Case 14480

Eurorad

Isolated absence of the right pulmonary artery with advanced emphysema in the right lung.

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ISSN: 1563-4086
Section: Chest imaging
Area of Interest: Cardiovascular system Thorax
Procedure: Diagnostic procedure
Imaging Technique: CT-Angiography
Special Focus: Congenital Chronic obstructive airways disease Case Type: Anatomy and Functional Imaging
Authors: Isabeau Hermie, Geert Verswijfel
Patient: 54 years, female

Clinical History:

A 54-year-old women who had to undergo cardiac ablation because of atrial tachycardia presented at our radiology department for a preoperative cardiac CT-scan. Previous history showed recurrent respiratory tract infections. She recently quit smoking (31 pack years).

Imaging Findings:

The cardiac CT-scan revealed an absent right main pulmonary artery. Despite this abnormality the ablation procedure was performed but failed and subsequently she received a pacemaker implantation.
Afterwards, she underwent a computed tomography pulmonary angiogram (CTPA) to evaluate the abnormal hilar configuration. The CTPA showed the absence of the right main pulmonary artery, right-sided displacement of the heart and the mediastinum, volume loss of the right lung and little collateral circulation. Remarkably, there were advanced emphysematous changes with multiple bullae and bronchiectasis in the right lung. No pulmonary embolism, obstructing malignancy or surgical alterations around the right hilum were noticed.

Discussion:

Unilateral absence of the pulmonary artery (UAPA) is a rare congenital cardiovascular malformation with an estimated prevalence of 1 in 200000. There is no gender or race predilection. UAPA is thought to be the result of failure of the fusion between the pulmonary trunk and the sixth aortic arch during embryologic development. The right pulmonary artery is most frequently affected, accounting for two thirds of the cases. Left-sided agenesis seems to be more frequently associated with cardiac abnormalities such as tetralogy of Fallot or cardiac septal defects [1].

Clinical presentation of isolated UAPA is diverse. It can be asymptomatic until late adulthood, but more often symptoms such as dyspnoea, chest pain, decreased exercise tolerance, haemoptysis and recurrent respiratory tract infections are reported. Haemoptysis is caused by large collateral circulations, which exposes the venous systems to abnormally elevated pressures. Other complications are pulmonary hypertension, right heart failure and bronchiectasis.

Typical chest radiographic findings are absent hilar shadow, displacement of cardiac and mediastinal shadow, elevation of the diaphragm and contralateral lung hyperinflation.
The definitive diagnosis of UAPA can be made by CT or MRI. Cross-sectional imaging shows the absence of the

pulmonary artery with collateral circulation, cardiac and mediastinal displacement, lung volume loss, mosaic parenchymal alterations and bronchiectasis due to recurrent infections. Transthoracic echocardiogram can also be used to establish the diagnosis. Angiography is considered the golden standard but is invasive and is rarely performed unless a surgical intervention is considered.

A remarkable aspect of our case is the presence of advanced asymmetric emphysematous changes in the right lung. Currently, it is assumed that emphysema is the result of an imbalance in protease-antiprotease activity. Cigarette smoking is associated with the induction of proteases, but in our case the emphysema presented mainly in the hypoperfused right lung. A possible explanation could be that hypoperfusion of the right lung prohibits the antiproteases in the blood to reach the proteinases produced locally in the lung, hereby inducing an imbalance [2]. Currently, there is no consensus regarding treatment of patients with isolated UAPA. Treatment options include revascularisation surgery, pneumonectomy or lobectomy and embolization of collateral haemorrhage.
In conclusion, UAPA is an extremely rare congenital disorder and is sometimes asymptomatic until late adulthood, postponing the diagnosis. Multiple imaging modalities are available to establish the definite diagnosis. Our case describes the rare finding of advanced asymmetric emphysematous changes in the affected lung.
Differential Diagnosis List: Right UAPA with advanced emphysema in the right lung., Primary pulmonary hypertension, Pulmonary embolism

Final Diagnosis: Right UAPA with advanced emphysema in the right lung.

References:

Ten Harkel AD, Blom NA et al. (2002) Isolated unilateral absence of a pulmonary artery: a case report and review of the literature. Chest 122(4):1471-7. (PMID: 12377882)
Roman J, Jones S (1995) Case report: congenital absence of the left pulmonary artery accompanied by ipsilateral emphysema and adenocarcinoma. Am J Med Sci 309(3):188-90. (PMID: 7879825)

Figure 1

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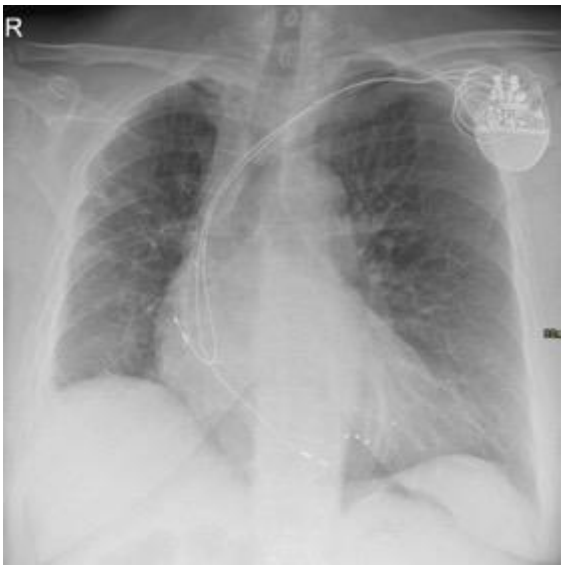

Description: Plain radiograph after pacemaker implantation revealed the absence of the right hilar shadow with diminished right pulmonary vascular markings, cardiac and mediastinal displacement to the right and loss of volume of the right lung. **Origin:** Verswijfel G., Department of Radiology, East Limburg Hospitals, Genk, Belgium.

Figure 2

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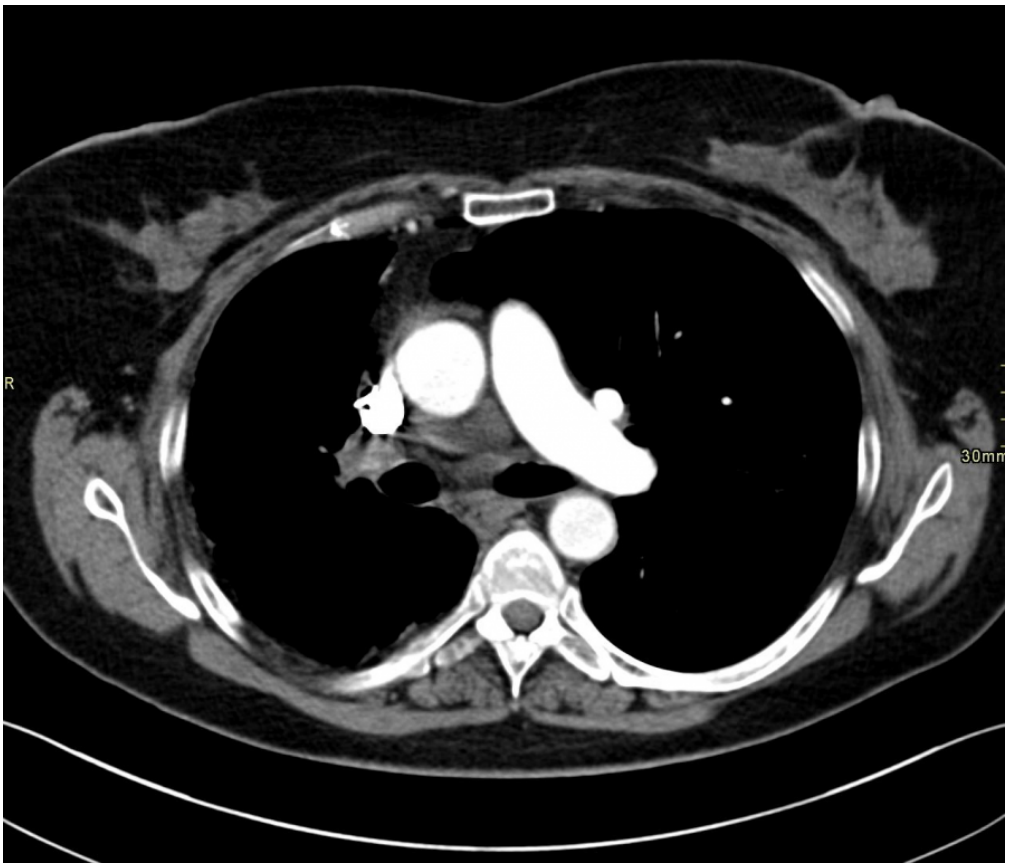

Description: CTPA with soft window settings shows the absence of the right main pulmonary artery. The right internal thoracic artery is enlarged. **Origin:** Verswijfel G., Department of Radiology,East Limburg Hospitals, Genk, Belgium.

Figure 3

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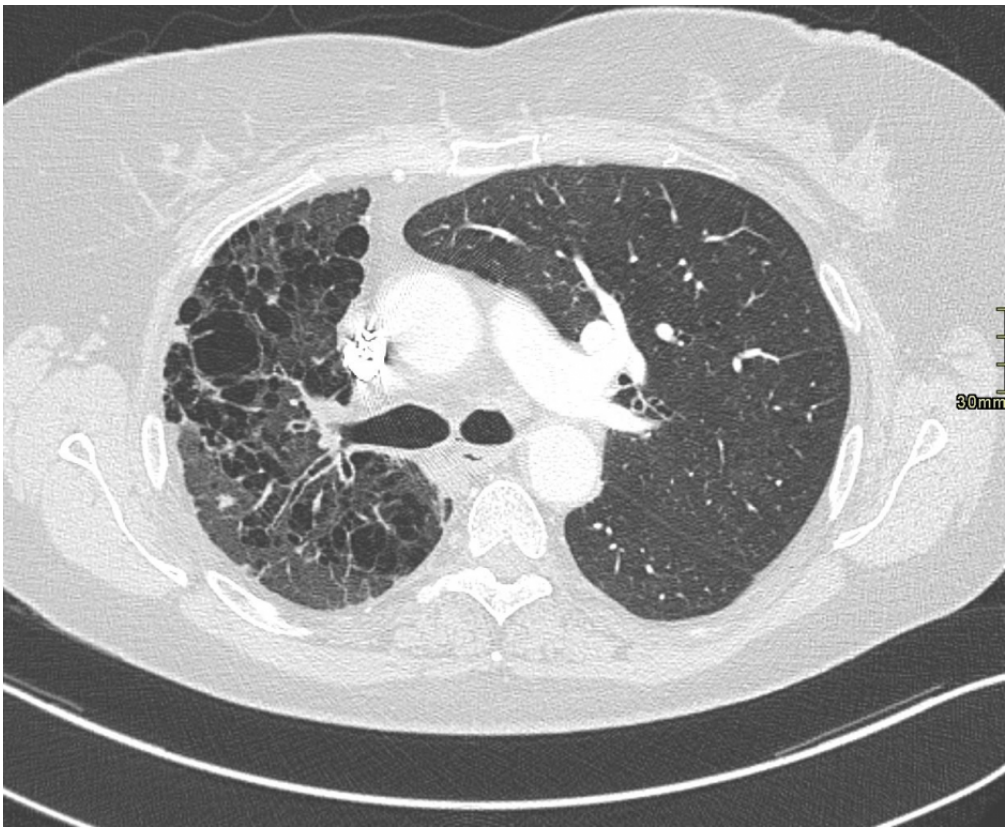

Description: CTPA with lung window settings showing the advanced emphysematous changes and bronchiectasis in the right lung. The mediastinum is displaced to the right. **Origin:** Verswijfel G., Department of Radiology,East Limburg Hospitals, Genk, Belgium.

Figure 4

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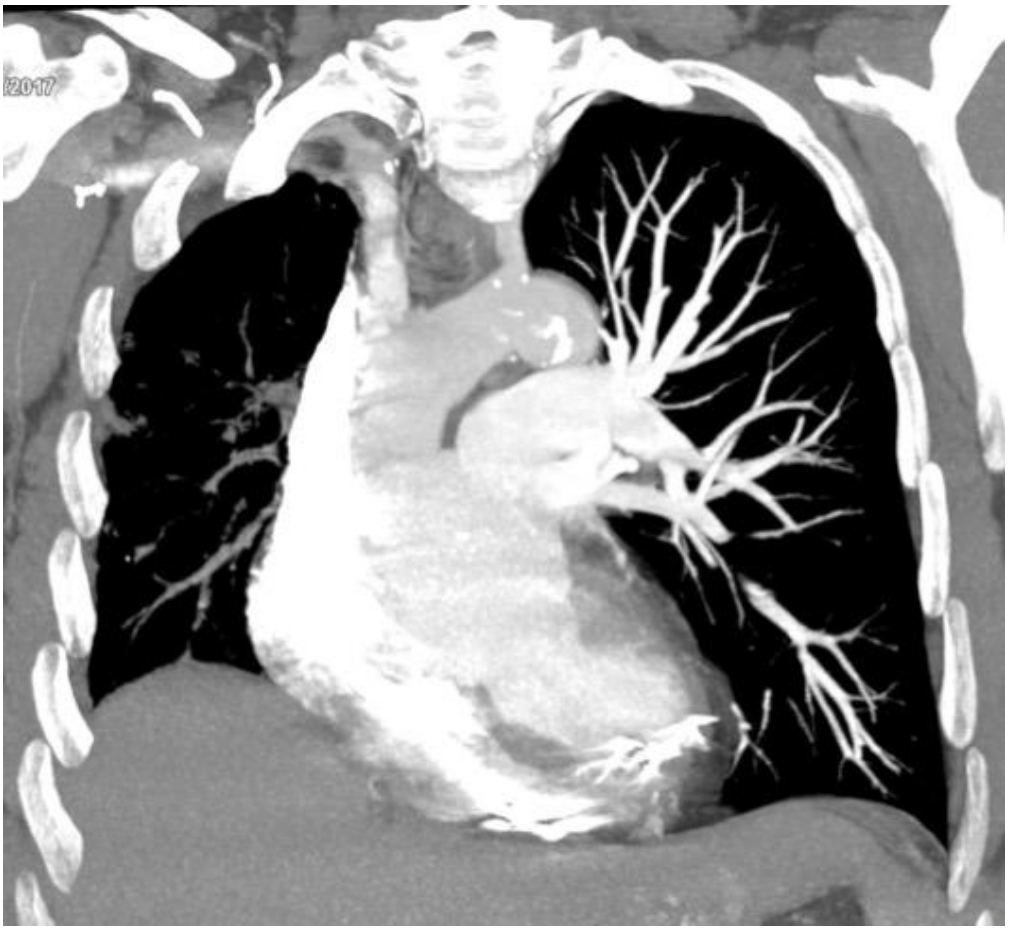

Description: This coronal MIP shows the absent arterial pulmonary vasculature in the right lung with prominent venous structures. Very little collaterals are seen. The vasculature in the left lung is normal.
Origin: Verswijfel G., Department of Radiology, East Limburg Hospitals, Genk, Belgium.