Case Report

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TANAFFOS

Unilateral Pulmonary Artery Agenesis in an Adult Patient with Cough and Hemoptysis: A Case Report

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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 and is sometimes associated with cardiac anomalies. We report a patient with a history of repaired ventricular septal defect, who presented with cough and hemoptysis and the diagnosis of UPAA was made.

Key words: Pulmonary artery agenesis, Ventricular septal defect, Hemoptysis

INTRODUCTION

Blood flow through the central pulmonary arteries (i.e., the main pulmonary artery, right pulmonary artery, and left pulmonary artery) may be interrupted due to pulmonary artery agenesis, pulmonary atresia, or pulmonary stenosis (1). UPAA is a rare congenital anomaly, which is frequently diagnosed during childhood and is usually associated with other cardiovascular abnormalities. Fallot's tetralogy, intracardiac septal defects, coarctation of the aorta, right aortic arch and Eisenmenger's syndrome are types of cardiac abnormalities that can be associated with left pulmonary artery atresia. UPAA may be on the left or the right side while left-sided pulmonary artery anomalies have been reported less frequently. Most patients without any associated cardiac anomaly manifest only minor or no symptoms and survive into adulthood (2). We report a case of unilateral pulmonary artery agenesis associated with ventricular septal defect who had no pulmonary symptoms until adulthood.

CASE SUMMARIES

A 24 year-old male presented with hemoptysis and cough since 2 weeks earlier. He was well until about two weeks before the evaluation when he experienced dyspnea on exertion which, was gradually aggravated and a few days later, cough and hemoptysis developed. He had no history of chest pain, fever, weight loss, night sweat or loss of appetite.

He was a non-smoker college student with no history of illicit drug consumption. Family history was negative but past medical history was remarkable for a cardiovascular surgery for ventricular septal defect repair while he was 17 years old. 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 (Figure 1) revealed decreased volume of the left lung and leftward shifting of trachea; thus, pulmonary CT angiography

(Figures 2-4) was performed and the findings were compatible with unilateral congenital left pulmonary artery agenesis and hypertrophied bronchial arteries. After the diagnosis was made, the patient received conservative treatment. In subsequent visits, he had minor respiratory symptoms with no limitations in physical activities.

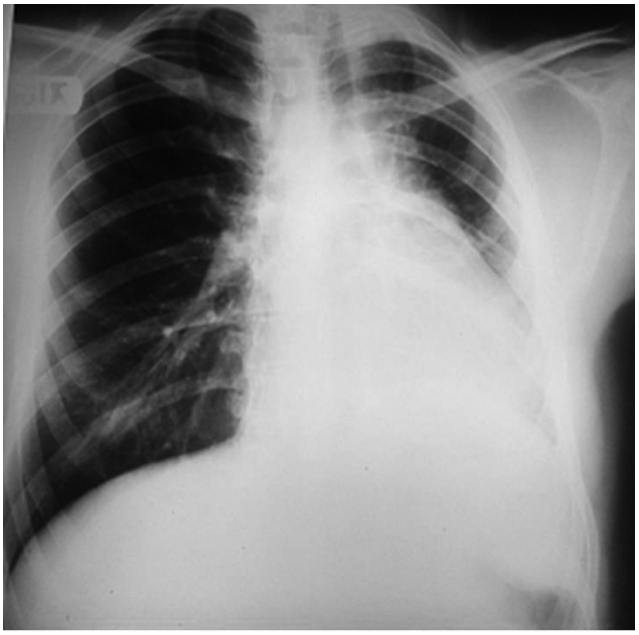

Figure 1. Chest X-ray (posteroanterior view) shows leftward shifting of the trachea and decreased rib distances in the left side. Compensatory hyperinflation in contralateral hemithorax was also noted.

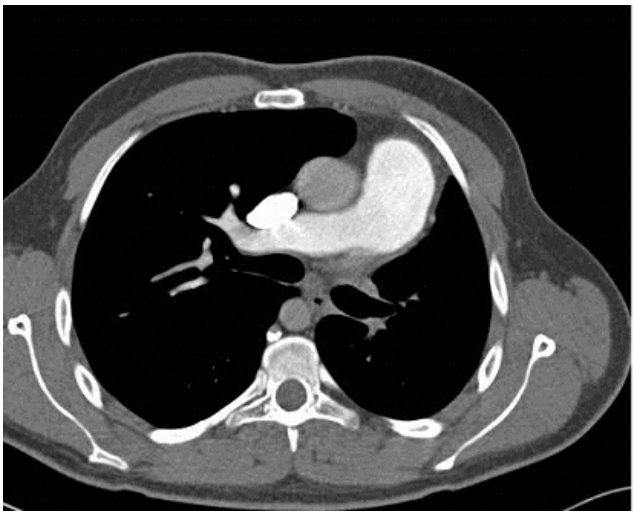

Figure 2. Pulmonary CT angiography revealed absence of left pulmonary artery at the bifurcation level (axial image).

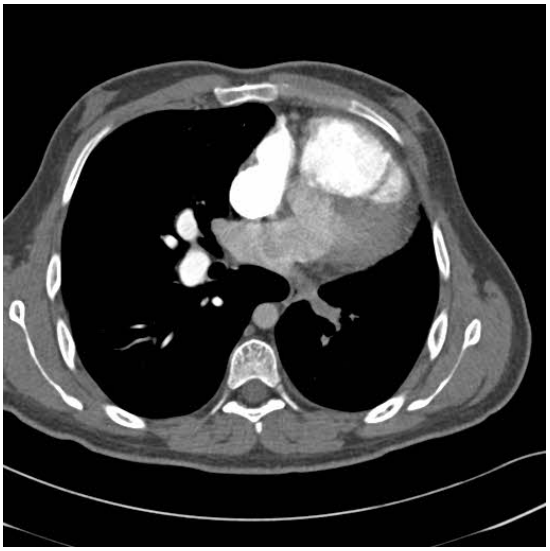

Figure 3. Pulmonary CT angiography shows enhancement of right branches of the pulmonary artery while there is no enhancement in the other side.

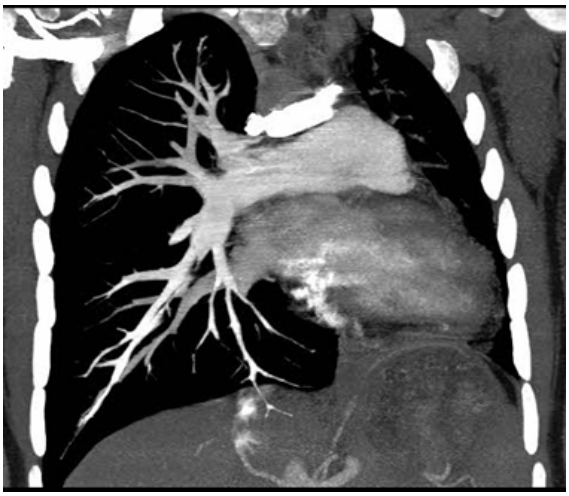

Figure 4. Pulmonary CT angiography: reformatted coronal image.

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 (1,2). The most common presenting symptoms in patients with pulmonary artery atresia include recurrent pulmonary infection, mild dyspnea and

decreased exercise tolerance (2). Hemoptysis has been described as a clinical symptom in up to 10% of patients (3,4). Clinicians and radiologists should be well aware of the possibility of undiagnosed cases in adults, with many atypical characteristics. Our patient was well until 17 years of age, when he was diagnosed with ventricular septal defect, which was repaired surgically. Later on, when he was 24 years old, respiratory symptoms appeared urging him 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, absent ipsilateral and enlarged contralateral pulmonary artery shadow and ipsilateral shift of the mediastinum (5). Most of these findings were present on the chest X-ray of our patient. In the first visit, possible diagnoses such as pneumonia, pulmonary tuberculosis, bronchiectasis and pulmonary thromboembolism were considered; thus, pulmonary CT angiography was done and revealed a diagnosis, which is often not included in the list of differential diagnoses. The diagnosis of UPAA can be made by chest radiography and echocardiography but to confirm the diagnosis, usually computed tomography and MR angiography are needed. We reported this case to mention 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.

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