PULMONARY AGENESIS: A CASE REPORT

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ABSTRACT

Pulmonary agenesis is a very rare congenital anomaly. Clinical symptoms vary depending on the other anomalies and severity of pulmonary hypoplasia. Although it is usually diagnosed in infancy and childhood, some cases do not show any symptoms until adolescence. Diagnosis in adulthood is much rarer. Our case is rarer as it is presented in the adolescent age group.

KEYWORDS

Pulmonary Agenesis, Pulmonary Aplasia, Pulmonary hypoplasia, VACTERL (Vertebral, Ano-Rectal, Cardiac, Trachea-Oesophageal, Renal and Lung) Anomalies.

INTRODUCTION

Bilateral pulmonary agenesis is incompatible with life. Unilateral agenesis is seen in 34 per 1 million live births. It is frequently associated with other congenital anomalies. The condition was first discovered accidentally at the autopsy of an adult female in 1673, by De Pozze. In India the first case was reported by Muhamed in 1923, of a left sided pulmonary agenesis in a medico-legal autopsy. We are here presenting a rarest of rare case of a unilateral agenesis as about 200 cases so far are reported.

CASE REPORT

A 16-year-old female patient presented with progressive shortness of breath of one week and frequent episodes of cough with mucopurulent sputum for 10 days. General examination revealed pallor, low grade fever and tachypnoea. On clinical examination of the chest, accessory muscles of respiration were working, drooping of shoulder was seen on the left side and scoliosis with convexity to the right was noticed. Intercostal suction was also seen. On palpation, movement was diminished on the left side with crowding of ribs, trachea was deviated to the left side and the apex beat was noted in the left 6th intercostal space in mid axillary line.

Vocal fremitus diminished throughout the left side. On percussion, left side had impaired note in the 7th intercostal space downward along mid axillary line and scapular line, it was resonant in the rest of the areas. On auscultation, bilateral vesicular breath sounds were heard. Reduced vocal resonance noted on the left side. Bilateral coarse crepitations were heard in the inter and infrascapular areas and right axillary region. Chest X-ray revealed an opaque left hemithorax with signs of volume loss and compensatory hyperinflation of the right lung [Figure 1]. A Computed Tomogram (CT) of thorax, virtual bronchoscopy, bronchogram, pulmonary angiogram were performed and a diagnosis of Type 1 lung agenesis was made.
DISCUSSION

Pulmonary agenesis is a rare congenital anomaly with a reported prevalence of 34 per 1 million live births.\(^1\) It is frequently associated with other congenital anomalies. The condition was first discovered accidentally at the autopsy of an adult female in 1673, by De Pozze.\(^2\)

In India, the first case of Pulmonary agenesis was reported by Muhamed (In 1923), of a left sided pulmonary agenesis in a medico legal autopsy.\(^3\)

Clinical symptoms vary depending on the other system anomalies and severity of pulmonary hypoplasia. Although it is usually diagnosed in infancy and childhood, some cases do not show any symptoms until the adolescent ages. Diagnosis in adulthood is very rare. Schneider classified agenesis into three groups, which has been subsequently modified by Boyd.\(^4,5\)

Depending upon the stage of development of the primitive lung bud, pulmonary agenesis is classified into three categories:

Type 1 (Agenesis): Complete absence of lung and bronchus and no vascular supply to the affected side.

Type 2 (Aplasia): Rudimentary bronchus with complete absence of pulmonary parenchyma.

Type 3 (Hypoplasia): Presence of variable amounts of bronchial tree, pulmonary parenchyma and supporting vasculature.

Our patient is classified as Type 1 pulmonary agenesis. Nearly 50% cases of pulmonary agenesis have associated congenital defects.\(^6\) Associated anomalies include the VACTERL syndrome (Vertebral segmentation anomalies, anorectal atresia, tracheo-oesophageal fistula, oesophageal atresia, radial ray and renal anomalies).\(^7\) Left sided agenesis is more common and these subjects have a longer life expectancy than those with right sided agenesis.\(^8\) Pulmonary aplasia (Agenesis) is thought to result from the negative effects that occur on the 4th week of foetal life. Although its aetiology is not fully understood, vitamin A or folic acid deficiency or the use of salicylates may be responsible for it.\(^9\) In our case, the diagnosis was made with non-invasive methods such as Chest radiograph and CT thorax.

On chest radiograph, mediastinum was found to be shifted to the left with intercostal narrowing and elevated hemidiaphragm were seen (Figure 1). In our case HRCT (High Resolution Computed Tomography) revealed rudimentary left main bronchus (Figure 2).

The NCCT (Non-Contrast Computed Tomography) confirmed chest x-ray findings (Figure 3). It revealed normal right pulmonary artery however left pulmonary artery was absent (Figure 4). No lung parenchymal tissue was detected on the left side. Heart and the main vascular structures were noted in the left hemithorax (Figure 4 & 5).

To conclude we would like to say that pulmonary agenesis is a rare congenital abnormality in which there is unilateral or bilateral absence of lung tissue. It has to be differentiated from pulmonary aplasia, a similar condition, the main difference being that there is a short-blind ending bronchus in aplasia.

REFERENCES