UNILATERAL PULMONARY ARTERY AGENESIS: CASE REPORT

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ABSTRACT Unilateral pulmonary artery agenesis (UPAA) is an uncommon congenital anomaly and most patients present in neonatal period with respiratory symptoms. Left-sided pulmonary artery agenesis is less frequent than right-sided. We report a patient who presented with hemoptysis and bronchial dilatation. The diagnosis of UPAA was made.

KEYWORDS Pulmonary artery agenesis, Hemoptysis, Malformation

Introduction

Unilateral pulmonary artery agenesis (UPAA) is a rare congenital anomaly, first described by Fraentzel in 1968[1]. The diagnosis is often made during childhood but may go unnoticed; it is based mainly on the CT angiography of the chest. This anomaly may be isolated or associated with other cardiovascular abnormalities. It most often affects the right pulmonary artery[1]. Through an observation and a review of the literature we will focus on the agenesis of the pulmonary artery.

Case report

A 31 year-old female with history of recurrent pneumopathy in adolescence, presented since 2 years; hemoptysis, cough and dyspnea on exertion, which gradually aggravated. She had no notion of fever, weight loss, night sweat or loss of appetite. On physical examination, vital signs were stable with no respiratory distress or use of accessory muscles. Normal breath sounds had decreased over the left lung but the heart, abdomen and extremities were normal.

On evaluation, chest x-ray revealed decreased volume of the left lung and leftward shifting of trachea (figure 1); thus, pulmonary CT angiography was performed and the findings were compatible with unilateral congenital left pulmonary artery agenesis and hypertrophied bronchial arteries as well as a restriction of the homolateral pulmonary volume, which is the site of bronchiectasis involving mainly the upper lobe and the base of the lower lobe (figure 2-3). Spirometry studies and complete blood gas studies were within normal limits, the rest is without particularity, notably the complete blood count. The patient underwent a left pneumonectomy, per operatively we noted a decreased lung size with a mediastinal shift, absence of a pulmonary artery, atypical vascular markings, a higher than normal diaphragm and the absence of visualization of the left pulmonary artery. The patient stayed in intensive care unit for 7 days. She presented a postoperative hemothorax of the residual cavity that spontaneously resorbed.

Discussion

UPAA is a rare congenital anomaly due to a failure in the connection of the sixth aortic arch with the pulmonary trunk. Since the common age of presentation is the neonatal period and childhood, it is frequently misdiagnosed in the adulthood and is often not included in the list of differential diagnoses of the unilateral hyperlucent lung [2-3]. The most common presenting symptoms in patients with pulmonary artery atresia include recurring pulmonary infection, mild dyspnea and decreased exercise tolerance [2]. Hemoptysis has been described as a clinical symptom in up to 10% of patients [4-5]. Clinicians and radiologists should be well aware of the possibility of undiagnosed cases in adults, with many atypical characteristics. Our patient presented a notion of recurrent lung infections in adolescence.
Figure 1. Chest X-ray (posteroanterior view) shows slight leftward shifting of the trachea, sites of bronchiectasis. Compensatory hyperinflation in contralateral hemithorax was also noted.

Figure 2. Pulmonary CT angiography: reformatted coronal image.

At 29 years old, the patient presented an aggravation of the respiratory symptoms urging her to seek medical care. Chest X-ray often shows a reduction in the size of the affected hemithorax, compensatory hyperinflation of the contralateral hemithorax, elevation of the ipsilateral hemidiaphragm, and ipsilateral shift of the mediastinum [6]. Possible diagnoses such as pneumonia, pulmonary tuberculosis, bronchiectasis and pulmonary thromboembolism can be considered.

The pulmonary CT angiography reveals and confirms the diagnosis of UPAA [7], which is often not included in the list of differential diagnoses. This reported case showed that we should consider unilateral pulmonary agenesis in an adult patient with radiographic findings such as asymmetric aeration of the lungs, even in the absence of major respiratory symptoms[8]. Surgery with pneumonectomy is considered in case of failure of the endovascular treatment of hemoptysis or in case of recurring infections [7].

Conclusion

UPAA is a rare malformation with a potentially fatal prognosis. Its assessment and the differential diagnostics require imaging based on a pulmonary CT angiography that allows simultaneous analysis of vessel opacification and pulmonary parenchyma. These patients require careful follow-up due to the risk of pulmonary arterial hypertension and hemoptysis.

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Conflict of interest

There are no conflicts of interest to declare by any of the authors of this study.

References


3. Isolated unilateral absence of a pulmonary artery: a case report and review of the literature