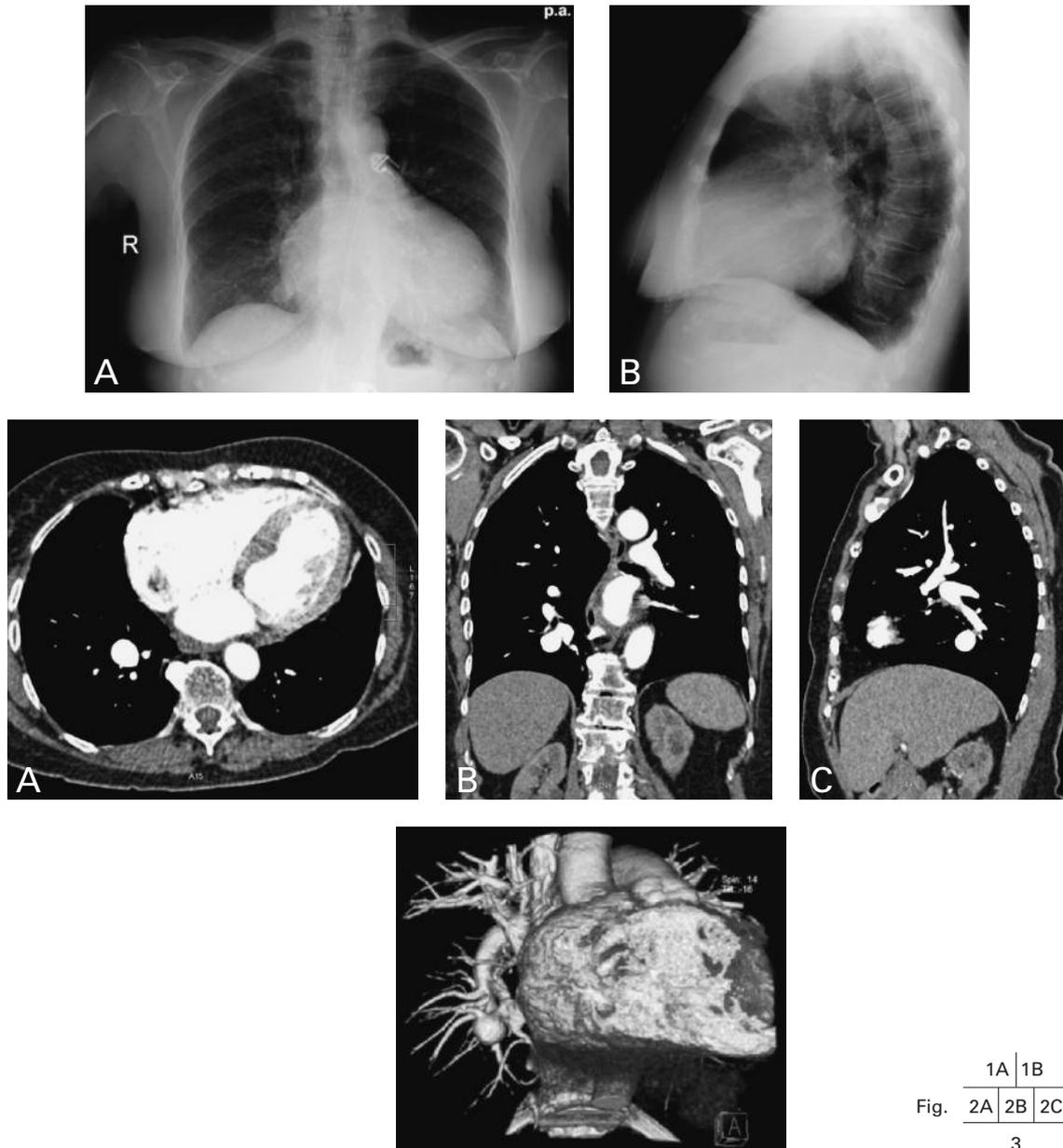


SACCIFORM VARIX OF RIGHT PULMONARY VEIN

T. Couvreur, B. Ghaye¹

Key-word: Varices

Background: An 83-year-old woman was admitted initially for evaluation of cardiac failure, secondary to persistent atrial fibrillation. She presented with increasing dyspnea on exertion and oedema of lower limb for some days.



	1A	1B
Fig.	2A	2B 2C
	3	

¹. Department of Medical Imaging, University Hospital Sart-Tilman, Domaine Universitaire du Sart-Tilman Liège, Belgium

Work-up

Chest radiography (Fig. 1) shows on the PA view (A) and profile view (B), besides cardiomegaly, a right paracardiac rounded opacity of 15 mm is seen.

Contrast-enhanced CT scan of the thorax (Fig. 2) includes a transverse section (A), a reformatted image in the coronal plane (B) and a reformatted image in the sagittal plane (C). All images reveal a sacciform vascular structure measuring 18 mm in diameter, that expands from the inferior root of the right inferior pulmonary vein.

Contrast-enhanced CT scan with volume rendering (Fig. 3) clearly shows the sacciform vascular structure, arising from the right inferior pulmonary vein.

Radiological diagnosis

Based on CT-findings, the diagnosis of *sacciform pulmonary varix* originating from the inferior root of the right inferior pulmonary vein is made.

Discussion

Congenital anomalies of the pulmonary veins presenting with abnormal diameter can be classified in stenosis, atresia and varices. Pulmonary varix (PV) is defined as a localized enlargement or an aneurysmal dilatation of a segment of a pulmonary vein that drains normally into the left atrium. The first case of a PV was reported by Puchlet in 1843, as a fortuitous finding in a newborn autopsy. This anomaly is rare as only 71 cases have been reported up to 1988. There is no gender, nor age predilection. PV is generally considered to be congenital, but mitral valvular disease was noted in 26-33%. Other congenital diseases can coexist, including hypoplastic pulmonary artery, anomalous pulmonary venous drainage, patent ductus arteriosus, and atrial or ventricular septal defect.

Symptoms are unusual, but rare cases of dyspnea or hemoptysis have been reported. Two fatal spontaneous ruptures into pleural cavity have been reported. Another patient died from a varix communication with a bronchus. Two cases of systemic embolism consecutive to in situ thrombosis in a pulmonary varix have been observed.

According to imaging, PV can be morphologically classified in saccular, tortuous and confluent types.

These varices are preferentially situated in the right lower lobe (60%), followed by the left upper lobe (17%), the right upper lobe (8%), the right middle lobe (4%) and the left lower lobe (4%). This lesion is a fortuitous finding at chest x-ray and appears as a mass with smooth, well-defined, and sometimes lobulated margins. Differential diagnosis is similar as for all coin lesions, including bronchogenic carcinoma, pulmonary arteriovenous fistula, granulomatous disease, etc. CT scan and MRI provide the correct diagnosis. Spiral CT is now defined as the method of choice in the evaluation of congenital anomalies of the pulmonary venous system. This method is non-invasive and can also estimate other eventual congenital malformations in a single examination. Bartram and Strickland have described the five angiographic criteria necessary to diagnose a PV: normal arterial phase, opacification of the varix at the pulmonary venous phase, direct drainage into the left atrium, delayed emptying, and anomaly located in the proximal portion of the pulmonary vein.

According to literature, a follow-up with chest x-ray is necessary and sufficient to monitor the size of the varix. The size of the varix does not increase when there is no pulmonary venous hypertension, while an acute increase in size indicates an elevation of the left atrial pressure and then surgical treatment must be considered. Radiological disappearance of the pulmonary varix has been reported in 4 cases and reduction of the size in 1 case, after prosthetic valvular replacement.

Bibliography

1. Andrikakos P, Gallis P, Christopoulos A., et al.: An asymptomatic pulmonary hilar mass: a case report. *Acta Radiol*, 2004, 45: 516-518.
2. Asayama J., Shiguma R., Katsume H., et al.: Pulmonary varix. *Angiology*, 1984, 35: 735-739.
3. Bartram C., Strickland B.: Pulmonary varices. *Br J Radiol*, 1971, 44: 927-935.
4. Uyama T., Monden Y., Harada K., et al.: Pulmonary varices: a case report and review of the literature. *Jpn J Surg*, 1988, 18: 359-362.