Absence of Left Pulmonary Artery

Case report

*Dilip Sankhla, Samir Hussein, Jojy George, Ranjan William, Sinan Al-Azawi, Badriya Al-Qassabi

Department of Radiology and Molecular Imaging, Sultan Qaboos University Hospital, Muscat, Sultanate of Oman.

To whom correspondence should be addressed. Email: sankhladilip@gmail.com

Absence of Left Pulmonary Artery

Case report

A 21 year old lady was admitted to Sultan Qaboos University Hospital, Oman, via its outpatient clinic, for the evaluation of a small left hemithorax and a mild cough. She had a history of mild attacks of shortness of breath, especially after exertion, and occasional left-sided chest pain. Her symptoms had got worse during the previous year. There was no history of associated fever, sweating, chills, loss of appetite or weight loss. There was no past history of allergy or contact with patients with pulmonary tuberculosis. On examination, she was observed to be in good condition and her vital signs were within normal ranges. Examination of the respiratory system showed a tracheal shift to the left, reduced left chest movements and reduced breath sounds and dull percussion in the left lung.

Her chest X-ray [Figure 1] showed evidence of volume loss in the left hemithorax, a mediastinal shift to the left and crowding of the left ribs. The right lung showed compensatory hyperinflation. The left lung showed less vascular marking compared to the right side. The right main pulmonary artery

**abstract:** Agenesis and hypoplasia of left-sided pulmonary artery anomalies have been infrequently reported. The majority of cases are diagnosed in childhood, but occasionally some asymptomatic cases are first recognised in adulthood when detected by an abnormal chest radiograph. We report a twenty-one year old female patient with left pulmonary artery agenesis who was asymptomatic till adulthood, but presented with mild respiratory symptoms and an abnormal chest X-ray. A contrast enhanced computerised tomography (CECT) scan helped to establish the diagnosis. Early diagnosis of this condition is essential to avert potentially lethal complications.

**Keywords:** Unilateral Pulmonary artery agenesis; Left pulmonary artery; Congenital abnormalities; Case report; Oman.
was normal and the left was not seen. These findings were suggestive of either a hypoplastic left lung or secondary to chronic obstruction in the left main bronchus. Bronchoscopy, however, revealed normal carina and main bronchi. A pool of secretions was noted in left main bronchus. The bronchial aspirate was negative for acid fast bacilli and its culture was negative. A contrast enhanced computerised tomography (CECT) scan [Figure 2] was performed on this patient to visualise the pulmonary vasculature. It revealed the absence of her left pulmonary artery, a right-sided aortic arch and a dilated main pulmonary artery with evidence of collaterals at the left hilum and in the apical region. The lung parenchyma was normal apart from volume loss of the left lung with compensatory hyperinflation of the right lung. The bronchi on either side were patent. A perfusion-ventilation scan (V/Q) [Figure 3] was also performed. The perfusion scan showed normal perfusion of the right lung and no uptake in the left lung. The ventilation scan showed hypoventilation in the left lung and a normally ventilated right lung. These findings led to the diagnosis of left pulmonary artery agenesis.

Discussion

The unilateral absence of a pulmonary artery is a rare condition and is believed to originate from a single embryological defect in the proximal sixth aortic arch. It refers to proximal interruption of those vessels, but the vessels are usually intact in the lungs. The incidence is about 1 in 200,000. Left-sided pulmonary artery anomalies have been reported even less frequently. Right pulmonary artery agenesis is twice as common as left pulmonary artery agenesis.1, 2 The majority of cases are diagnosed in childhood, but occasionally some cases with very few symptoms are first recognised in adulthood through the detection of an abnormal chest radiograph. Chest X-rays frequently show a 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,
a right-sided aortic arch and an ipsilateral shift of
the mediastinum. Most of these findings were
present in the chest X-ray of our patient.

Fallot’s tetralogy, intracardiac septal defects,
coarctation of aorta, right aortic arch and
Eisenmenger’s syndrome are associated with left
pulmonary artery atresia (75%). The most common
presenting symptoms in patients with pulmonary
atresia are recurrent pulmonary infection, mild
dyspnea and decrease exercise tolerance. Our patient had no major complaints until a year
before her admission. In these asymptomatic
patients, the blood supply distal to the interruption
is provided by a patent ductus arteriosus, bronchial
collaterals, and an artery arising directly from the
aorta or, less commonly, by transpleural intercostal
collaterals. Many bronchial collaterals were visible
on the CECT of our patient. The presence of a
hypoventilated left lung with no perfusion on that
ting has been reported in the literature. This
also explains the late onset of her symptoms. The
unilateral absence of the pulmonary artery (UAPA)
may be suspected by the presence of recurrent
respiratory infections, haemoptysis, bronchiectasis
or pulmonary hypertension in the absence of other
known causes. The parenchymal abnormality is
extremely rare.

The diagnosis of UAPA can be made by chest
radiography and echocardiography. The definitive
diagnosis, anatomic details and the presence of
hilar collaterals, however, can be discerned by
CECT scanning and MR Angiography. Cardiac
angiography and pulmonary venous wedge
angiography have been considered in establishing
the diagnosis of UAPA and identifying collateral
vessels. However, with the advent of CECT and
MR angiography, cardiac catheterisation should
be reserved for patients requiring embolisation
for recurrent haemoptysis or being considered
for revascularisation surgery. If sufficiently
large hilar arteries are found, revascularisation
may significantly improve the outcome. However,
large collateral arteries can also lead to pulmonary
hypertension or haemoptysis which may require
embolisation. The early diagnosis of UAPA and
management can avert the above complications and
it can also avert the potentially devastating effects
of high altitude or pregnancy.

Conclusion

Agenesis of the left pulmonary artery is a rare
anomaly and the patient may remain asymptomatic
till adulthood. Imaging plays a major role in the
diagnosis and detecting the associated findings
in heart and lungs. For patients who present with
a unilateral small hemithorax, recurrent chest
infections and an abnormal chest X-ray, a UAPA
anomaly should be considered in the differential
diagnosis. An early diagnosis can avert serious
complications.

References

1. Fraser RS, Müller NL, Colman N, Pare PD.
Developmental anomalies affecting the pulmonary
vessels. In: Fraser RS, Pare PD, Eds. Fraser and Pare’s


