Left Pulmonary Artery Hypoplasia with Left Upper Lobe Atelectasis

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Abstract

A 67 year-old female patient was admitted to our hospital with the complaints of dyspnea, cough, expectoration, hemoptysis and chest pain. Her CT examination revealed left upper atelectasis but could not demonstrate left pulmonary artery. Perfusion scan showed extremely reduced perfusion on the left. In concert with these, MR angiography demonstrated proximal interruption of left pulmonary artery and very thin immediate branches.
These findings led to the diagnosis of left pulmonary artery hypoplasia with upper lobe atelectasis. This pathology is presented here for its extreme rarity.

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Key words: *left pulmonary artery hypoplasia, left upper lobe atelectasis*

Introduction

Agenesis and hypoplasia of the right or left pulmonary artery are among the rarest pulmonary artery anomalies. Left sided pulmonary artery anomalies have even been less frequently reported (1). We present a 67 year old female patient with left pulmonary artery hypoplasia and with left upper lobe atelectasis.

Case Report

A female patient presented to the clinic with complaints of dyspnea, cough, expectoration, hemoptysis and chest pain. These complaints had started 10 days ago. The chest X-ray showed a left upper lobe atelectasis and the patient was hospitalized.

Vital signs were normal. On inspection, the left hemithorax was less developed than the right. Auscultation revealed bronchial breathing sounds in the left upper zone. No other noteworthy findings were detected in the physical examination. Except for mild leucocytosis (11 100 per mm^3) and a high LDH level (1355 mg/dl), the patient's haematological and biochemical evaluation were completely normal. She had never smoked and her detailed past medical history did not reveal any specific information. There was no history of chronic chest symptoms or any pulmonary disorder.

In the chest X-ray the left hemidiaphragm was elevated, and volume loss and hyperlucency were noted in the left

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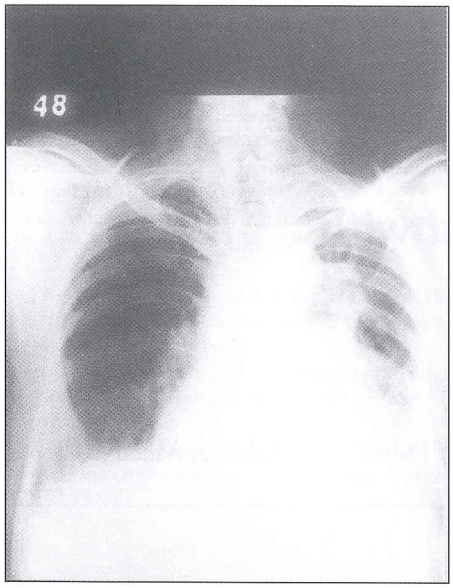

Figure 1. *Chest x-ray on admission.*

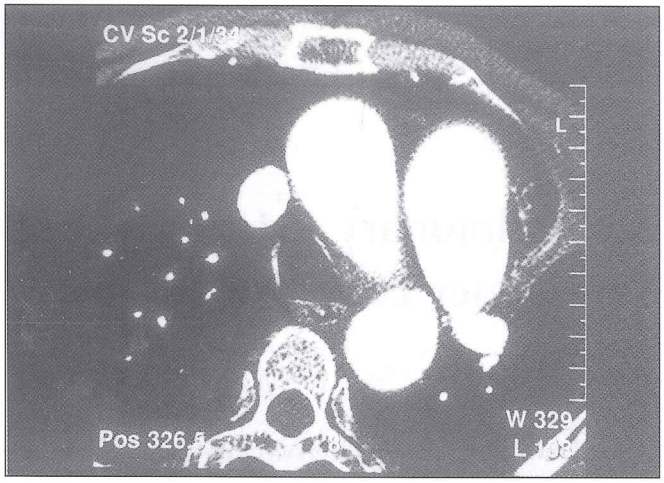

Figure 2. *Thorax computed tomography on admission.*

hemithorax. There was a left hilar retraction toward the upper zone and a homogenous opacity in the left upper zone (Figure 1). During the bronchoscopic examination, the bronchial tree was found to be completely patent, and no endobronchial lesions were detected. In the computerised tomography (CT) of the thorax, the main and right pulmonary arteries were dilated, but the left pulmonary artery could not be visualised (Figure 2). Many enlarged collateral vessels were easily visible on CT. Any lymph node which might cause hypoperfusion by compression was not detected (Figure 3). Backing the bronchoscopy findings, CT examination also clearly demonstrated that although the left upper lobe was totally atelectatic, its bronchus was intact.

MR angiographic examinations revealed a proximal interruption of the left pulmonary artery and its immediate branches were very thin and blood flow to distal regions was found to be extremely reduced in all pulmonary artery branches (Figure 4). The perfusion scan demonstrated a normal perfusion pattern on the right side, but perfusion of the left lung was extremely reduced (Figure 5). Ventilation scan of both lungs were normal. To better define the cardiac status, ECG and Doppler echocardiography were performed. ECG revealed atrial fibrillation. Echocardiography revealed an ejection fraction of 68% and a pulmonary artery pressure of 43 mmHg. The right and left atria showed a dilation of 1st to 2nd degree, tricuspid insufficiency and a 1st degree mitral insufficiency. The patient responded well to a low-salt diet and antibiotic, diuretic, calcium channel blocker and low dose ACE inhibitor medication. Her complaints disappeared in 1 week, and she was discharged to be followed up on an ambulatory basis.

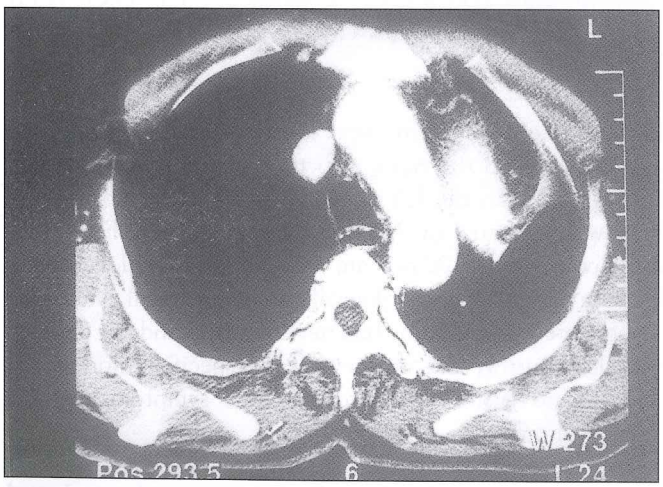

Figure 3. *Thorax computed tomography showing many enlarged collateral vessels.*

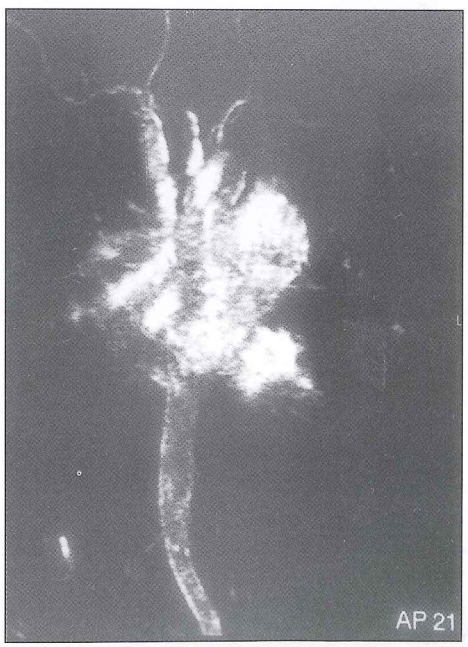

Figure 4. *MR angiographic examination.*

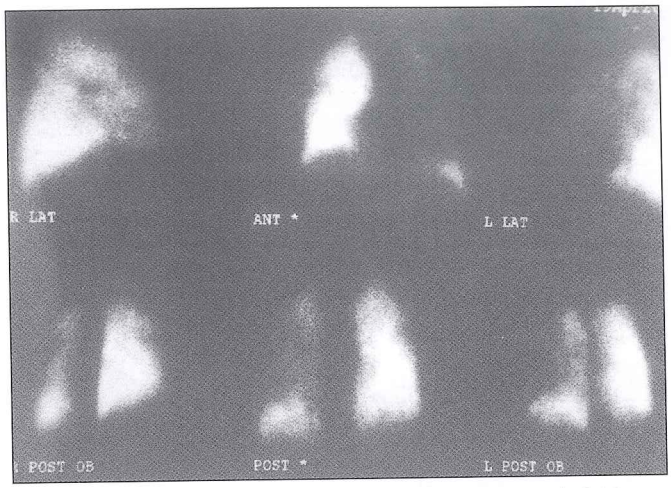

Figure 5. *Scan showing an extremely reduced perfusion pattern in the left lung.*

Discussion

Although there are rare examples of complete absence of right or left pulmonary arteries, this anomaly is better defined as a proximal interruption, because the vessels are usually intact in the lungs (1). The majority of cases are diagnosed in childhood, but occasionally some asymptomatic cases are first recognised in adulthood through the detection of an abnormal chest radiograph (2,3). Our patient had no complaints until 10 days before her admission. In these asymptomatic patients, the blood supply distal to the interruption is provided by a patent ductus arteriosus, bronchial collaterals, an artery arising directly from the aorta or less commonly, by transpleural intercostal collaterals (4). In this case many bronchial collaterals were also visible on the chest CT. Although some concomitant congenital cardiovascular anomalies were described and particularly tetralogy of Fallot, there is one rare case report demonstrating an associated pulmonary parenchymal abnormality (1,5). In this case report, a left upper lobe bronchiectasis was described as an anomaly concomitant to a left pulmonary artery hypoplasia.

The most frequent findings encountered in chest x-rays are decreased size of the affected hemithorax, compensatory hyperinflation of the contralateral hemithorax, elevation of the ipsilateral hemidiaphragm, absent ipsilateral and enlarged contralateral pulmonary artery shadow and ipsilateral shift of the mediastinum (2,3). Although diminished or total absence of ipsilateral perfusion is demonstrable by perfusion scintigraphy, ventilation scan of these patients are normal (3). Almost all of these radiological and scintigraphic changes were detected in our patient.

The definitive diagnosis of this condition can be made by MRI, contrast-enhanced CT or angiography (6,7). In our case the diagnosis was made by a combination of CT and MRI. We presented this patient because the associated pulmonary parenchymal abnormality is extremely rare in patients with right or left pulmonary artery hypoplasia. It should also be stressed that gross congenital pulmonary vascular abnormalities can sometimes be asymptomatic and may go unnoticed until an advanced age.

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