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CASE REPORT

Absence of Left Pulmonary Artery
Case report

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غياب الشريان الرئوي الأيسر
تقرير حالة

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الملخص: عدم تخلق وقصور نموّ الشريان الرئوي الأيسر الناجّ عن العيوب الخلقية عادة ما يكون نادر الذكر في التقارير الطبية. يتم تشخيص معظم الحالات في مرحلة الطفولة. وأحيانا ما تكون هذه الحالات بدون أعراض ويتم التعرف عليها للمرة الأولى في سن البلوغ من خلال أشعة الصدر. نعرض هنا حالة مريضة تبلغ من العمر واحدا وعشرين عاما تعاني من عدم تخلق الشريان الرئوي الأيسر الذي كان بدون أعراض حتى البلوغ . حيث بدأت تعاني من أعراض بسيطة في الجهاز التنفسي وكانت صورة الأشعة السينية للصدر غير طبيعية. ساعد الاسْتِعْزازُ التَبايُنِيّ للتَّصْويرُ المَقْطَعِيُّ المُحوسَب في تأكيد التشخيص. من الجدير ذكره أن التشخيص المبكر لهذة الحالة المرضية ضروري لتجنب المضاعفات التي يمكن أن تكون قاتلة.

مفتاح الكلمات: عدم تخلق الشريان الرئوي وحيد الجانب . الشريان الرئوي الأيسر . العيوب الخلقية . تقرير حالة . عمان.

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.

The unilateral absence of a pulmonary artery is a rare condition with diverse clinical presentations. Routine investigations including chest X-rays and echocardiography may suggest the diagnosis of absence of a pulmonary artery, but confirmation requires computed tomography (CT) scans and magnetic resonance angiography (MRA). An early diagnosis of a unilateral absence of pulmonary artery and appropriate intervention may significantly improve the outcome. We report a rare case of a young woman with a unilateral absence of the left pulmonary artery who presented with a recent history of shortness of breath and chronic cough.

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

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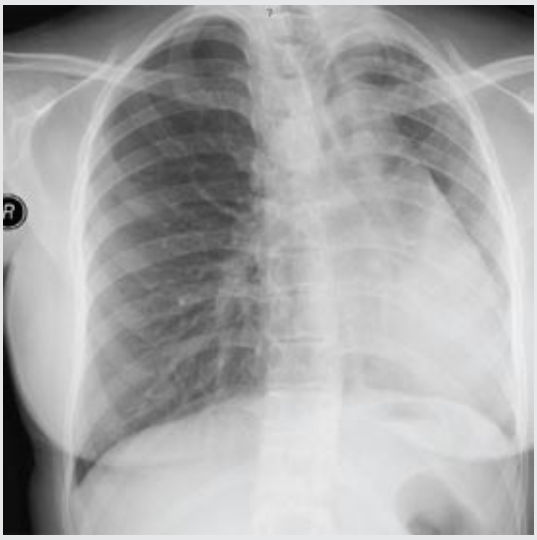
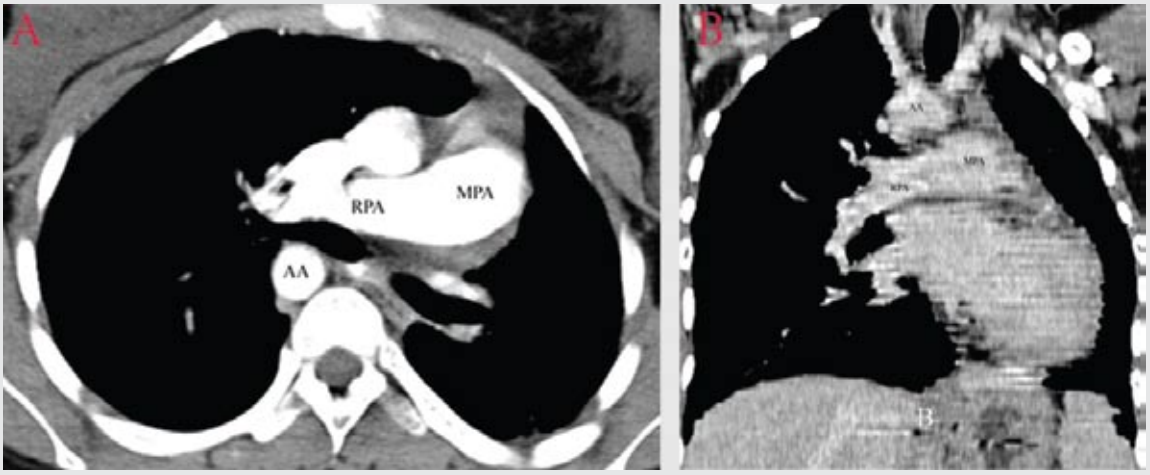


Figure 1: Chest X-ray posterior anterior view shows hypoplastic left lung, decreased vascular markings with ipsilateral mediastinal shift

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.

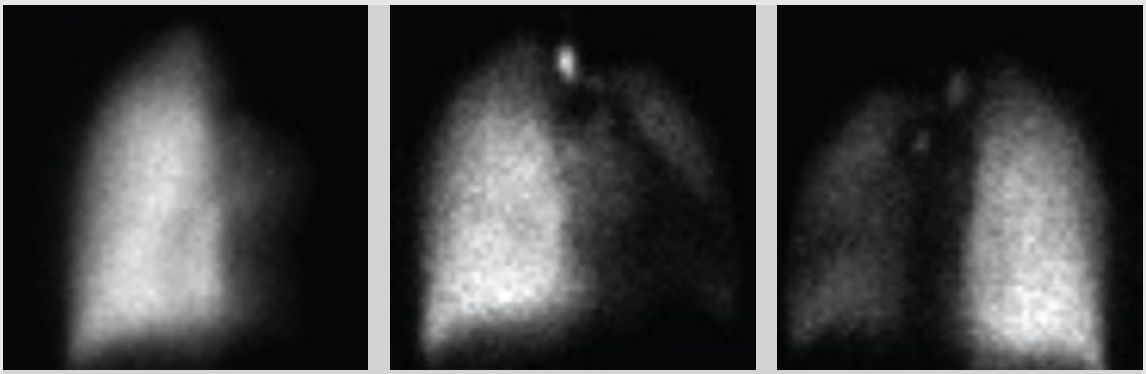
Discussion

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,



Legend: RPA = right main pulmonary artery; MPA = main pulmonary artery; AA = Aorta

Figures 2a & 2b: Contrast enhanced computed tomography axial image (A) and reformatted coronal image (B) shows absence of left pulmonary artery and right-sided aortic arch.



Perfusion Anterior View | Ventilation Anterior View | Ventilation Posterior View

Figure 3: Perfusion and ventilation scans. The perfusion scan shows no activity in the left lung and normal activity in right lung. Ventilation scans display mild activity in the left lung and normal activity in right lung

a right-sided aortic arch and an ipsilateral shift of the mediastinum.[2, 3, 4] 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%).[5] The most common presenting symptoms in patients with pulmonary atresia are recurrent pulmonary infection, mild dyspnea and decrease exercise tolerance.[5]

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.[6] Many bronchial collaterals were visible on the CECT of our patient. The presence of a hypoventilated left lung with no perfusion on that side also explains the presence of collateral vessels supplying the left lung. Diminished or total absence of ipsilateral perfusion and normal or mildly reduced ventilation has been reported in the literature.[3] 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.[7, 8] 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.[4, 5] 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.

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