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A 38-Year-Old Woman with Scimitar Syndrome

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ABSTRACT

Scimitar syndrome is a rare clinical syndrome which consists of anomalous right pulmonary venous return to the inferior vena cava. Scimitar syndrome classically involves the right lung and is most commonly reported in very early infancy. However, it is occasionally seen in adults. We present a 38 year-old woman complaining of dry cough and exertional dyspnea during moderate exercise with right lung scimitar syndrome. The chest radiography showed increased radiolucency of the left lung and heart dextroposition with a characteristic appearance of scimitar sign which is a curvilinear density in the right middle and lower pulmonary fields resembling a curved Turkish sword. (Tanaffos2011; 10(3): 63-66)

Key words: Scimitar syndrome, Anomalous right pulmonary venous connection, Heart dextroposition

INTRODUCTION

The scimitar syndrome is a part of a congenital disorder called “partial anomalous pulmonary venous connection (PAPVC) (1). 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 a rare clinical syndrome which consists of anomalous right pulmonary venous return to the inferior vena cava (1-3). Other associated anomalies include hypoplasia of the right lung, heart dextroposition, small or absent right pulmonary artery, bronchial isomerism, cystic diverticula or

hypogenesis of the right bronchial tree, and abnormal systemic arterial supply from abdominal aorta or rarely descending thoracic aorta to the right lower lung and most commonly to the posterobasal segment of the lower lobe(3,4). Although scimitar syndrome classically involves the right lung, left-sided and bilateral scimitar syndrome have also been reported (5,6).

CASE SUMMARIES

A 38 year-old woman presented with dry cough of 3 months duration. She also complained of exertional dyspnea during moderate exercise which she had for about 3 years. She denied any fever, weight loss, chest pain, or wheezing. Her past medical history was unremarkable. On physical examination, she was an alert woman in no distress with normal vital signs. The jugular veins were not distended. The cardiac and lung examinations were unremarkable. She had

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no clubbing or cyanosis. The remainder of the physical examination was normal.

The chest radiography showed increased radiolucency of the left lung and right sided shifting of the heart shadow with a curvilinear density in the right middle and lower pulmonary fields (Figure 1).

As shown in Figure 2, chest CT scan revealed hypoplasia of the right lung with a small right pulmonary artery, and absence of right pulmonary venous return to the left atrium. Anomalous right pulmonary venous return to the infradiaphragmatic inferior vena cava was revealed in reconstructed images as shown in Figure 3.



Figure 1. Chest radiography showed increased radiolucency of the left lung, right sided shifting of heart shadow, and scimitar shadow parallel to right heart border.

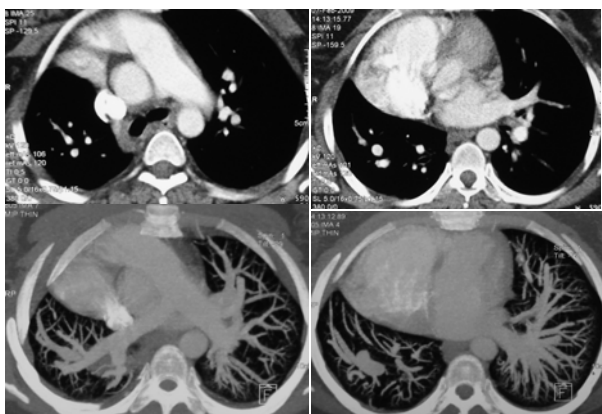


Figure 2. Chest CT scan with contrast and MIP images revealed a hypoplastic right lung, small right pulmonary artery, and absence of right pulmonary venous return to the left atrium.

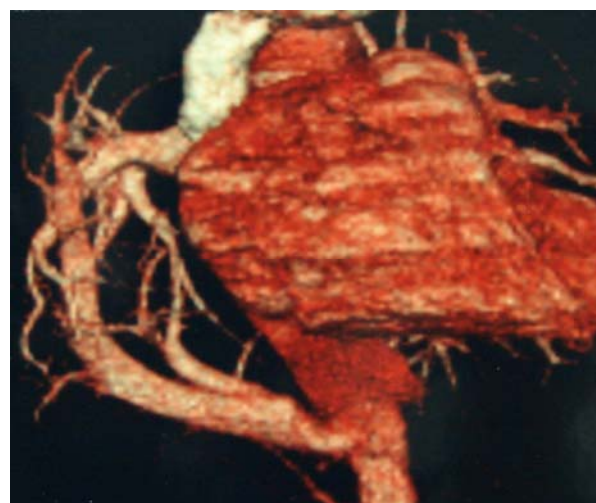
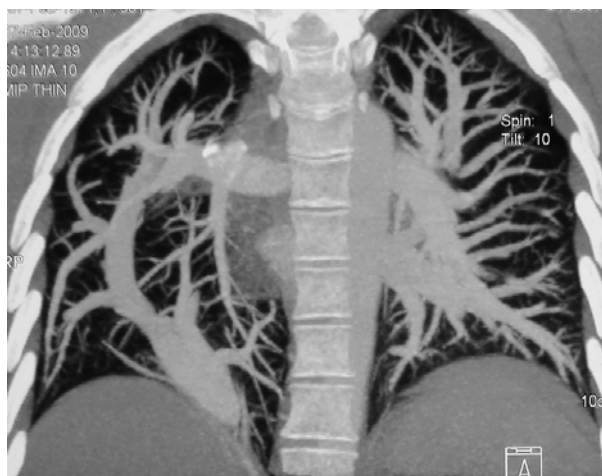


Figure 3. MIP and reconstructed CT images demonstrating anomalous right pulmonary venous return to infradiaphragmatic inferior vena cava.

DISCUSSION

Although this 38 year-old woman presented with minimal and nonspecific symptoms and signs, the radiographic findings were characteristic. Scimitar syndrome is identified on chest radiography by a characteristic appearance of scimitar sign which is a curvilinear shadow parallel to the right cardiac border that resembles a curved Turkish sword (7). A wide range of coexisting congenital cardiac abnormalities can also occur, most commonly secundum type atrial septal defect (8). Another common associated anomaly is horseshoe lung which is fusion of the posterior basal segment of the lungs behind the pericardial reflection (6).

Scimitar syndrome is most commonly reported in very early infancy with a mean age of seven months, but is occasionally seen in adults (9). The infantile form that is diagnosed in cases under one year of age, is associated with a variety of thoracic abnormality and vascular malformations (1,10). The infantile form is associated with respiratory failure, recurrent wheezing, coughing, failure to thrive, and recurrent pulmonary infections (1,11,12). Symptoms of cardiac failure and pulmonary hypertension may develop with a large left-to-right shunt (12,13). The childhood/adult form is associated with a smaller shunt and a less severe pulmonary hypertension (1). This group is characterized by minimal symptoms as in our present case and lack of coexisting anomalies (10). Although, some patients present with recurrent infection, hemoptysis, and hemothorax, the majority are identified on routine chest radiography (7,8). However, other imaging techniques including Doppler US, conventional X-ray angiography, transthoracic Doppler ECHO, computed tomography, Cine MRI and gadolinium enhanced 3-D MR angiography are necessary to confirm the diagnosis or visualize the associated anomalies (4,7,10). In patients who are asymptomatic or have minor symptoms with small left-to-right shunt and normal

pulmonary artery pressure, conservative management and close observation are indicated (4,7,8). The two main indications for surgical intervention are large left-to-right shunt greater than 2:1 resulting in pulmonary hypertension or heart failure, and recurrent pulmonary infection associated with sequestered lungs (1,10,13).

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