREATMENT

Because of the severe pulmonary hypertension, the 4-month-old patient (case 2) underwent surgical occlusion of the abnormal aortopulmonary collaterals at 9 months of age.

OUTCOME AND FOLLOW-UP

The 3-month-old girl (case 1) was followed up for 10 months and remained in a good condition. The second patient (case 2) got a partial benefit from the surgery. She was under the follow-up of the division of paediatric cardiology.

DISCUSSION

Both horseshoe lung and scimitar syndrome are rare congenital anomalies; however, the association of the horseshoe lung with scimitar syndrome is common, with the incidence as high as 80%–85%.[5–8] In horseshoe lung, posterobasal segments of lungs fuse by a narrow isthmus behind pericardial reflection between heart and aorta.[9 10] Fusion happens through common parietal pleural defect that provides communication of the pleural cavities.[9] This may happen directly without fissure or indirectly through intervening visceral pleura.[1 11] Figa *et al*[10] proposed a horseshoe lung classification based on the pleural anatomy. According to this classification, three patterns are available: (1) the lungs show a complete fusion in baseline without intervening pleura, (2) a lung segment extends towards opposite haemithorax, and there is a unilateral pleural layer and (3) there are bilateral pleural layers, and horseshoe lung is a separate lobe in its own visceral pleura. In our cases, the pattern is consistent with the type II since there was a unilateral pleural layer in each one. Radiographic appearance of the horseshoe lung depends on the presence of pleura around isthmic parenchyma and its layout.[3] This pleural line appears as 'well demarcated faint linear or curvilinear density' in the medial part of basal region of the lung.[12] Pleural line around the horseshoe lung can readily be seen on CT scan.[2] In horseshoe lung, arterial and bronchial supply of the isthmic portion is invariably from the hypoplastic lungs.[1 2 6 8–10 13] Historically, conventional angiography and bronchography were used to show this. A branch from inferior pulmonary artery of hypoplastic lung is seen to extend to the base of opposite lung passing through midline on pulmonary arteriography; bronchus of the horseshoe segment is demonstrated to originate from hypoplastic lung on bronchography.[2] Nowadays, MDCT could potentially replace these conventional imaging methods. The embryology of horseshoe lung has not been completely understood. The development of the lower respiratory system begins during the fourth week of gestation as an outgrowth from the ventral wall of the foregut (respiratory diverticulum). The cartilagenous, muscular and connective tissue of the trachea and lungs are derived from splanchnic mesoderm. The respiratory diverticulum is induced by a mass of splanchnic mesoderm, and the whole mass divides into left and right lung. It is assumed that the horseshoe lung could result from non-seperation of the mesodermal mass.[4 13] Horseshoe lung is associated with different foregut and bronchopulmonary malformations such as bilateral intralobar pulmonary sequestration, congenital cystic adenomatoid malformation, oesophageal atresia with tracheoesophageal fistula and oesophagobronchial fistula.[3 5 9 14 15] Scimitar syndrome

or hypogenetic lung syndrome is the most common abnormality accompanying the horseshoe lung.[5–8] It is characterised by lung and pulmonary arteries hypoplasia, ipsilateral pulmonary venous return anomaly (drains into the systemic veins fully or partially), abnormal bronchial branching and lobulation and systemic arterial presentation to the lower lobe of the same lung.[16 17] The combination of these abnormalities is usually unilateral and usually involves the right side. In the literature, there is no reported case with left-sided scimitar syndrome associated with horseshoe lung. However, there are a few reported cases of bilateral scimitar syndrome with horseshoe lung.[11 16 17] The scimitar veins are commonly multiple, and these drain the supradiaphragmatic IVC in horseshoe lung associated with scimitar syndrome in childhood.[11 18 19] In a study of 16 patients diagnosed with scimitar syndrome, the scimitar vein drained to the infradiaphragmatic IVC in 10 patients, but the majority of the patients were adult and none of them had horseshoe lung.[20] The scimitar vein was both single and infradiaphragmatic in our first patient. Dupuis *et al*[21] found six cases of horseshoe lung in a group of 147 scimitar syndrome cases. They reported that 'the prognosis of scimitar syndrome does not seem to be worse when associated with horseshoe lung'. Horseshoe lung associated with scimitar syndrome can easily be diagnosed by MDCT. Although CT is seriously affected by respiratory motion artefact in young children, this limitation may be overcome with faster image acquisition time or respiratory-gated examination.[2] Examination by using a 128-section CT scanner significantly shortens the scanning time for larger body parts and so helps resolve the problem of respiratory artefacts. It also offers non-invasive diagnostic evaluation. Obtaining higher quality reformed images, three-dimensional MIP, MinIP, VR and shaded surface display images is the superiority of MDCT. Simultaneous evaluation of parenchymal, vascular and bronchial abnormalities increases the diagnostic value of CT as distinct from MRI. The major disadvantage of MDCT is the associated radiation exposure. However, the use of single phase study can reduce radiation dose. Additionally, CT angiography can obviate the necessity for catheter angiography, which leads to substantially higher radiation exposure. In conclusion, horseshoe lung associated with scimitar syndrome is a rare and complex anomaly. Many facets of this anomaly can be identified by MDCT without the need for more invasive techniques such as catheter angiography and bronchoscopy.

Learning points

- Both horseshoe lung and scimitar syndrome are rare congenital anomalies.
- Involvement in this syndrome is usually unilateral and affects mostly the right side.
- As the degree of pulmonary hypoplasia and associated various congenital anomalies are essential in prognosis and surgical planning, accurate and early diagnosis of these complex bronchovascular anomalies is important.
- The scimitar veins are commonly multiple, and these drain the supradiaphragmatic inferior vena cava in horseshoe lung associated with scimitar syndrome in childhood.
- Multidetector CT is a useful and non-invasive diagnostic imaging procedure in demonstrating vascular and bronchial anomalies in young children with horseshoe lung and therefore eliminates more invasive methods such as catheter angiography, bronchography or bronchoscopy.

Contributors KAG drafted the article and approved the final version. YC and OMB contributed to the conception and design and prepared the paper initially. ZY revised the manuscript critically for important intellectual content.

Funding The authors have not declared a specific grant for this research from any funding agency in the public, commercial or not-for-profit sectors.

Competing interests None declared.

Patient consent for publication Parental/guardian consent obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

REFERENCES

1 Yildiz AE, Oguz B, Haliloglu M. Horseshoe lung associated with left lung hypoplasia and single left pulmonary vein; a rare combination. *Clin Radiol* 2012;67:86–8.
2 Goo HW, Kim YH, Ko JK, Jk K, *et al*. Horseshoe lung: useful angiographic and bronchographic images using multidetector-row spiral CT in two infants. *Pediatr Radiol* 2002;32:529–32.
3 Goldberg S, Ringertz H, Barth RA. Prenatal diagnosis of horseshoe lung and esophageal atresia. *Pediatr Radiol* 2006;36:983–6.
4 Jeewa A, Culham JA, Human DG. A case of horseshoe lung and complex congenital heart disease in a term newborn. *Pediatr Radiol* 2010;40:206–9.
5 Wales PW, Drab SA, Connolly B, *et al*. Horseshoe lung in association with other foregut anomalies: what is the significance? *J Pediatr Surg* 2002;37:1205–7.
6 Takahashi M, Murata K, Yamori M, *et al*. Horseshoe lung: demonstration by electron-beam CT. *Br J Radiol* 1997;70:964–6.
7 D'Alessandro L, Kovesi T, Massoud S, *et al*. Horseshoe lung and facio-auriculo-vertebral sequence: a previously unreported association. *Pediatr Pulmonol* 2006;41:592–6.
8 Tilea B, Garel C, Delezoide AL, *et al*. Prenatal diagnosis of horseshoe lung: contribution of MRI. *Pediatr Radiol* 2005;35:1010–3.
9 Hawass ND, Badawi MG, Fatani JA, *et al*. Horseshoe lung with multiple congenital anomalies. Case report and review of the literature. *Acta Radiol* 1987;28:751–4.
10 Figa FH, Yoo SJ, Burrows PE, *et al*. Horseshoe lung--a case report with unusual bronchial and pleural anomalies and a proposed new classification. *Pediatr Radiol* 1993;23:44–7.
11 Akay HO, Kervancioglu M, Nazaroglu H, *et al*. Horseshoe lung associated with rare bilateral variant of scimitar syndrome: demonstration by 64-slice MDCT angiography. *Pediatr Radiol* 2008;38:563–6.
12 Frank JL, Poole CA, Rosas G. Horseshoe lung: clinical, pathologic, and radiologic features and a new plain film finding. *AJR Am J Roentgenol* 1986;146:217–26.
13 Oguz B, Alan S, Ozcelik U, *et al*. Horseshoe lung associated with left-lung hypoplasia, left pulmonary artery sling and bilateral agenesis of upper lobe bronchi. *Pediatr Radiol* 2009;39:1002–5.
14 Cerruti MM, Marmolejos F, Cacciarelli T. Bilateral intralobar pulmonary sequestration with horseshoe lung. *Ann Thorac Surg* 1993;55:509–10.
15 Jamieson DH, Fisher RM. Communicating bronchopulmonary foregut malformation associated with esophageal atresia and tracheo-esophageal fistula. *Pediatr Radiol* 1993;23:557–8.
16 Kabbani M, Haider N, Abu-Sulaiman R. Bilateral scimitar syndrome. *Cardiol Young* 2004;14:447–9.
17 Kamijoh M, Itoh M, Kijimoto C, *et al*. Horseshoe lung with bilateral vascular anomalies: a rare variant of hypogenetic lung syndrome (scimitar syndrome). *Pediatr Int* 2002;44:443–5.
18 Bharati A, Merchant SA, Garekar S, *et al*. Horse-shoe lung-rediscovered via volume rendered images. *Indian J Radiol Imaging* 2013;23:297–300.
19 Dische MR, Teixeira ML, Winchester PH, *et al*. Horseshoe lung associated with a variant of the 'scimitar' syndrome. *Br Heart J* 1974;36:617–20.
20 Masrani A, McWilliams S, Bhalla S, *et al*. Anatomical associations and radiological characteristics of Scimitar syndrome on CT and MR. *J Cardiovasc Comput Tomogr* 2018;12:286–9.
21 Dupuis C, Rémy J, Rémy-Jardin M, *et al*. The "horseshoe" lung: six new cases. *Pediatr Pulmonol* 1994;17:124–30.