Pulmonary arteriovenous malformation

E. Janssens, P. Leyman, T. De Beule, L. Ardies, P. D’haenens

A 37-year old patient with Rendu-Osler-Weber syndrome presented to our hospital with hypoxia.

Chest radiography shows on the PA view (Fig. A, arrow) a sharply margined branching lobular mass of uniform density in the right upper/lower lobe.

A multislice spiral contrast-enhanced CT scan of the thorax shows multiple conglomerations of lobulated contrast-filled vessels (Fig. B, arrows) with feeding artery and draining vein.

Anterior view of 3D volume-rendered image (Fig. C) displays the largest lesion in the right lower lobe with 2 feeding arteries (arrows) one draining vein (arrowhead). The lesion is 4.6 cm in diameter with 2 feeding arteries of 0.7 and 0.9 mm and one draining vein of 10 mm.

Based on the imaging features and the clinical symptoms, the diagnosis of pulmonary arteriovenous malformations (AVMs) was made.

Comment

Pulmonary AVM, is a condition in which there is an abnormal direct communication between pulmonary arteries and pulmonary veins. The most common cause of pulmonary AVM is congenital.

Acquired pulmonary AVM is seen in patients with prior congenital cyanotic heart surgeries, liver disorders (hepatopulmonary syndrome), systemic diseases and venous anomalies.

10% of the cases are identified in infancy or childhood with male predominance in newborns.

Pulmonary AVMs are uncommon lesions, but are the cause of considerable morbidity and occasional mortality.

Patients become symptomatic at the age of 40-60 years. Epistaxis and hemoptysis are common clinical features. Pulmonary AVMs with high flow or anatomic right-to-left shunt may result in cyanosis, digital clubbing, congestive heart failure and paradoxic embolism with substantial risk for stroke and cerebral abscess.

A pulmonary AVM with a diameter less then 2 cm usually does not cause any symptoms.

Rendu-Osler-Weber syndrome, is an autosomal dominant disorder and is frequently associated with pulmonary AVM (35%). For this reason, family members with Rendu-Osler-Weber should be screened for pulmonary AVMs.

Contrast pulmonary angiography remains the gold standard in the diagnosis of pulmonary AVM, and is usually necessary if resectional or oblitative therapy is being considered.

CT is the method for choice for routine detection of pulmonary AVMs, for diagnosis in patients unable to undergo conventional angiography, for follow-up of patients with proven pulmonary AVM and in confirming the presence of residual AVMs at follow-up after embolotherapy.

All symptomatic pulmonary AVMs, pulmonary AVMs larger than 2 cm in diameter and pulmonary AVMs with feeding arteries larger than 3 mm should be treated with embolotherapy or surgery. The decision to perform surgical excision or transcatheter embolization planning rests on reaching an accurate estimate of the number, size, and course of vessels.

1. Department of Radiology, Onze-Lieve-Vrouweziekenhuis, Aalst, Belgium.