Scimitar and Dyspnea

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Introduction: Scimitar syndrome, also known as hypogenetic lung syndrome or pulmonary venolobar syndrome, is a partial anomalous pulmonary venous connection of the right lung to the inferior vena cava (IVC) or to the portal or hepatic veins. Annual incidence is 1 to 3 in 100,000 live births, with a 2:1 female predominance. The affected lung and airway are often hypoplastic. Case: 29-year-old female with Scimitar syndrome diagnosed during infancy, was exhibiting mild chronic dyspnea without functional limitations. Pulmonary function testing showed total lung capacity 70% of predicted, restrictive spirometry and normal diffusing capacity. Computed tomography (CT) of the chest showed dextrocardia, congenital absence of the right pulmonary artery and right pulmonary veins, anomalous venous drainage of the right lung into the superior vena cava via the azygous vein and anomalous arterial feeder emanating from the abdominal aorta supplying the right lower lobe. There was complete agenesis of the right upper and middle lobes with normal appearance of the left lung and associated hyperexpansion. Echocardiogram had limited windows without ability to visualize the right and left ventricles, which were described on CT as dilated with a small possible ventricular septal defect (VSD). The patient had undergone surveillance imaging throughout childhood. Discussion: In Scimitar syndrome, the affected lung is small and may be associated with a small or absent pulmonary artery and systemic arterial supply. Many cardiovascular and pulmonary anomalies, such as bronchopulmonary sequestration, pulmonary vein stenosis, dextrocardia, may be associated with this syndrome. Patients are either diagnosed incidentally in infancy or may present with fatigue, dyspnea, pulmonary hypertension, recurrent pneumonia, hemoptysis etc. Hemodynamic abnormalities usually develop when more than half of the pulmonary blood flow is recirculated back to the pulmonary vasculature. Diagnosis is mostly made via imaging. Chest X-ray may show the shadow of the descending pulmonary vein, hypogenetic lung and dextrocardia. Diagnostic work up includes getting an echocardiogram as well as Magnetic Resonance Imaging (MRI) or CT Angiography of the chest to better characterize the vasculature, bronchial tree and cardiac chambers. Cardiac catheterization helps determine pulmonary vascular resistance, the degree of left-to-right shunt, and better characterize anatomical anomalies. Corrective surgery may be performed safely, but patients requiring lobectomy or pneumonectomy may have higher morbidity and mortality.
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