

Unilateral left pulmonary agenesis with the presence of bilateral accessory lobes of lung

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Abstract

Lobar agenesis is uncommon anomaly of lung. It presents with varied clinical manifestations. The manifestation of agenesis depends on what stage of development the growth of lung is arrested. It can be bilateral complete, unilateral or lobar type. In present case, in routine cadaveric dissection we found unilateral lobar agenesis on one side (left) and marked hyperplasia of lung on the opposite side (right). The mediastinal space was extremely narrow and not seen before the removal of lung. Heart was not present in the middle mediastinum. It was more to left side and was lodged on the concavity formed by the anterior surface of the left lung. Left lung had no fissure and overlying parietal pleura was inseparable. Right lung had only one fissure which divided the lung into two lobes. We also found presence of accessory lobes of lung on both side. There were two accessory lobes (one above and one below bronchus) on left side and one accessory lobe on right side.

Key Word: lobar agenesis, lung hypoplasia, accessory lobes

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INTRODUCTION

Lungs are the major part of pulmonary circulation. The space between the two lungs is called as mediastinum. In the middle mediastinum heart is located. Normally right lung has two fissures (one oblique and one horizontal) and divided into three lobes while left lung is having single oblique fissure which divides the left lung into two lobes. The root of right lung consists of two bronchi, one pulmonary artery and two pulmonary veins (superior and inferior) while the root of the left lung consist of only one bronchus, one pulmonary artery and two pulmonary veins.^{1,2} Lobar agenesis is an uncommon lung anomaly. When it occurs in both the lungs called as Bilateral type while when only one lung is involved it is called as

Unilateral.³ According to Vetraino *et al*³ Bilateral lobar agenesis is incompatible with extra uterine life while in unilateral agenesis mediastinal contents are shifted to hypo plastic side and the opposite lung is hyper extended.³ The prevalence of pulmonary agenesis along with other congenital anomalies reported among hospital admissions is 0.0034.⁴ Our present case is of unilateral lobar agenesis in which, left sided lung is hypo plastic without any fissure. Heart was shifted more to the left side and was lodged on concavity formed by anterior surface of left lung While on the right-side hyper plastic lung was present with single oblique fissure dividing it into two lobes. Mediastinal space was not seen before the removal of lungs. In addition, there were accessory lobes present two on the left side and one on the right side. Pulmonaryagenesis is having varied aetiology (maternal and foetal factors). The manifestation of agenesis also varies. It can be the cause of recurrent respiratory infections in children. Sometimes it remains asymptomatic throughout the life and found out as autopsy finding. Lung agenesis has definite embryologic basis and in most of the cases (60%), it is associated with other anomalies.⁵ Though rare, clinical manifestation of this anomaly makes it significant. Present case is the case of unilateral lobar agenesis with bilateral accessory lobes.



Figure 1:

Figure 1: Image showing hyperplastic right lung with two lobes and hypo plastic left lobe;



Figure 2:

Figure 2: Image showing hyperplastic right lung with one accessory lobe and hypo plastic left lung with two accessory lobes

DISCUSSION

In the routine cadaveric dissection, we found two malformed lungs in a male cadaver of 25 years. The left lung was hypoplastic while the right lung was hyperplastic and mediastinal contents were shifted to hypoplastic side(left). Pulmonary agenesis has embryologic basis. When respiratory bud fails to develop, agenesis of lung can occur.^{6,7} In case of aplastic or hypoplastic lung on one side, hyperplastic lung on other side is found. According to Dobremez *et al*⁸ whenever lobar agenesis is there, maximum times (70%) it is on left side. There is no sex predilection in the incidence of agenesis.³ Lung hypoplasia can be primary or secondary. Primary is of unknown aetiology while secondary is due to some maternal and foetal factors. Our case is of unilateral left lung agenesis with bilateral accessory lobes. Our findings match with the study by Bashir Ahmed³. Bashir Ahmed³ found hypo plastic left lung with single lobe without any fissure. In our case, the left lung was inseparable from the parietal pleura covering it and this hypo plastic lung was having single lobe without any fissure. Root of this lung showed single pulmonary artery and vein. Anterior border was straight without any cardiac notch. Lingula was not obvious. Anterior surface of this lung was concave forming concavity on which heart was present. When tracheobronchial tree was observed in the left lung only one principal bronchus was found. Pleura covering this left lung was very strongly adherent to underlying lung tissue and was inseparable from it. We also found the presence of accessory lobes of lung on both the sides. Two accessory lobes were present, one above and one below the principal bronchus in the left lung. These accessory lobes were not supplied by any bronchus. Right lung was hyperplastic with only two lobes separated by single oblique fissure. Fissure was not complete. Horizontal fissure was absent. Anomalies of fissure are not uncommon. Okamoto K⁹ in his case report mentioned the presence of anomalous horizontal fissure which was incomplete and deep and was not reaching to the posterior surface. Ueyama *et al*¹⁰ also reported a case

with two aberrant fissures. One accessory lobe was present on the right side which may be hypoplastic middle lobe. We observed one pulmonary artery and two pulmonary veins in the root of this lung. When tracheobronchial tree was observed in right lung we found three principal bronchi. Out of these three, two were for hyperplastic lower lobe and single one was for upper lobe. Lobar agenesis can occur due to decreased thoracic cavity as found in cases of Diaphragmatic hernia. The herniated contents also compress the lung resulting in decreased fluid volume. Therefore, such cases are also associated with other anomalies like renal agenesis, oligohydramnios and other genitourinary anomalies.^{11,12} In our case, when lungs were removed and thoracic cavity was observed, right sided part of thoracic cavity from the midline was larger than the left part. Accessory lobes which were found in our study were not supplied by any bronchus both in the right and left lung. The presence of such accessory lobe was also mentioned by Okamoto K⁹ in one case report where this lobe was described as some gland like structure representing hypo plastic middle lobe by author. Bergman¹³ *et al* also mentioned similar case and described accessory lobe as vestigial middle lobe. Spencer (1977) modified the earlier given classifications of pulmonary agenesis. Presently the classification given by Boyden (Schaffer, 1960) is followed who divided pulmonary agenesis into following types three groups.¹⁴ Group 1: The lung its pulmonary artery and bronchus are absent. Group 2: Absent lung and pulmonary artery with a rudimentary bronchus coming off from trachea. Group 3: Hypoplastic lung with pulmonary vessels and a fully formed bronchus. Our case as per the above classification comes under Group –3- showing hypoplastic left lung with pulmonary vessels and fully formed bronchus. In addition, there was presence of bilateral accessory lobes. Though uncommon, the presence of such anomaly can lead to varied manifestations from asymptomatic to fatality and death. Pulmonary agenesis is also associated with cardiac and renal anomalies. The prognosis of such cases depends on the presence of associated congenital

anomaly. The Commonest cardiac malformation associated with pulmonary agenesis is the patent ductus arteriosus which was not found in the present case.³ In paediatric age group it can also present as a cause of recurrent respiratory infection. Hence the knowledge of such anomaly is significant.

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