A rare case of horseshoe lung with scimitar syndrome and persistent left superior vena cava

Sir,

Horseshoe lung was first described by Spencer in 1962,[1] as a malformation in which an isthmus of pulmonary parenchyma extends from the right to the left lung bases across the midline posterior to the heart and anteriorly to the esophagus and spinal column. The term "horseshoe lung" was coined in analogy to "horseshoe kidney."[2] Nonseparation of the splanchnic mesoderm will cause persistence of connection between the two lungs and pleural cavities.[3] So far, about forty-odd such cases have been reported, largely associated with scimitar syndrome.[4] The resulting isthmus of pulmonary parenchyma receives the blood supply from the right pulmonary artery, and the respective bronchi, from the right bronchial tree. The isthmus presents its own pleural layer that may be incomplete, allowing communication between the right and left pleurae.[5]

A 31-year-old female presented with complaints of breathlessness for 10 days associated with exertional chest pain, anorexia, and vomiting. No other significant past history or complaints were noted. Physical examination reveals normal pulse rate and blood pressure, and on auscultation, loud P_2 was noted. Echocardiogram shows dilated right ventricle, pulmonary artery, and mild tricuspid regurgitation. The interatrial septum was intact. No other significant findings were noted. The patient was referred for contrast-enhanced computed tomography (CT) thorax for the evaluation of pulmonary hypertension.

A plain chest posteroanterior radiograph demonstrated the decreased volume of the right thorax, mild mediastinal shift to the right, and a hazy right cardiac border, all suggestive of hypoplastic right lung with well-defined scimitar vein [Figure 1a]. In the lateral projection, a linear oblique line that parallels the posterior margin of the sternum with increased density anterior to the line was noted [Figure 1b].

The findings of abnormal arterial branch of the right pulmonary artery 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 were demonstrated on the volume rendered CT(VRCT) images [Figures 2-4]. Incidentally, coronal butterfly vertebra was noted involving T11 vertebral body as coexisting segmentation anomaly.

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.[6-8] Association with other cardiovascular abnormalities, such as atrial septal defect, interventricular septal defect, persistent arteriosus ductus, and single left ventricle, may occur.[9] In patients with horseshoe lung, tracheal and bronchial stenosis and anomalous bronchial bifurcation are also reported.[10,11]

Congenital pulmonary venolobar syndrome is a constellation of different congenital anomalies of the thorax. Major components are hypogenetic lung (69%), partial anomalous pulmonary venous return (scimitar syndrome) (31%), pulmonary sequestration (24%), absence of the pulmonary artery (14%), systemic arterialization of the lung without sequestration (10%), absence or interruption of the inferior vena cava (7%), and duplication of the diaphragm (7%).[12]

Scimitar syndrome, also known as hypogenetic right lung, venolobar syndrome, Halasz syndrome, mirror image lung syndrome, epibronchial right pulmonary artery syndrome, or vena cava bronchovascular 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.[13] This vein presents an oblique caudal course, resembling the short, curved sword called scimitar, and in 15% of cases, it is associated with horseshoe lung.[4,6]

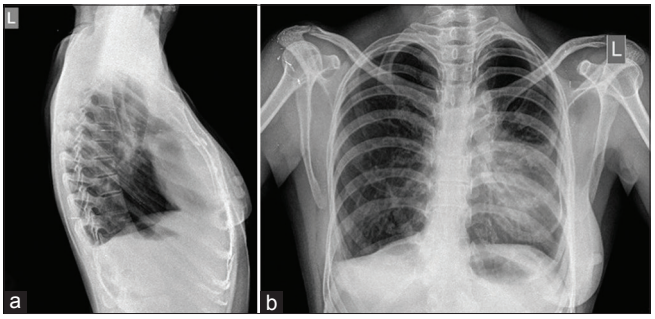

Figure 1: (a and b) Lateral and frontal (posteroanterior) chest radiographs showing obliteration of retrosternal space and increased retrocardiac clear space. Mild flattening of right hemidiaphragm is noted

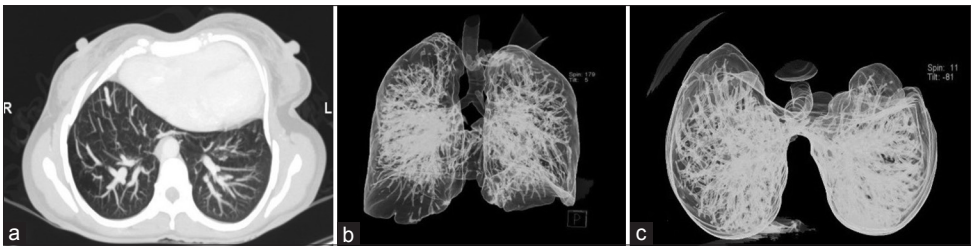

Figure 2: (a) Axial noncontrast computed tomography maximum intensity projection image showing the posterior fusion of lung segments. (b and c) Volume rendered computed tomography images demonstrating the fusion of the right and left lung confirming diagnosis of horseshoe lung

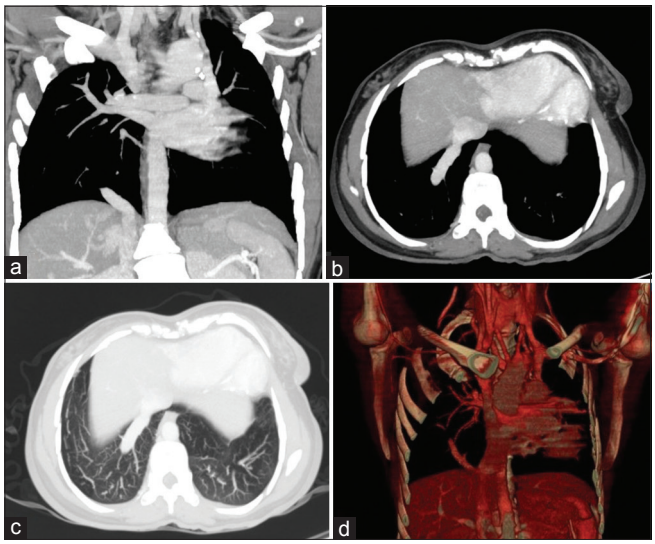

Figure 3: (a-c) Axial computed tomography with coronal reformation, (b) maximum intensity projection images and lung window, (c) anomalous right pulmonary vein draining into Inferior vena cava and persistent left superior vena cava, (d) coronal volume rendered computed tomography demonstrating partial anomalous pulmonary venous return with persistent left superior vena cava

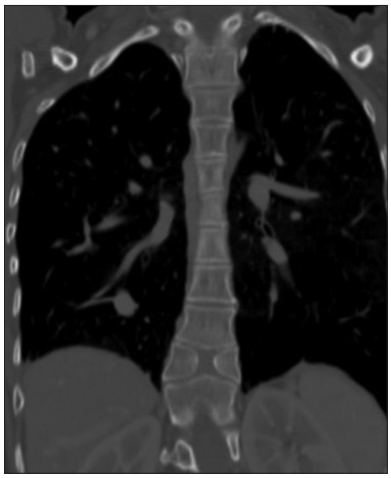

Figure 4: Coronal computed tomography reformation showing butterfly vertebra at T11 level in the same patient

In the lateral radiography, linear opacity can be identified due to extrapleural areolar connective tissue filling the space of absent lobe. Differentiation of scimitar syndrome from horseshoe lung can be made on the plain chest film, if, in addition to the typical radiographic findings of scimitar syndrome, there is evidence of a lucent area in the medial aspect of the left base demarcated laterally by a fine linear density. The isthmus of pulmonary tissue seen at the left base is often lucent, presumably because it is hypovascular compared to the adjacent left lower lobe. The presumption is made that intervening pleura is the linear density identifiable on plain films demarcating the lateral extent of the isthmus.[6]

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.[14,15] In the setting of this anomaly, the right lung is abnormal in terms of size, lobe, and bronchial tree.[16] The right pulmonary artery is usually abnormally hypoplastic, and in some cases, is totally absent; on the other hand, in 40% of cases, its size is almost normal.[17]

A controversy exists regarding the presence of intervening visceral pleura between the isthmus and the surrounding normal lung.[6,18] However, both pleural cavities always communicate through a common parietal defect.[15] The pleural membrane if visible on CT imaging will suggest pseudohorseshoe lung. Nonexistence of the pleural membrane is indicative of a true horseshoe lung malformation.[12,19,20] Pseudohorseshoe lung resulting from inflammatory changes such as necrotizing pneumonia or from destruction of pulmonary parenchyma, which leads to compensatory hypertrophy of the other lung and largely occurs in pediatric population and is just a basal segment herniation of the right pulmonary lobe.[19] It is not certain whether any entity such as congenital pseudohorseshoe lung exists despite reports.[20]

In our case, CT imaging was sufficient for diagnosis, and no further invasive imaging, either catheter angiography or bronchography, is required. The case highlights the importance of VRCT images for comprehensive diagnosis of such rare and complex congenital disorders. These anomalies needed confirmation in the past with invasive angiography or bronchography, but with the advent of multidetector CT (MDCT), the need

for invasive imaging may be completely obviated and VR images would be sufficient to depict the complex anatomy much more clearly than the conventional imaging techniques.[7]

The prognosis in cases of horseshoe lung is dependent on the pulmonary artery pressure. Usually, the types of horseshoe lung associated with normal pulmonary artery pressure are asymptomatic.[2] The two main indications for surgical intervention are large left-to-right shunt >2:1, resulting in pulmonary hypertension or heart failure, and recurrent pulmonary infection associated with sequestered lungs.[13,21,22]

In 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. Our case was unique in having multiple anomalies associated with horseshoe lung.

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Conflicts of interest
There are no conflicts of interest.

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