Horse-shoe lung-rediscovered via volume rendered images

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Abstract

Horseshoe lung, usually associated with pulmonary venolobar syndrome, is a rare congenital anomaly involving the fusion of the postero-basal segments of the right and left lungs across the midline. The fused segment or the isthmus lies posterior to the pericardium and anterior to the aorta.The associated pulmonary venolobar syndrome involves anomalous systemic arterial supply and anomlaous systemic venous drainage of the right lung. With the advent of MDCT imaging, we can diagnose this rare condition as well all its associated anomalies non-invasively. Volume-rendered techniques greatly simplify the complex anatomy and provide easy understanding of the same.

Key words: Horse-shoe lung; scimitar syndrome; volume rendered reconstructions

Introduction

Horse-shoe lung is a rare congenital anomaly usually associated with scimitar syndrome. Before the advent of multidetector CT imaging (MDCT), invasive techniques such as bronchoscopy and catheter angiography were used for the diagnosis. With the advent of newer MDCT scanners, this need has been obviated. A comprehensive diagnosis of complex congenital cardiovascular anomalies can be obtained in a single study which can delineate malformations of the heart, major vascular structures as well as lung and tracheo-bronchial tree. We report a case of horse-shoe lung associated with pulmonary venolobar syndrome identified on MDCT imaging. The case highlights the role of volume rendering techniques that can play a role in addition to the standard axial images for diagnosing and depicting the complex anatomy.

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Case Report

A 1-month-old male child initially presented with complaints of irritability and continuous crying. The child subsequently developed fever and chest infection. A chest radiograph done for the same revealed dextrocardia, and further evaluation by 2D echocardiography was done to look for congenital heart disease. The 2D echo findings showed dextroposed heart with lack of normal four pulmonary veins draining into the left atrium. A suspicion of scimitar syndrome was raised and further evaluation by CT scan was advised to assess the arterial supply and venous drainage of the right lung. CT imaging was preferred over MR angiography as there was a need to exclude lung sequestration besides delineating the pulmonary venous drainage.

CT pulmonary angiography was performed on a 64 detector GE Lightspeed VCT scanner using non-ionic contrast media and bolus-tracking method. Contrast was injected through an intraven intervenues (IV) line placed in the right hand as lower limb venous access was small and would limit the flow rate. The study was performed under short acting general anesthesia wherein 1.5 cc/kg of contrast was injected through a dual head pressure injector with a flow rate of 0.4 cc/s. Images were acquired on 64-detector CT scanner without ECG gating, using a slice thickness of 0.7 mm and collimation 0.6 mm, followed by overlapping reconstruction.

The acquired CT images were post-processed with focus on establishing the arterial supply to both lungs, the size and location of the main pulmonary artery (MPA), the right and left pulmonary arteries (RPA and LPA), and the venous drainage of both lungs. Volume-rendered (VR) reconstructions were done for depicting the pathology in 3D to aid understanding of the anomaly.

A scout topogram of the chest taken for planning of CT imaging showed the heart shadow in the right hemithorax [Figure 1]. Evaluation of CT images showed that the right lung was small in size with compensatory emphysema of the left lung [Figure 2C]. There was associated dextroposition of the heart with atrio-ventircular and ventriculo-arterial concordance.

All components of the pulmonary venolobar syndrome, also known as hypogentic lung or scimitar syndrome, were identified. These findings included systemic supply to the right lung base by two large collaterals arising from the upper abdominal aorta, one of which was a branch of the coeliac trunk [Figure 3]. These collaterals were seen supplying the basal segments of the right lower lobe. No collateral supply from the thoracic aorta was identified. There was no systemic collateral supply to the left lung. The right lung showed venous drainage to the suprahepatic inferior vena cava (IVC) through multiple small channels (at least three) [Figure 3]. In addition to these findings, there was also seen an anomalous tissue of lung extending in the retrocardiac region across the midline [Figure 2]. On standard axial images, both on mediastinal and lung windows, this lung tissue looked no different from the hyperinflated left lung tissue. However, on VR imaging with tracing of the main and branch pulmonary arteries and evaluation of the bronchi, peculiar findings of horseshoe lung were apparent [Figures 2 and 4]. These included a branch of the right pulmonary artery (RPA) extending across the midline to the left side. An associated bronchial supply also extending to the left was seen on VR imaging. These findings were reassessed on axial images and diagnosis of horseshoe lung was made in association with pulmonary venolobar syndrome. This highlights the importance of VR imaging in the evaluation of congenital anomalies. In this case, it helped in depicting the findings of a rare association of horseshoe lung with pulmonary venolobar or scimitar syndrome.

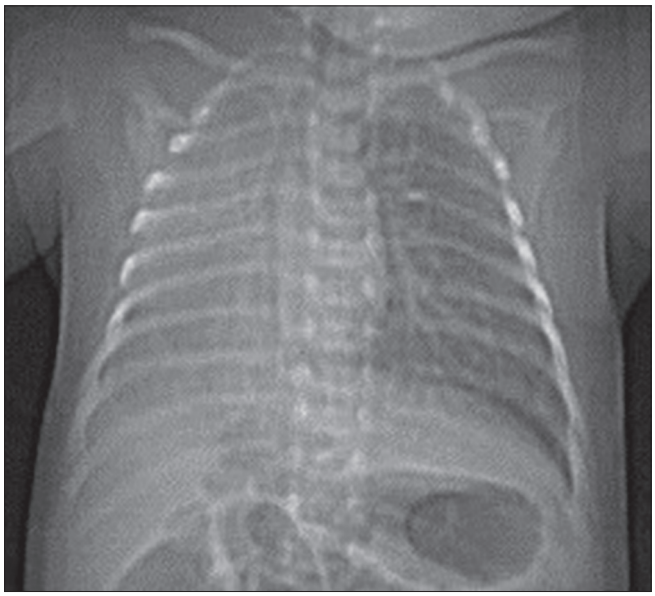

Figure 1: Scout topogram of the chest shows the heart shadow occupying the right hemithorax consistent with a dextroposed heart. It is difficult to identify dextrocardia versus dextroposition on the AP view

Discussion

The term horseshoe lung was first used by Spencer in 1962.[1] It is a rare congenital anomaly associated with a midline isthmus formed of lung tissue bridging the right and left lungs. Eighty percent of horseshoe lung cases are associated with scimitar syndrome, also known as the congenital pulmonary venolobar syndrome.[2] The scimitar syndrome is characterized by abnormal pulmonary venous return from a part of or the entire right lung to the inferior vena cava, anomalous arterial supply to the right lung base, and right lung hypoplasia. The diagnosis of horseshoe lung is confirmed with the finding of a bronchus arising from either right or left bronchus and crossing the midline to the contralateral side along with a corresponding branch pulmonary artery. The bronchial and arterial branches supplying the isthmus invariably arise from a hypoplastic pulmonary artery and bronchus, which supply the hypoplastic lung.[3,4] A controversy exists regarding the presence of intervening visceral pleura between the isthmus and the surrounding normal lung;[5-7] however, both pleural cavities always communicate through a common parietal defect.[4] Our case clearly showed the pleural line partly extending between the two, which was not traceable anteriorly, suggesting continuity between the pleural spaces. In a study by Frank *et al*., striking resemblance of clinical and radiographic features was found in patients

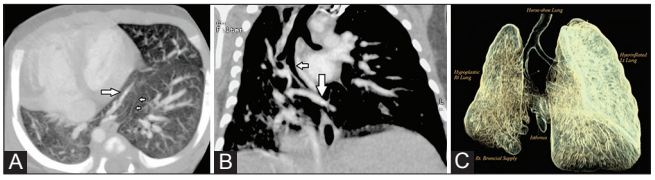

Figure 2 (A-C): (A) Axial image showing branch of the right pulmonary artery (long arrow) crossing the midline and supplying the isthmic tissue. Thin fibrous band (small arrows) was seen extending partially between the right and left lungs. (B) Coronal MPR image shows branch from right bronchus (horizontal arrow) supplying the isthmic tissue.Vertical arrow points to the arterial branch supplying the isthmus. (C) Volume-rendered image showing right lung hypoplasia with compensatory emphysema of the left lung. Branch from right bronchus supplying the isthmic tissue

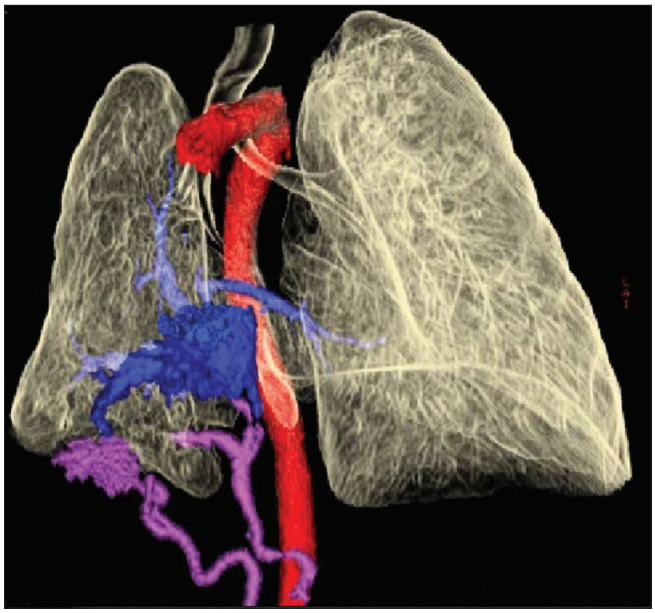

Figure 3: Systemic supply to the right lung base by two large collaterals (purple) from the upper abdominal aorta (aorta, red), one of which is a branch of the coeliac trunk, and right lung venous drainage to the suprahepatic IVC through multiple, at least three, channels (blue)

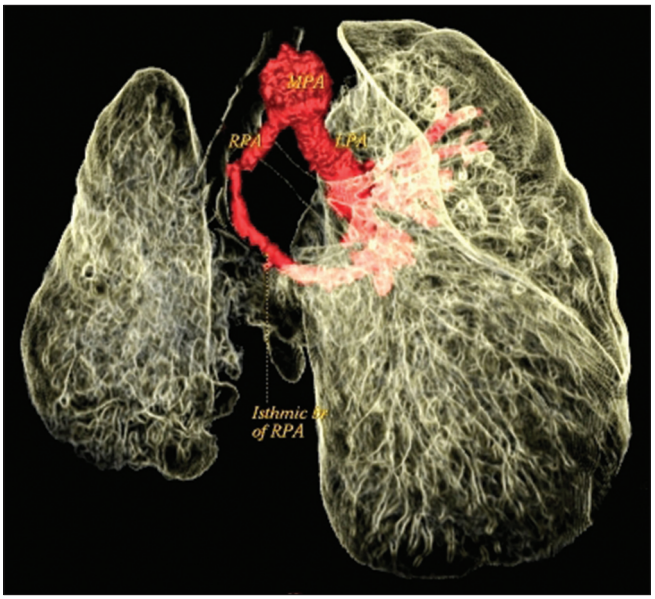

Figure 4: Volume-rendered imaging reveal findings of horseshoe lung with connecting isthmus and a branch of the right pulmonary artery coursing across the midline to the left to supply the pulmonary isthmus

presenting with scimitar syndrome with horseshoe lung.[7] All cases of horseshoe lung were under the age of 5 years and mostly below the age of 1 year. A plain chest radiograph demonstrated hypoplastic right lung in all cases with well-defined scimitar vein in 8 of the 11 cases.[7] The lung hypoplasia is recognized in the frontal projection by decreased volume of the right thorax, a mediastinal shift to the right, and a hazy appearance of the right cardiac border. Frank *et al*. also proposed that the finding of a linear density subtending a triangular or circular area in the medial apsect of the left lung base in scimitar syndrome may be a strong indication of coexisting horseshoe lung on frontal chest radiograph .[7]

Electron beam CT was used to clearly demonstrate the important features of horseshoe lung in 1997 by Takahashi *et al*. with free breathing in a 2-year-old child.[8] With the advent of MDCT scanners, excellent VR images can be generated to optimally depict the vascular distribution and eliminate the need for invasive imaging techniques such as angiography and bronchography. We evaluated the infant on 64-MDCT which could show the hypoplastic right lung; however, the scimitar vein was absent, instead there were multiple small veins draining into the IVC.

Features of scimitar syndrome in the form of anomalous pulmonary venous return from the right lung to IVC and anomalous systemic arterial supply from the abdominal aorta to the right lung base were seen in 11 of the 14 cases studied by Frank *et al*.[7] as well as in our case, which could be clearly demonstrated on CT images. There was also associated hypoplasia of the RPA. An abnormal lower branch of the RPA crossing the midline to the left was clearly seen on the VR images, confirming the presence of an isthmus connecting the right and left lungs. An abnormal bronchus arising from the right and extending toward the midline to the left was also apparent on VR images. The findings of abnormal arterial branch of RPA and a bronchial branch, both extending toward the left, and an obvious bridging isthmic lung tissue connecting the right and left lung bases confirmed the diagnosis of horseshoe lung and was demonstrated on the VRCT images. In our case, CT images were sufficient for confident diagnosis and no further invasive imaging, either catheter angiography or bronchography, was required. The case highlights the importance of VR imaging for comprehensive diagnosis of rare and complex congenital disorders. These anomalies once needed confirmation by invasive angiography or bronchography, but with the advent of MDCT, the need for invasive imaging may be completely abviated and VR images would be sufficient to depict the complex anatomy much more clearly than the conventional imaging techniques.

Conclusion

Horseshoe lung is a rare congenital anomaly, usually associated with scimitar syndrome. It can be demonstrated by MDCT imaging and the complex anatomy can be accurately demonstrated using VR techniques, thus obviating the need for more invasive conventional modes of diagnostic imaging such as catheter angiography or bronchography.

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