Pulmonary Artery Agenesis: A Rare Case Report

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Abstract

Pulmonary artery agenesis is one of the rare anomalies of unknown etiology and approximately 50% can be associated with congenital malformations such as cardiovascular, gastrointestinal, neuro-musculoskeletal and genitourinary system. Pulmonary artery agenesis is a rare congenital anomaly in neonatal period or later in life. It often causes respiratory distress syndrome in the neonatal period. Differential diagnosis includes total atelectasis, congenital diaphragmatic hernia, congenital cystic adenomatoid malformation, pulmonary sequestration, chylothorax, pneumothorax, bronchogenic cyst and soft tissue tumors. Computed tomography is the preferred imaging method for diagnosis. Prognosis depends on the presence of other concomitant anomalies and functional capacity of the lung. We detected right pulmonary artery agenesis in a patient with recurrent lung infection. Because of the vital importance of early diagnosis in these patients, we aimed to draw attention to such variation diseases.

Keywords: Pulmonary artery agenesis; Child; Recurrent pneumonia

Introduction

Pulmonary artery agenesis is one of the rare anomalies of unknown etiology. It was first described by D. Pozze in 1673 during a woman's autopsy [1]. Bilateral pulmonary artery agenesis is incompatible with life. Genetic and teratogenic factors have been implicated in the unilateral pulmonary artery agenesis. Approximately 50% of pulmonary artery agenesis may be associated with congenital malformations such as cardiovascular, gastrointestinal, neuro-musculoskeletal and genitourinary system [2,3]. The diagnosis of unilateral pulmonary artery agenesis can be made by various radiological imaging methods such as posteroanterior and lateral lung direct radiography, computed tomography (CT), magnetic resonance (MR), bronchography and angiography. It is common opinion that the majority of isolated unilateral pulmonary artery agenesis is infant-child and is a very rare congenital pathology [1]. In this disease, it does not contain a specific hereditary component but in some case reports it has been shown that there are different chromosomal abnormalities [2]. It is thought to be caused by the disturbance of pulmonary root and aortic arch connection during embryological development [3]. Before the development of the pulmonary artery, the blood circulation of the lung is provided by the transitional branches of the dorsal aorta, which are collaterals, and then regressed. These branches are replaced by enlarged supradiaphragmatic and infradiaphragmatic aortic branches or bronchial arteries as in cases where there is no pulmonary artery development [1]. It is frequently associated with abnormal lung development and cardiovascular abnormalities in its pathophysiology. Left pulmonary artery agenesis is more rare than right. Coexistence of cardiovascular anomalies such as fallot tetralogy, ventricular septal defect and right arch aorta is common among congenital heart defects [3]. In this article, we present a rare case of right pulmonary artery agenesis without any additional malformation.

Case Presentation

The fourteen months old girl sick, she presented with complaints of spontaneous fever, rapid breathing and decreased appetite. Oral clarithromycin was started at the external center. He was born 39 weeks, 2600 g, normal vaginal delivery. There was no need for any intervention in the delivery room. No respiratory distress. Immediately fed with breast milk, Meconium was released in the first 24 h. No newborn jaundice. At admission, the vaccines are compatible with the age. Two times a day, a thick, green poop was doing. He had a history of hospitalization for pneumonia twice at 9 and 12 months of age. His appetite had decreased for two months and oral formula was started. There was no hospitalization in the neonatal period and pediatric intensive care unit. There was no trait in the family history. In physical examination: Height: 78 cm 50 p weights: 7.6 kg 1 p to 3 p pulse 139/min, respiratory rate 46/min, and blood pressure 100/60 mmHg and oxygen saturation...
was 89%. The patient’s breathing was tachypnea. He had bilateral diffuse crepitant, rales and wheezing. The other system examination was normal. In laboratory tests, Hb: 9.4 g/dL, white blood cell: 13900/mm³, platelet: 284000/mm³, there were no atypical cells in peripheral smear, platelets were abundant and clustered, and erythrocytes were normochromic microcytes. Sedimentation: 65 mm/h, C-reactive protein: 81.9 mg/L, pH 7.36 in blood gas, PaCO₂: 32 mmHg, PaO₂: 98 mmHg, Hco₃: 17.7 mmHg. Skin prick test was negative. Food specific IgE: negative. The TORCH panel was negative. Immunological values were normal according to the age of the patient. BCG scar was present. Nasopharyngeal aspirate PCR showed rhinovirus and enterovirus positivity. Cold agglutination test: Negative. Presence of perihilar and paracardiac infiltrations on postero-anterior and lateral chest radiographs (Figure 1A and 1B) and thoracic computed tomography revealed parenchymal infiltrations in the upper lobe posterior and lower lobe, ground glass attenuation in the left lung, right lung deviation, right lung deviation and right pulmonary artery agenesis (Figure 2A-2C). Thorax ultrasound and oesophageal gastric duodenum graphy were normal. Electrocardiography and ECO water were normal. The patient was admitted to the pediatric chest diseases department with the diagnosis of right pulmonary artery agenesis and pneumonia. Free flow oxygen, inhaler therapy and antibiotherapy were started. Oxygen saturation was normal, respiratory distress did not develop and no complication was observed. The patient whose symptoms regressed was discharged with healing by prescribing inhaler treatment with the suggestion of polyclinic. The patient came to the outpatient clinic for control. No complaints. Routine examination and examinations were normal.

**Discussion**

Pulmonary artery agenesis is one of the rare anomalies of unknown etiology. The age of diagnosis of the reported cases ranged from one month to 58 years. The reason for delay in diagnosis is due to the lack of symptoms in some cases and non-specific findings in symptomatic cases [4,5]. In some patients, as in our case, recurrent lung infections, dyspnea or congestive heart failure may occur with findings such as [3,5]. Pulmonary hemosiderosis and pulmonary hypertension have been described in cases developing. In fact, recurrent lung infections can be seen in 40% of patients [4]. The mortality rate due to this disease is 7% [3]. Absence of pulmonary artery and hypoplastic lung should be separated from Swyer-James-MacLeod syndrome by perfusion scintigraphy or pulmonary angiography [6,7]. In Swyer-James-MacLeod syndrome, pulmonary artery is hypoplastic and perfusion cannot be observed. In the absence of unilateral pulmonary artery, perfusion is observed in the peripheral periphery of the lung due to collaterals, although it is not observed on the side with pulmonary artery involvement [7]. Since pulmonary artery agenesis is a rare disease, the treatment approach is different [8]. Treatment options include surgical resection of the lung lobe and endothelin receptor

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**Figure 1A:** View of perihilar and paracardiac infiltrations on postero-anterior chest X-ray.

**Figure 1B:** The appearance of perihilar and paracardiac infiltrations on lateral chest X-ray.

**Figure 2A:** CT scan of the pulmonary arteries showed mediastinal sections and left main pulmonary artery and its branches in normal calibration. Right main pulmonary artery is not observed from the outflow.

**Figure 2B:** CT scan for pulmonary arteries showed mediastinal sections and left main pulmonary artery and its branches in normal calibration. Right main pulmonary artery is not observed from the outflow.

**Figure 2C:** CT scan of the pulmonary arteries showed mediastinal sections and left main pulmonary artery and its branches in normal calibration. Right main pulmonary artery is not observed from the outflow.
antagonists, prostacyclin, and nitric oxide as medical treatment and in most cases are followed by symptomatic treatment [9]. Most cases are diagnosed in the first year of life and surgical treatment is performed if indicated [4]. The opinion that there is no need for treatment is dominant in asymptomatic cases [5]. As in our case, a congenital pathology should be considered on the basis of recurrent lung infections and necessary investigations should be performed for suspected cases [10-13].

References

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