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"PULMONARY AGENESIS" EMBRYOLOGICAL BASIS AND ITS CLINICAL IMPORTANCE

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ABSTRACT

Deficiency of the lungs results from malfunction of the breathing bud to develop. Agenesis of one lung is more regular than bilateral agenesis, however both conditions are unusual. Unilateral pulmonary agenesis is compatible with life. The heart and other mediastinal edifices are shifted to the implicated side, and the present lung is stretched out. Lung agenesis is an uncommon but a severe condition which ought to be early diagnosed and followed up. Prognosis, apart from abnormal formation, is that of a single functional lung. Diagnosing a developmental abnormality of the lungs in adults is often challenging for clinicians.

Introduction

Pulmonary agenesis appears as an obscure hemithorax on the affected side. There is a mediastinal swing into the implicated side, and compensatory hyperinflation of the residual lung. In the right lung agenesis, malposition of the aortic arch and descending aorta are the reason of the extreme dextroposition consequential from the absence of the right lung. The distal trachea is normally compressed and deformed posteriorly by the dextroposal crossing aortic arch. The left mainstem bronchus is also recurrently compressed between the engorged solitary left pulmonary artery anteriorly and the posterior malpositioned descending aorta. The ipsilateral lung parenchyma, blood vessels, and airways are deficient at CT (computed tomography).

Imaging results in pulmonary aplasia and agenesis are comparable, except for the presence of a short blind-ending bronchus in aplasia. Postnatal radiography display diffuse opacification of the implicated hemithorax with ipsilateral mediastinal shift, and computed tomography helps authenticate the absence of the lung parenchyma, bronchus, and pulmonary artery on the implicated side [Abel et la., 2006].

Incidence

The congenital anomalies are usually the cause of deaths in infants under the age of one year [Ajit e.t, 2014; Ganesh Elumalai and Sushma chodisetty, 2016]. Pulmonary Agenesis is a very uncommon congenital anomaly representing failure of development of the primal lung bud or complete absence of the lung parenchyma, bronchus and pulmonary vasculature. Approximately 1 in 15 000 children are born with innate absence of one lung and the linked bronchus. Unilateral pulmonary agenesis most possibly results from an in-utero insult during the 4th week of gestation.

It has been theorized that irregular blood flow in the dorsal aortic arch during the 4th week of gestation (embryonic phase) causes pulmonary agenesis. Pulmonary agenesis occurs with one and the same incidence on the left and right. However, right sided agenesis is associated with a much shoddier prognosis due to greater anatomic deformation of the airway and great vessels, intermittent infections and tracheobronchomalacia. Associated anomalies are regularly found, in more than 50% of implicated fetuses, and that include congenital heart disease, vertebral abnormalities, imperforate anus, renal agenesis and tracheoesophageal fistula.

Ontogenesis of the normal development of the bronchi and lungs

Development of the lungs start on day 22 with formation of a ventral outpouching of the endodermal foregut called the respiratory diverticulum. The respiratory bud that developed at the caudal end of the laryngotracheal diverticulum during the fourth week soon divides into two outpouchings, the primary bronchial buds. These buds grow laterally into the pericardioperitoneal canals, the primordial of the pleural cavities. Secondary and tertiary bronchial buds soon develop. Along with the surrounding splanchnic mesenchyme, the bronchial buds differentiate into the bronchi and their ramifications into the lungs. Early in the fifth week, the connection of each bronchial bud with the trachea enlarges to form the primordial of main bronchi. The embryonic right main bronchus is slightly larger than the left one and is more vertically oriented. This embryonic relationship continues in the adult; as a result, a foreign body is more liable to enter the right main bronchus than the left one. The major bronchi give rise to secondary bronchi that form lobar, segmental, and intrasegmental branches. On the right, the superior bronchus will supply the upper lobe of the lung, whereas the inferior bronchus subdivides into two bronchi, one is to the middle lobe of the right lung and the other one to the lower (inferior) lobe. On the left, the two secondary bronchi supply the upper and lower lobes of the lung. Each lobar bronchus undergoes progressive branching. The segmental bronchi, 10 in the right

lung and 8 or 9 in the left lung, begin to form by the seventh week. As this occurs, the surrounding mesenchyme also divides. Each segmental bronchus with its surrounding accumulation of mesenchyme is the primordium of a bronchopulmonary segment. By 24 weeks, approximately 17 orders of branches have established and respiratory bronchioles have developed. An additional seven orders of airways develop after birth.

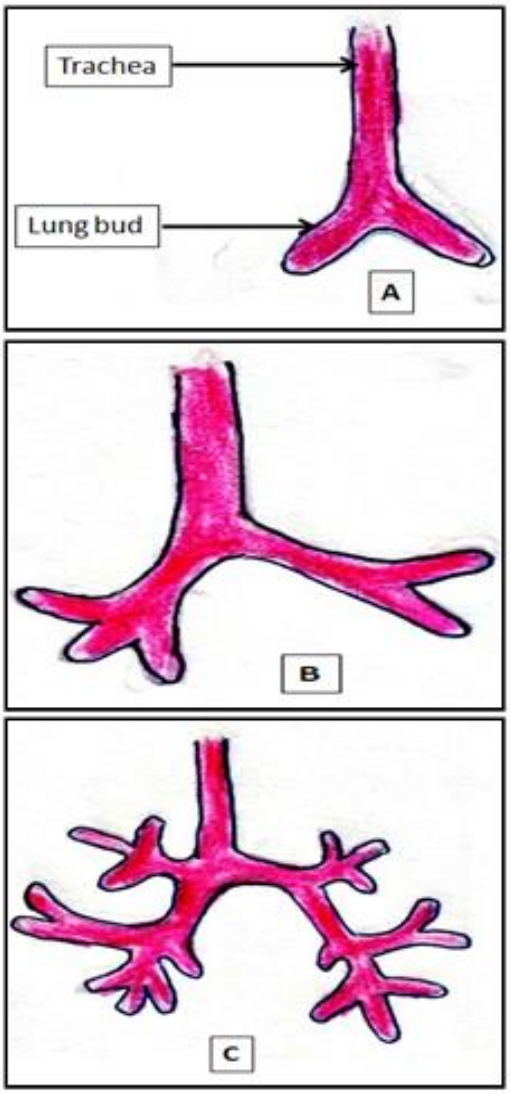

Fig 1: Development of lung buds. (A) Right and left buds begin to appear. (B) They divide into lobar bronchi (three on right side and the two on the left side). (C) Segmental bronchi are established.

Ontogenesis for the pulmonary agenesis

Absence of fissures that are normally present leads to a reduction in the number of lobes, for example, absence of the transverse fissure of the right lung results in a right lung with only two lobes. A transverse fissure may be present on the left side with the result that the left lung has three lobes (A), and the medial basal segment of the left lung may be separated by a fissure from the rest of the lower lobe (B). the superior segment of the lower lobe may be similarly separated(C) and also, a part of the upper lobe of the right lung may come lie medial to the azygos vein (D). This part is called azygos lobe. Accessory lobes are usually connected to bronchi that are not part of the normal bronchial tree. Such bronchi may rise from trachea above its bifurcation and oesophagus. Occasionally, the lobe may not have any bronchus. An area of embryonic lung tissue may separate from tracheobronchial tree. Such tissue may form a complete lobe, which may have an independent pleural covering.

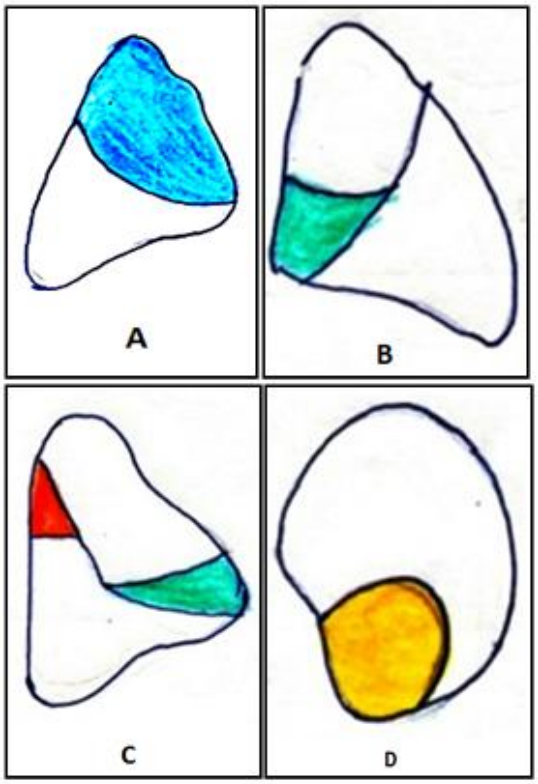

Fig 2: Abnormal lobes of the lungs: (A) Right lung with only two lobes. (B) left lung with three lobes. (C) Apical segment of lower lobe is separate. (D) Medial segment is separate.

Discussion

Lung agenesis is an enormously rare abnormality whose commonness is estimated at 34 per million live births. The true incidence of this abnormality is unidentified because 50% of cases are stillborn and more than 20% die at birth or during their first few months. Right agenesis is rarer but more severe than the left, probably because of an excessive carina shift and an improper drainage of the functioning lung. Lung agenesis is associated in 50 -75% with other malformations, especially cardiovascular, gastrointestinal, urogenital and osteoarticular ones. Mortality is higher and earlier in these cases, generally within the first 5 years of life. Pulmonary agenesis can occur between the 4^{th} and 5^{th} week of gestation in the embryonic phase, before the pseudo glandular period, when the primitive lungs are forming as a diverticulum protruding from the foregut. The precise etiology of this circumstance is mysterious, but hereditary factors, viral agents, and dietary insufficiency of vitamin A throughout pregnancy are suspected to be accountable. It is hypothesized that ether a simple arrest of development occurs, bilateral agenesis, or there is malfunction to maintain the developmental balance of the two lung buds. Out of the two lungs, the left lung is affected more often, with a male high proportion. Nevertheless, many reports have recognized that right-sided agenesis is typically associated with other congenital anomalies, mainly cardiac malformations, and because of these manifold abnormalities most patients pass on within the first year of life. The diagnosis of agenesis of the lung is habitually alleged from the chest radiograph, which commonly reveals a homogeneous opacity possessing most of the

implicated hemithorax and displacement of the mediastinal contents into the empty hemithorax.

In certain cases the opacity may occupy mainly the middle and lower zone on the right side, along with a shift of the mediastinal contents towards the pretentious side. Differential diagnosis on X-ray include collapse, destroyed lung, post-pneumonectomy, thickened pleura, and agenesis.

Concerning management, cases with no symptoms need no interference, but prevention of infection in the introverted lung is of principal importance. Chest infections of the introverted lung can be life-threatening and should be treated without delay and aggressively with antibiotics, bronchodilators, and physiotherapy. Cases with no symptoms and patients with minimal symptoms have good prognosis. Clinical presentation is outstandingly variable. Lung agenesis is often revealed in childhood after recurring pulmonary infections, or as part of polymalformative syndrome. The absence of lung parenchyma is not typically evident at the x-ray chest because of a controlateral upper lobe hypertrophy crossing the midline. The resultant image of basithoracic opacity may mimic lobar collapse, pleural effusion, or diaphragmatic hernia.

Conclusion

Lung agenesis is an uncommon but a severe condition which ought to be early diagnosed and followed up. Prognosis, apart from abnormal formation, is that of a single functional lung. Pneumothorax occurrence is treacherous and should be surgically treated. Pulmonary agenesis of the right side, without any other congenital anomaly, is extremely rare. Diagnosing a developmental abnormality of the lungs in adults is often challenging for clinicians.

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