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Pulmonary Artery Agenesis with Bronchial Asthma

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ABSTRACT

Unilateral Pulmonary Artery Agenesis (UPAA) is a rare congenital anomaly during the 4th week of gestational age. It is defined as an absence of pulmonary parenchyma and its supporting artery. A 9-year-old girl was admitted to our hospital because of chronic cough. Chest examination showed a decrement in lung sound of right hemi-thorax with expiratory wheeze. Chest radiography (CXR) revealed a semi-opaque right hemi-thorax. Chest CT with intra-venous contrast demonstrated absence of the right pulmonary artery and lung parenchyma with hyper-inflated left lung and dextro-position of mediastinum. This case emphasizes that in patients with respiratory compliant and chronic cough CXR must be done to rule out similar diagnosis other than asthma.

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Introduction

Unilateral Pulmonary Artery Agenesis (UPAA) is a rare congenital anomaly with prevalence of 1/200,000 to 1/300,000 of patients (1-3). It is commonly coexisted with other congenital anomalies such as cardiovascular defects (2). The embryologic reason for the absence of pulmonary artery is involution of the proximal sixth aortic arch and persistence of the connection of the intrapulmonary artery to the distal sixth aortic arch during the 4th week of gestation (3, 4). UPAA can remain asymptomatic till adolescence with median age of 14 years at diagnosis (2, 3). Symptoms may include dyspnea, chest pain, recurrent pulmonary infections, hemoptysis, and pulmonary hypertension (5, 6). Decreased sounds in common clinical finding in the affected lung (3). Pulmonary function tests (PFT) reveal restrictive patterns (3). Prognosis of pulmonary artery agenesis depends on coexisting anomaly. However, dextrocardia and respiratory failure due to pulmonary arterial hypertension may be a common cause of morbidity and mortality (2).

Case report

A 9-year-old girl was referred to our hospital with chronic productive cough. She was born by normal vaginal delivery at 38 weeks of pregnancy. Her birth weight was 4300 gr with no need for ventilator support. There was no family history of atopy. The patient had non-smoker parents, with a history of recurrent lower respiratory tract infections. The heart sounds were heard in right side. Lung auscultation revealed decreased respiratory sounds on the right hemi-thorax with expiratory wheeze, while the rest of clinical examinations were normal. The laboratory tests are listed below.

WBC: 6200/ml: (Noutrophil; 49%, Lymph; 41%
Monocyte; 2%, Eosinophil; 8%
RBC: 4.4 mil/ml
Hb: 12 g/dl
Hct: 37%
MCV: 84fl
Platelet count: 329000/ml
ESR <7 mm/h
Sweat test: 45 (Normal: 0-60)
CRP <0.6

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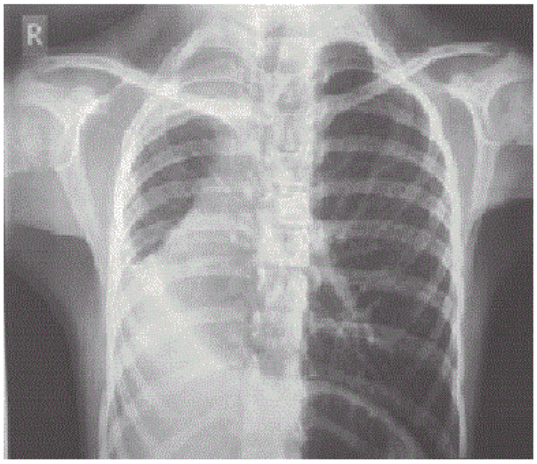

Figure 1. CXR (Right mediastinal shift and opacity of the right hemithorax except for right lung apex)

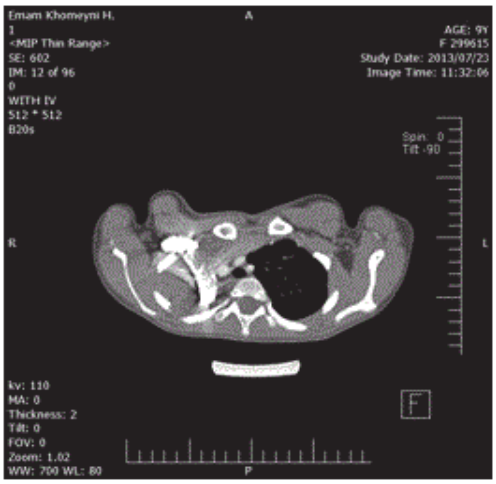

Figure 2. Absence of right lung parenchyma

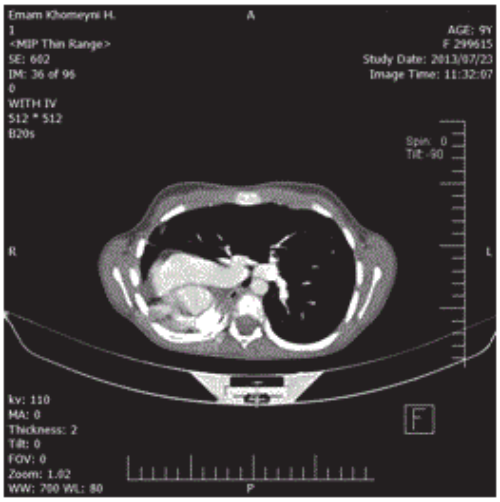

Figure 3. Absence of right lung parenchyma and right main bronchus with hypertrophy of left lung on chest CT

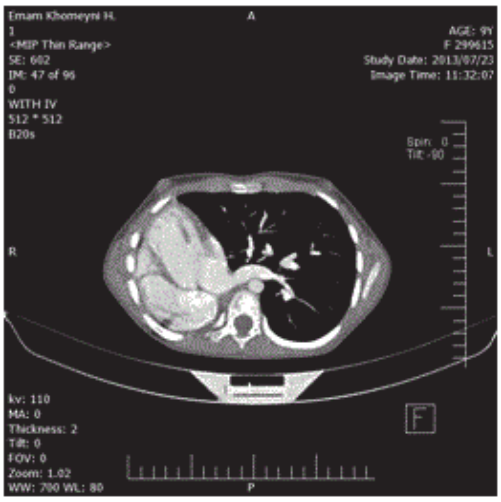

Figure 4. Absence of right lung parenchyma and right main bronchus with hypertrophy of left lung on chest CT

IgA <0.6 (Normal 0.71-3.5)
IgM 0.5(Normal 0.4-2.63)
IgE 112(Normal <90, 6-9 years)
IgG (Serum) 7.7g/Lit (Normal 4.5-13.6)

CXR demonstrated mediastinal shift to the right side with opacity of the right hemi-thorax except in apex, absence of the pulmonary artery shadow and hyperlucency in the left lung (Figure 1). Pulmonary function tests showed a reduced airflow with reversibility of the bronchodilatory test according to ATS Guidelines (Table 1).

Chest CT scan with IV contrast showed absence of right pulmonary artery and lung parenchyma with mediastinal and cardiac shifts to the right side and absence of the right pulmonary artery suggesting UPAA (Figure 2-4).

Echocardiogram revealed dextrocardia with absence of right pulmonary artery and ejection fraction of 60 percent.

Discussion

UPAA is a rare congenital anomaly compared with the other congenital anomalies such as cardiac disorders. This anomaly is commonly asymptomatic, while in other patients the symptoms are nonspecific (6).

There is no sex predilection and both sides of lung are affected equally (3). Left-sided agenesis is always isolated; however, unilateral agenesis of

Table 1. PFT parameters of case

Parameter	Predicted	Prebronchodilator		Post bronchodilator		Change	
		Actual	% Predict	Actual	% Predict	Actual	Percent
FEV1	1.82 Lit	0.80 Lit	44	1.04 Lit	57	0.24 Lit	+13
FVC	2.12 Lit	0.89 Lit	42	1.16 Lit	55	0.27 Lit	+13
FEV1/FVC	86%	-	89	-	89	-	- 0.01
FEF 25-75*	2.41 L/s	0.96 L/s	40	1.25 L/s	52	0.31 L/s	+12

*Forced expiratory flow at 25-75% in liters per second

the right lung is frequently associated with other congenital anomalies (5, 6). This is probably due to extremely mediastinal shift and malrotation of carina (7).

UPAA classified into three groups.

1. Agenesis, with complete absence of lung and bronchus and its supporting vasculature.
2. Aplasia, with primitive bronchus and complete absence of pulmonary parenchyma.
3. Hypoplasia, with presence of variable amounts of bronchial tree, pulmonary parenchyma, and supporting vasculature (8, 9).

Patients with UPAA may have a normal pulmonary trunk and unilateral absence of supporting pulmonary artery. Pulmonary trunk may develop normally but due to inadequate vascular supply from the bronchial vessels, resulting in a small lung at the affected side (2).

Symptoms may include dyspnea, recurrent pulmonary infections and chest pain. The anomaly may be presented by life-threatening hemoptysis (10-12).

Clinical examination is non-diagnostic, except from the decreased sounds in the affected lung. Pulmonary function tests reveal a mild restrictive pattern with normal single breath diffusion capacity (13).

A number of imaging techniques are available for diagnosis. CXR findings may include cardiac and mediastinal displacement, absence of the pulmonary artery shadow on the affected side, an ipsilateral elevation of the diaphragm and mediastinum shift as well as, a contra-lateral compensatory hyperinflation of the hemithorax, and herniation across the midline. Pulmonary CT angiography and digital subtraction angiography are the gold standard diagnostic tests (13, 14).

Common causes of death include right heart failure, respiratory failure, massive pulmonary hemorrhage and severe pulmonary edema (5. 15).

Treatment of UPAA comprises surgical, pharmacological and behavioral management. Pneumonectomy and surgical revascularization are considered in cases of recurrent pulmonary infections, life threatening hemoptysis and pulmonary hypertension (15).

We presented a case of UPAA with chief complaint of chronic cough. Our patient was a female child with right sided pulmonary agenesis without any cardiac, vertebra and rib malformation.

She was asymptomatic until 2 years ago, other symptom of UPAA such as dyspnea, chest pain and hemoptysis was not reported. A decreased right hemithorax sounds was noticed in physical examination. Despite bronchial reversibility at PFT, due to lung volume reduction secondary to right pulmonary absence, forced vital capacity (FVC) was decreased and the ratio of forced expiratory volume in one second (FEV1) to FVC increased and the PFT pattern presented as a restrictive pattern. Bronchoscopy was not done due to patient refusal.

Diagnosis UPAA as a rare disease from other etiology may save patient's life and decrease complication resulting from inappropriate treatment.

Conclusion

Although UPAA is a rare congenital anomaly, in cases of decreased unilateral lung sound, this disorder should be considered as one of differential diagnosis. Furthermore, in patients with clinical suspicious of UPAA, CXR and CT angiography must be done to confirm or exclude the diagnosis.

Conflict of Interest

Authors declare there was not any conflicting interest.

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