Clinical Quiz

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A rare cause of recurrent dyspnea

Clinical Presentation

A 58-year-old man, who had been a chronic smoker for 25 years, presented with gradually progressing exertional breathlessness of 3 years duration. He has had recurrent episodes of productive cough and fever, which was diagnosed as chronic obstructive airway disease and treated symptomatically in various hospitals for 5 years. **Figure 1** shows his chest radiograph in posteroanterior view. **Figure 2** shows his chest computed tomogram image.

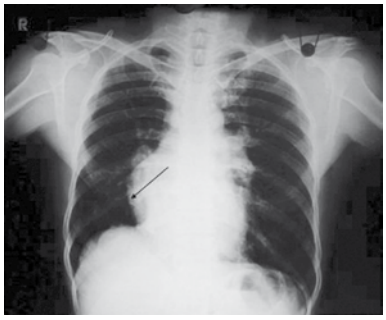

Figure 1 - Chest radiograph in posteroanterior view, showing an opacity in the right para cardiac region.

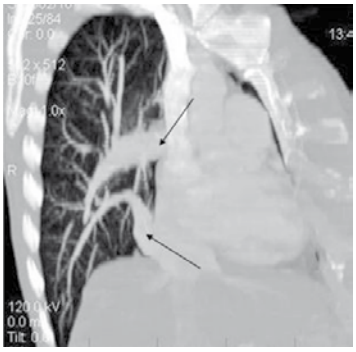

Figure 2 - Computed tomogram image showing right upper lobe pulmonary vein opening into the superior venacava and the right lower lobe pulmonary veins draining into the infra diaphragmatic inferior venacava.

Questions

1. Describe the chest radiograph and the CT image.
2. What is your diagnosis? What other defect do you expect along with this anomaly?
3. What further test would you like to do to confirm the findings?

Clinical Quiz

Answers

1. The chest radiograph in the posteroanterior view shows opacity in the right paracardiac region. The right heart border is sharply defined and clearly visible throughout its entire length. The cardiac axis was rotated to the right, suggesting a possible cardiac anomaly. The CT image shows the right upper lobe pulmonary vein opening into the superior venacava (SVC) and the right lower lobe pulmonary veins draining into the infra diaphragmatic inferior venacava (IVC).

2. The diagnosis is partial anomalous pulmonary venous connection (PAPVC) or Scimitar syndrome. In the above radiography the scimitar vein was obscured by the right heart border, mimicking a middle lobe collapse. Partial anomalous pulmonary venous connection are usually associated with atrial septal defects (ASD).

3. A trans-thoracic echocardiogram followed by cardiac catheterization needs to be carried out in order to demonstrate the ASD.

Discussion

First described by Chassinat in 1836,[1] this is a complex constellation of cardio-pulmonary anomalies. In its complete form, the Scimitar syndrome consists of partial or total anomalous pulmonary venous drainage of the right lung into the IVC, hypoplasia of the right lung, dextro-rotation of the heart and hypoplasia or other malformation of the right pulmonary artery. Cardiac abnormalities such as interatrial septal defect and patent ductus arteriosus are also associated.[2] Scimitar syndrome is usually right-sided; however, rare cases have been reported that involved the left side.[3] It shows the abnormal vein draining into the inferior vena cava as a curved vascular shadow with a lateral convexity in the right lower zone, called the Scimitar sign. Three forms of scimitar syndrome have been described on the basis of age at presentation.[4] These are the infantile form, the adult form and a rare third form characterized by additional cardiac and extra cardiac malformations.

In the reported chest roentgenogram, the Scimitar vein was obscured by the heart shadow, which created problems in the diagnosis. The CT imaging showed an anomalous connection with the right pulmonary vein from the upper lobe, draining into the SVC and a vein from the lower lobe draining into the IVC. Partial anomalous pulmonary venous connection (PAPVC) of the right lung is usually associated with an ASD and an intact atrial septum is exceptional. There are a few reports of PAPVC without ASD in the literature.[5]

References

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