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LEFT PULMONARY ARTERY AGENESIS

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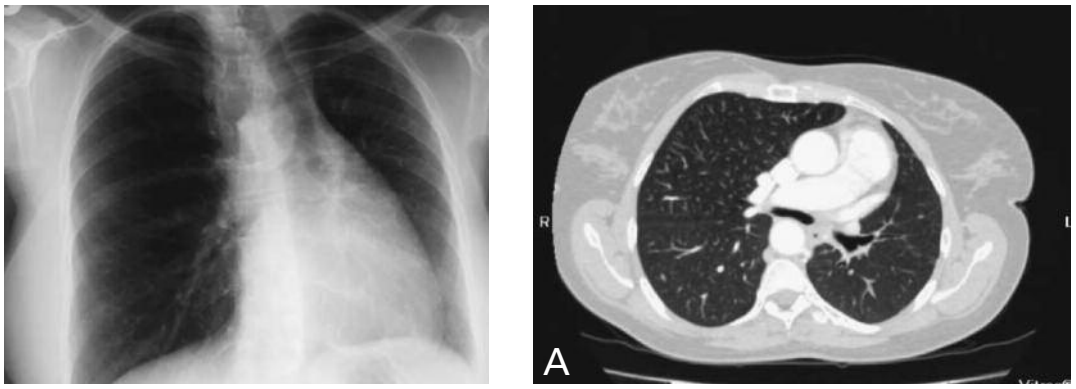
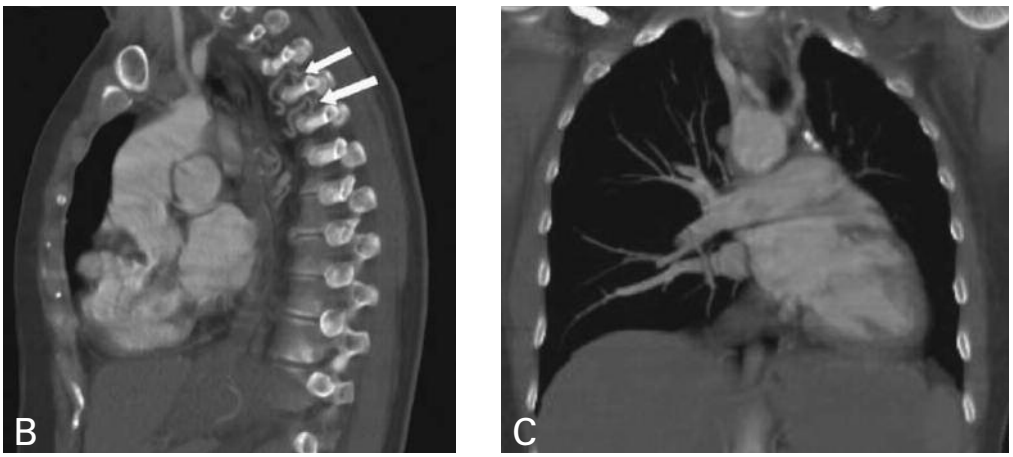
Key-word: Pulmonary arteries, abnormalities

Background: A 45-year-old woman without medical history presented to the first aid department with shortness of breath and some mild thoracic pain. At first the conventional radiography of the chest was interpreted as normal. D-dimer concentrations were raised and a pulmonary embolism was suspected.





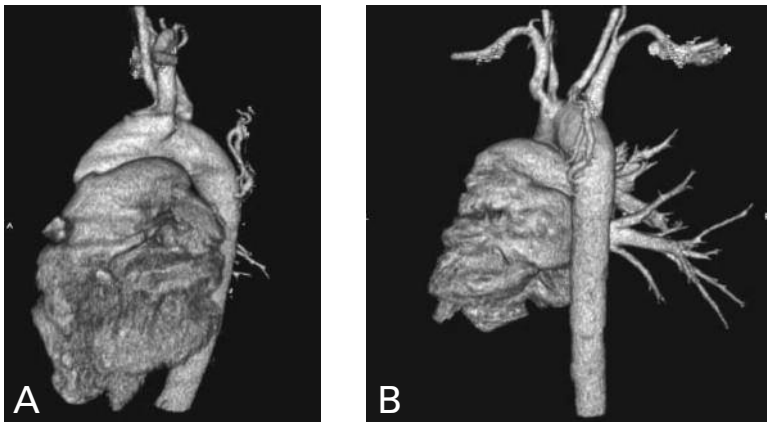


Fig. 1 2A 2B 2C 3A 3B

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Work-up

Conventional radiography of the chest (PA-view) (Fig. 1) shows a right sided aortic arch, deviation of the trachea to the left, asymmetric lung-volumes, the left lung volume being smaller than the right, and abnormal pulmonary artery branching pattern on the left side. No signs of pulmonary infarction nor alveolar consolidation are noted.

Contrast-enhanced CT scan of the thorax (Fig. 2) included a transverse image at the level of the pulmonary trunk (A) that showed absence of left pulmonary artery, asymmetric lung volumes, and left lung smaller than right. Sagittal image of the left hemithorax (B) demonstrates hypertrophic intercostal arteries arising from descending aorta. On coronal reformatted images of chest CT (C), normal branching pattern of pulmonary artery on the right, and absence of pulmonary arteries on the left are noted.

MIP-images of contrast-enhanced CT scan of the thorax (Fig. 3) shows on the left lateral view (A) and posterior view (B) a right sided aortic arch and mirror-image branching pattern of cervical vessels. The first vessel to arise from the aorta is a left sided brachiocephalic artery (innominate artery). Hereafter the right sided common carotid artery and subclavian artery arise from the aortic arch. Hypertrophic intercostal arteries arise from the descending aorta. Note the absence of pulmonary arteries on the left side.

Radiological diagnosis

Radiological findings are corresponding with *absence of left pulmonary artery* combined with right mirror-image aortic arch. No congenital heart disease was found in this patient.

Discussion

Unilateral absence of the pulmonary artery is a rare congenital condition that can occur on both the left and right sides. Absence of the pulmonary artery at the pulmonary trunk or "unilateral non-confluent pulmonary artery" which is a more accurate description of the abnormality, causes under-development of the lung tissue. The abnormality may occur as an isolated lesion, but is frequently accompanied by congenital heart disease like tetralogy of Fallot or septal defects.

Findings on physical examination are unremarkable, exception made for decreased breath sounds on the affected side.

On imaging, the underdeveloped lung may give rise to an asymmetric chest appearance with mediastinal shift to the affected side as shown in the presented case. Elevation of the hemidiaphragm on the affected side may be seen. Systemic collateral vessels arising from the aorta e.g. bronchial arteries or intercostal arteries provide blood supply to the affected lung. Venous return to the left atrium assured through normal pulmonary veins. Rib notching due to enlarged intercostal arteries can occur.

Approximately 15% of patients are asymptomatic. About 40% of patients have mild symptoms and present with recurrent pulmonary infections and decreased exercise tolerance. The systemic flow to the affected lung causes a left-to right shunt., which can lead to pulmonary hypertension and right ventricular hypertrophy in 25% of patients. About 20% of patients present with haemoptysis due to bronchial artery hypertrophy.

One of the known accompanying congenital conditions in absence of a pulmonary artery is a right aortic arch, a condition that occurs in approximately 0.1% of the population. There are two main types of right aortic arch, classified according to its course. In type 1 a mirror-image branching pattern is seen.

First a left sided brachiocephalic artery (innominate artery) arises from the arch, then a right sided common carotid artery and finally the right sided subclavian artery. In type 2 an aberrant left subclavian artery running from the descending aorta, encircling the oesophagus and trachea is present. This artery, called a.lusoria, forms a vascular ring encircling the oesophagus and trachea. On radiographs a right sided arch is identified as an abnormal convex right mediastinal contour, absence of the aortic contour on the left side and deviation of the trachea to the left.

In our patient there were no clinical signs of congenital heart disease, and there was no cardiologic history. Besides the absence of the left pulmonary artery and the right mirror-image aortic arch no developmental (heart) conditions were found.

Bibliography

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