

IMAGES IN CLINICAL RADIOLOGY



Isolated unilateral absence of the right pulmonary artery

B.S. Smet^{1,2}, A. De Backer¹, F. Cottenie¹, D. Dewilde¹, F. Feyaerts¹

A 40-year-old man was referred for evaluation of cough of a few weeks duration and a history of recurrent respiratory tract infections for several years. Clinical examination revealed no abnormalities. Routine hematologic and biochemical evaluation were normal. Contrast-enhanced computed tomography of the thorax showed a hypoplastic right lung, hyperinflation of the left lung with cardiomeastinal shift to the right. Absence of the right pulmonary artery was noted and replaced by an extensive collateral network of hypertrophied vessels originating from bronchial, intercostal and mammaria interna arteries, and right arteria subclavia (Fig. A, B). Discrete bronchiectasis with thickened bronchial walls in a hypoplastic right lung was noted (Fig. C). A normal bronchial tree and normal parenchyma in the left lung was seen.

In our patient symptoms were mild and treated successfully with pulmonary rehabilitation.

Comment

Unilateral absence of a pulmonary artery (UAPA) is a very rare condition first described in 1868. Its prevalence has been estimated to be 1 in 200 000 persons. UAPA is caused by a congenital anomaly due to failure in the connection of the sixth aortic arch and the pulmonary trunk. UAPA is usually associated with other vascular abnormalities such as tetralogy of Fallot, right aortic arch, septal defects and patent ductus arteriosus and, as a consequence, diagnosis is made in early childhood. Less frequently UAPA occurs as an isolated finding and is detected later in life with a median age of 14 year, found by Ten Harkel et al. in a population of 108 patients with isolated UAPA, ranging from 0,1 to 58 years. Most patients have no or minimal symptoms consisting of mild dyspnoe on exertion, recurrent pulmonary infections and related manifestations of infection such as cough and pleural effusion. However, some patients may present with hemoptysis, congestive heart failure or pulmonary hypertension.

Imaging findings consist of a smaller hemithorax, ipsilateral hemidiaphragm elevation with ipsilateral cardiac and mediastinal shift and absence of the ipsilateral pulmonary artery. Hyperlucency and hyperinflation of the contralateral lung is seen. Bronchiectasis in the hypoplastic lung reflects the impact of unilateral absence of a pulmonary artery on the mucosal defense of the lung. Differential diagnosis has to be made with Swyer-James-MacLeod's syndrome or thrombo-embolic disease. Computed tomography and magnetic resonance imaging may accurately demonstrate the absent pulmonary artery and its compensatory collateral systemic vascularisation.

A therapeutic intervention may be considered in patients when serious complications occurs. In patients with hemoptysis, congestive heart failure or pulmonary hypertension endovascular or surgical occlusion of the collateral vessels, pneumectomy or even heart-lung transplant may be performed. Conservative therapy is considered when symptoms are mild.

Reference

1. Ten Harkel A.D., Blom N.A., Ottenkamp J.: Isolated unilateral absence of pulmonary artery. *Chest*, 2002, 122: 1471-1477.

1. Department of Radiology, General Hospital Sint-Lucas, Ghent, 2. Department of Radiology, University Hospital Ghent, Ghent, Belgium.