Eur Respir J 1998; 12: 989–992
DOI: 10.1183/09031936.98.12040989
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European Respiratory Journal
ISSN 0903 - 1936

CASE STUDY

Haemoptysis associated with pulmonary varices: demonstration using computed tomographic angiography

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Haemoptysis associated with pulmonary varices: demonstration using computed tomographic angiography. G.R. Ferretti, F. Arbib, B. Bertrand, M. Coulomb. ©ERS Journals Ltd 1998.

ABSTRACT: A 67 yr old female with mitral stenosis presented with an acute haemoptysis caused by the rupture of pulmonary varices. Chest radiography and bronchoscopy showed nonspecific abnormalities. The diagnosis of this rare but potentially lethal complication was made using computed tomographic angiography with three-dimensional volume rendering.

Eur Respir J 1998; 12: 989–992.

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Keywords: Computed tomography, haemoptysis, pulmonary veins, three dimensional imaging

Received: November 20 1997
Accepted after revision February 13 1998

Pulmonary varices are rare abnormalities of pulmonary veins that may exist either as an isolated malformation or in association with pulmonary venous hypertension. Pulmonary varices are usually asymptomatic and diagnosed on chest radiography [1]. Rupture and systemic embolization are uncommon, but potentially severe complications [1]. Computed tomography (CT) has been shown to be a valuable technique in demonstrating this abnormality [2].

The case is reported of a patient who presented with haemoptysis due to rupture of pulmonary varices in whom contrast enhanced spiral CT with three-dimensional (3D) volumetric rendering demonstrated the malformation.

Case report

A 67 yr old female was admitted to hospital for the evaluation of acute haemoptysis. Her past medical history was remarkable for systemic hypertension, smoking (47 pack-yrs), chronic obstructive pulmonary disease (COPD) and mitral stenosis secondary to rheumatic heart disease. She had had one episode of haemoptysis, coughing up about one glass of bright red blood. She had been taking 500 mg aspirin for 2 days before the haemoptysis because of back pain. The patient had no fever or chest pain but complained of dyspnoea. Physical examination revealed the typical mid-diastolic murmur of mitral stenosis (2/6). The blood pressure was 140/80 mmHg. Laboratory examinations disclosed a haemoglobin level of 121 $g \cdot L^{-1}$ and a haematocrit of 35%. Arterial blood gas studies on room air were pH 7.39, oxygen tension (PO_2) 10.4 kPa (78 mmHg) and carbon dioxide tension (PCO_2) 4.48 kPa (34 mmHg).

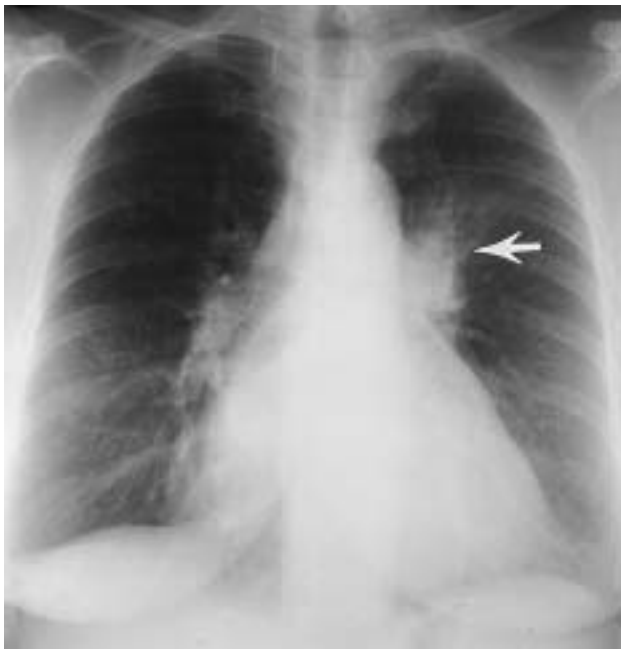
Chest radiography showed parenchymal air space consolidation in the left upper lobe (arrow) and prominent left atrium appendage (fig. 1a and b). Fibreoptic bronchoscopy demonstrated a blood clot in the anterior segmental bronchus of the left upper lobe, without any associated abnormality of the bronchial tree.

Spiral CT was conducted with a GE hiSpeed Advantage CT scanner (GEMS, Milwaukee, WI, USA) with 3 mm slice collimation and 1:1 pitch, during a single breath-hold acquisition. Contrast material (100 mL, 300 $mg \cdot mL^{-1}$) was injected with a power injector into an antecubital vein at the rate of 3 $mL \cdot s^{-1}$. Injection was started 20 s before CT acquisition. Axial imaging demonstrated tortuous vascular structures within the area of parenchyma air-space consolidation (fig. 2). However, it was difficult to analyse precisely the origin of this vascular malformation on axial images. Therefore, 3D images were reconstructed from the spiral CT data set (2 mm intervals, 20 cm field of view) using a real-time volume-rendering technique (work in progress software, GEMS) on an Advantage Windows workstation (Spark 20, Sun Microsystems, Mountain View, CA, USA). A voxel intensity histogram of the CT data set was first created. Image display parameters *i.e.* opacity, brightness, level and window, were chosen interactively to emphasize structures containing contrast material and deemphasize air-containing voxels [3]. A lateral display ofered an overview of the vascular malformation, demonstrating two tortuous vessels draining into the left upper pulmonary vein (fig. 3).

Digital subtraction angiography was performed and disclosed the final diagnosis of pulmonary varices, based on a normal pulmonary artery angiogram, no capillary shunting, and delayed opacification of the dilated veins in the venous phrase (fig. 4a and b). The left upper pulmonary vein entered normally into the left atrium.

Transthoracic echocardiography revealed a stenosed mitral valve (1.3 cm^2 on planimentry) without valvular calcification and a mean mitral valve gradient of 7 mmHg. Because

a)



b)

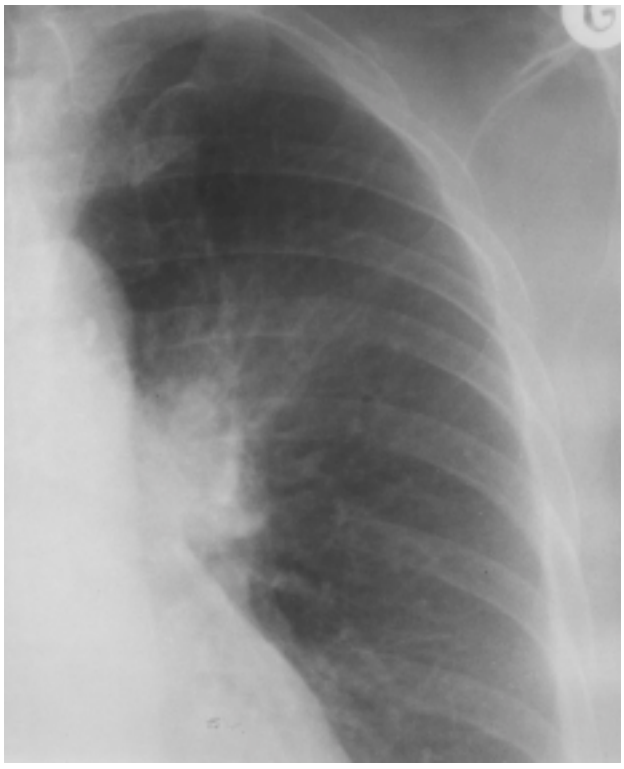


Fig. 1. – a) Posteroanterior chest radiograph showing the left upper lobe pulmonary infiltrate (arrow) and b) close-up of the left upper lobe.

the patient had mitral valve stenosis without mitral valve insufficiency, percutaneous balloon valvuloplasty was performed. The mitral valve area increased from 1.4 to 2.3 cm^2 and the mean mitral valve gradient decreased from 7 mmHg to 2.2 mmHg after dilatation. The haemoptysis did not recur. A spiral CT scan 2 months following mitral balloon valvuloplasty disclosed a decrease in the size of the pulmonary varices.

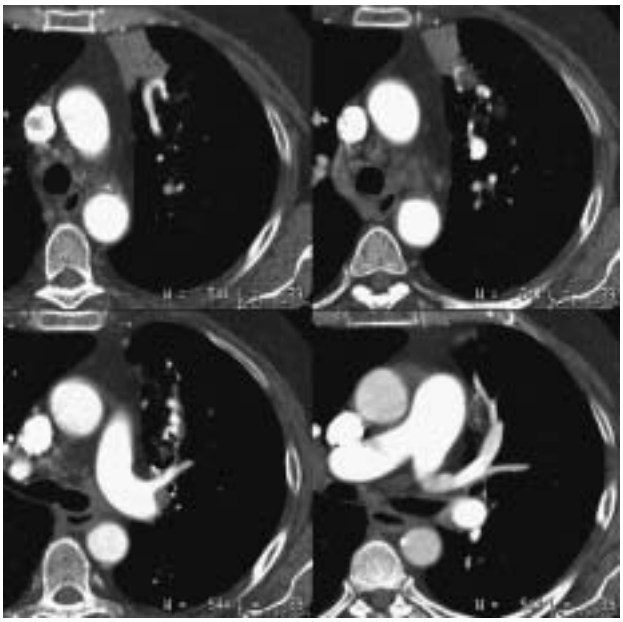


Fig. 2. – Four slices of contrast-enhanced spiral computed tomographic acquisition through the left upper lobe showing tortuous vessels within the anterior segment.

Discussion

The origin of pulmonary varices is unclear and may be congenital or acquired [1]. Acquired factors such as pulmonary venous hypertension secondary to mitral regurgitation or, less commonly, mitral stenosis are associated with pulmonary varices in about 50% of cases and may reveal congenital weakness of the wall of the veins [4]. Decrease in venous pressure after surgical treatment of mitral valve disease has been associated, in some cases, with regression of the varices [4, 5]. In the present patient, a decrease in pulmonary venous hypertension after mitral valvulotomy was associated with a decrease in the size of pulmonary varices, but the abnormality persisted.

Pulmonary varices are uncommon abnormalities that are usually asymptomatic and discovered incidentally on a chest radiograph [1]. Previous studies have shown that pulmonary varices are mostly localized in the right lower lobe (60% of cases), left upper lobe (17%) and right upper lobe (8.5%) [6]. Acute or chronic haemoptysis has been described only rarely in the past and may be fatal owing to variceal rupture into a bronchus or the pleural space [1]. In the patient in this study, haemoptysis could have been favoured by pulmonary venous hypertension, acute bronchitis and the ingestion of aspirin. Other rare clinical manifestations include dysphagia or middle lobe syndrome secondary to extrinsic compression [1], systemic emboli and rapid increase in size. According to the classification of Uyama *et al*. [6], the patient had a tortuous type of pulmonary varices, which is more frequently described in the right lower lobe and is often associated with mitral valve disease.

A diagnosis of pulmonary varices is usually obvious on the chest radiograph, when the mass presents as one or more oval or round, well-defined opacities within the medial third of the lung without any modification in size on

Fig. 3. – Volume-rendered image in lateral orientation demonstrating complex vascular malformation (arrows) draining into the left superior pulmonary vein (arrowhead).

follow-up [1]. The sensitivity and specificity of chest radio-graphy for the pulmonary varices have not been addre-ssed as the malformation is usually asymptomatic. The plain film differential diagnosis of centrally located round or lobulated mass includes all masses in the lung [1]. The plain film differential diagnosis of serpiginous tubular den-sities within lung parenchyma includes pulmonary arteriovenous fistulae and partial anomalous pulmonary venous return [1, 7]. Chest radiography with Valsalva or Mueller manoeuvres may increase the size of pulmonary varices. However, as chest radiography is limited in assessing the differential diagnosis, contrast-enchanced CT is usually performed and allows the vascular nature of the mass to be recognized [2, 7]. In the present case, the chest radiograph did not suggest the diagnosis because the varices were masked by alveolar haemorrhage. Fibreoptic bronchoscopy revealed a blood clot in the anterior bronchus of the left upper lobe but did not suggest the diagnosis. Contrast-enchanced CT allows for noninvasive diagnosis of pulmonary varices when it shows an enlarged vein draining into the left atrium [2]. However, axial CT images are sometimes difficult to analyse because the malformation is complex and depicted on several contiguous slices. In such cases, spiral CT with 3D reconstructions offers the opportunity of clarifying the anatomy. In this case, 3D volume rendering, a new technique for generating 3D im-ages from spiral CT data sets [8, 9], was used. Volume rendering has the advantage over maximum intensity projection (MIP) algorithms and 3D surface-shaded displays as it allows the display of the entire CT data set into 3D images. Volume rendering maintains the correct anatomical spatial relationships between displayed structures, unlike MIP images. Unlike surface-shaded display, volume rendering does not necessitate binary classification of the CT data set and, therefore, relative voxel attenuation is preserved in the final images. In this case, volume-rendered images offered an accurate representation of the venous malformation as demonstrated by comparison with angiographia images.

KUSZYK *et al*. [10] found the 3D volume-rendering technique useful in evaluating pulmonary artery and arteriovenous malformations. The case shows that this technique can also be applied to demonstrate pulmonary venous malormations. Although CT angiography with 3D volume rendering led to the suspicion of pulmonary varices, the definite diagnosis and the exclusion of pulmonary arteriovenous malformation required selective angiography in this particular case because CT angiography lacked dynamic information. However, dynamic information could have been obtained by using continuous CT acquisition at a single level through the malformation with contrast injected [11]. A pulmonary varix adjacent to the heart was diagnosed using transoesophageal echocardiography by TOHER *et al*. [12]. Echocardiography displayed the anatomy of the varix, its connection to the left atrium with

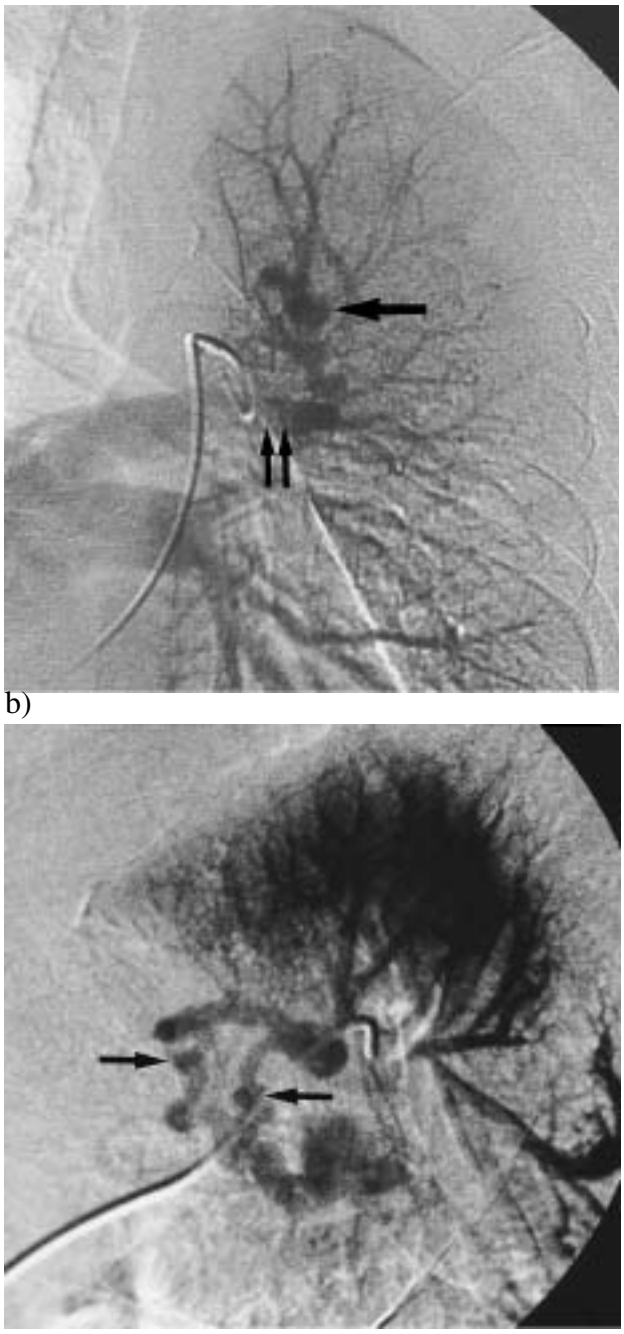


Fig. 4. – a) Venous phase of digital substraction pulmonary angiography in a frontal projection, showing left upper lobe serpiginous varices (arrow) draining into the left upper lobe pulmonary vein (double arrow). b) digital substraction pulmonary angiography in a lateral projection. Selective catheterism of the anterior segmental artery of the left upper lobe. During the venous phase of the injection, opacification of two tortuous veins occurred (arrows), draining into the left superior vein.

normally directed, low-velocity flow by pulsed and colour Doppler and the absence of contrast enchancement of the varix after *i.v.* bolus injection of microsonicated dextrose. In the patient studied here, because of the location of the pulmonary varices within the lung parenchyma with air interposition between the heart and the varices, transoesophageal echocardiography was not able to demonstrate the varices. To the authors knowledge, magnetic resonance angiography has not been used to diagnose pulmonary varices. However, in combination with gradient-echo imaging or contrast injection, this technique certainly has the potential to demonstrate such malformations because of the pulmonary varix [13].

In summary, this case demonstrates that spiral computed tomography with three-dimensional reconstruction has the potential to demonstrate complex pulmonary vessel malformation but, in this case, digital pulmonary angiography remained the ultimate technique for obtaining the diagnosis.

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