Pulmonary arteriovenous malformations (PAVMs) are abnormal direct communications between pulmonary arteries and pulmonary veins. These abnormal communications result in an anatomical right-to-left shunt that reduces the arterial oxygen saturation and may cause hypoxaemia and dyspnoea. Although PAVMs frequently remain undiagnosed, they are associated with severe morbidity in the form of ischaemic strokes and brain abscesses. We report a case of incidental CT angiography depiction of a PAVM within a pulmonary cyst. To the best of our knowledge, no such case has been described previously. On the basis of its appearance and lack of typical clinical features of hereditary haemorrhagic telangiectasia (HHT), we suggest that this PAVM actually represents an acquired fistula from a previously unrecognised focal pulmonary insult, such as trauma or infection, that simultaneously evolved into a pulmonary arteriovenous fistula (PAVF) within a traumatic pulmonary cyst or pneumatocele.

Received 26 April 2009
Revised 12 May 2009
Accepted 22 June 2009
DOI: 10.1259/bjr/39651947
© 2010 The British Institute of Radiology
between pulmonary arteries and veins may also be found in a variety of acquired conditions, and in these cases the correct nomenclature is PAVF. These acquired conditions include hepatic cirrhosis, and less commonly schistosomiasis, mitral stenosis, trauma, actinomycosis, Fanconi’s syndrome and metastatic thyroid carcinoma [5–9].

PAVMs may be single or multiple in occurrence, with the incidence of single PAVMs ranging from 42% to 74% of cases in different series. As in our case, most solitary PAVMs are seen in close proximity to the visceral pleura or embedded in the outer third of lung parenchyma in bilateral lower lobes. The left lower lobe is the most common location, followed by right lower lobe, left upper lobe, right middle lobe and right upper lobe [10].

Most PAVMs remain asymptomatic and do not present until the fourth decade of life [11]. However, PAVMs can be associated with a variety of complications, some of which are life-threatening. These life-threatening conditions include massive haemoptysis, haemothorax, stroke and brain abscess [12–14]. The incidence of neurological complications in general varied in different series from 19% to 59%, with a reported incidence of stroke and brain abscess of 18% and 9%, respectively [10].

The accepted radiological classification of PAVMs is determined mainly by the number of arterial pedicles to be occluded by the angiographer, whatever the number of draining veins. Therefore, simple-type PAVMs are defined by a single artery feeding into an aneurysmal communication (i.e. a malformation that is treatable with the occlusion of a single arterial pedicle). By contrast, complex-type PAVMs are characterised by more than one pulmonary arterial branch connecting into the aneurysmal communication (i.e. a malformation treatable with the occlusion of several arterial pedicles) [15].

Case report: CTA of pulmonary arteriovenous fistula within a pulmonary cyst

Figure 1. Anteroproximal portable chest radiograph demonstrates a left lower lobe oval hyperlucency (*), which is engulfed by two soft-tissue-density tubular structures, representing a feeding artery (dotted arrow) and a draining vein (solid arrow). These tubular structures converge with the left hilar region medially and with each other inferolaterally (curved arrow).

Figure 2. Axial non-enhanced CT images with (a) mediastinal window and (b) lung window settings at the level of the left lung base. Note a thin-walled pulmonary cystic lesion measuring 5.8 cm in the left lower lobe (*). This cystic structure contains multiple round or tubular structures, that have a soft-tissue density, adherent to its internal walls (arrows). Also of note, there is a large cystic structure of simple-fluid density within the right lobe of the liver. This is an incidental finding not related to the findings described.
Figure 3. Coronal multiplanar reformatted images of the CT pulmonary angiography study, using (a) soft tissue and (b) lung window settings through the previously described left lower lobe cystic lesion. Note the enhancing upper and inferior internal tubular structures, representing the feeding artery (dotted arrow) and a draining vein (solid arrow), which converge with each other inferolaterally (curved arrow) to create a pulmonary arteriovenous fistula.

Figure 4. Coronal multiplanar reformatted images of the CT pulmonary angiography study, using (a) soft tissue and (b) lung window settings at the plane of the descending thoracic aorta. Note the enhancing feeding artery (dotted arrow) and draining vein (solid arrow), which converge with the left hilar region medially.

Figure 5. Volume-rendered reformatted image of the CT pulmonary angiography study from (a) anterior and (b) posterior projections. Note the pulmonary arteriovenous fistula (curved arrow), the feeding artery (dotted arrow) and the draining vein (solid arrow).
There is no published association between PAVMs and pulmonary cysts. Further, there is no known association between congenital cystic lung lesions, such as congenital adenomatoid malformation, and PAVMs [16].

To the best of our knowledge, the depiction and analysis of a PAVF within a pulmonary cyst by multidetector CT (MDCT) angiography has not been reported in the medical literature. The occurrence of PAVF within a pulmonary cyst is, by itself, important for patient management as this pulmonary lesion is not a recognised manifestation of HHT. As a genetic disorder, HHT is by definition inherited in an autosomal dominant manner, and is the most common cause of PAVM. On the basis of this observation and a short clinical assessment, clinicians were able to exclude HHT from the differential diagnosis, therefore eliminating the need for long unnecessary evaluation of patient’s first-degree family members for possible asymptomatic HHT. In our opinion, the coexistence of PAVF within a pulmonary cyst suggests a prior unrecognised focal pulmonary insult, such as a trauma or infection, that simultaneously evolved into a PAVF within a traumatic pulmonary cyst or pneumatocele.

This case also illustrates the capability of MDCT angiography, with its inherent increased spatial and contrast resolutions, in the detailed analysis of complex vascular anomalies within the lung parenchyma. By delineating one feeding pulmonary artery, one draining pulmonary vein and the abnormal communication between the two, a diagnosis of a simple PAVF was established, making this lesion amenable to transcatheter embolotherapy as a definitive treatment [17]. Because the diameter of the feeding segmental pulmonary artery was larger than 3 mm, treatment with transcatheter embolotherapy would have been a reasonable approach because of the increased risk of neurological complications associated with the PAVF. Nonetheless, in this particular case, the lesion was an incidental finding that did not seem to contribute to the patient’s unstable angina pectoris. The triage of treatment for the diagnosed solitary PAVF was postponed until convalescence from the CABG surgery.

References


