PULMONARY AGENESIS AND BRONCHIAL ATRESIA

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The development of the lung follows the pattern of a tubular-acinous ductal system. Disturbance of the developing pulmonary anlage in early embryonic life may give rise to some uncommon anomalies like pulmonary agenesis or in very rare occasions, atresia of a segment of the respiratory tract associated with development of normal pulmonary tissue distal to the atresia. This paper reports on a case of pulmonary agenesis and a case of bronchial atresia.

CASE REPORTS

Case 1

A full-term, fresh, male stillborn was delivered by a Chinese woman A.N.T. on 14.6.1966 in Kandang Kerbau Hospital. The pregnancy, labour and peuperium were normal.

NECROPSY

The body was that of an achondroplastic male Chinese infant weighing 3175 gms. The right lung and pleura were absent and the right thoracic space was occupied by the enlarged heart and loose connective tissue (Fig. 1). The trachea was continuous with the left bronchus which led to a non-expanded left lung. The right bronchus was absent (Fig. 2).

The left lung was supplied by a normal sized pulmonary artery, but the pulmonary vein drained into the coronary sinus via the left superior vena cava. The right pulmonary artery was hypoplastic and ramified among the loose connective tissue in the right thoracic space.

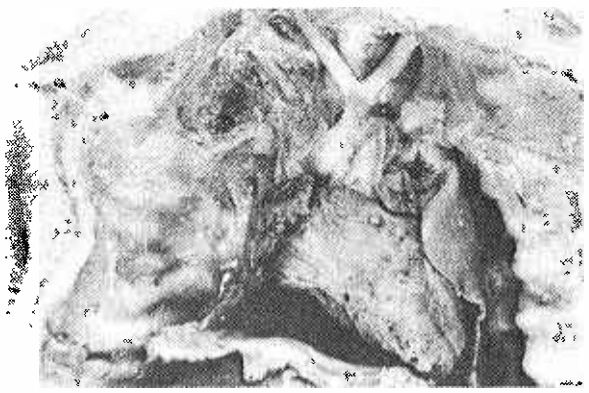

Fig. 1. Absence of right pleural space and right lung. The right thorax is occupied by the enlarged heart and some loose connective tissue.

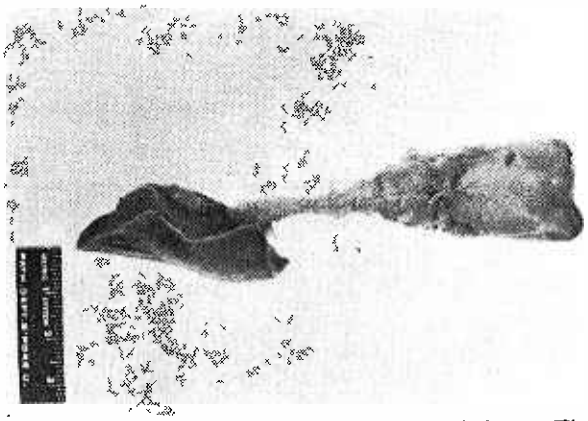

Fig. 2. Absence of right bronchus and lung. The trachea continues into the left bronchus.

Other findings were omphalocoele, incomplete fusion of the facial processes, adactyly, A-V communis with hypoplastic left atrium and atretic mitral valve, polycystic kidneys and hydranencephaly.

Case 2

L.K.H. a 36 year old Chinese woman was first seen on 3.3.1967 in the Antenatal Clinic, Kandang Kerbau Hospital, at 32nd week of her eighth pregnancy. She was anaemic with a haemoglobin level of 8·5 gm%. A mild hydramnios was detected which became progressively worse. Roentgenogram revealed an anencephalic foetus. She delivered a fresh female stillborn on 26.4.1967 uneventfully, 21 hours after artificial rupture of membrane when about 5 litres of clear liquor were drained.

NECROPSY

The body was that of an anencephalic female infant weighing 2380 gms. The larynx and the trachea were normal. The right main bronchus ended blindly one cm. from the carina and was connected by loose connective tissue to the hilar region of the right lung. This lung was large, single lobed, spongy, airless and was supplied by a normal sized pulmonary artery and drained by a large pulmonary vein (Fig. 3). On cutting the hilium of the right lung, a large bronchus was found arising blindly (Fig. 4). Dye injection revealed that this bronchus gave rise to three branches. resembling the distribution of a normal right upper lobe (Fig. 5).

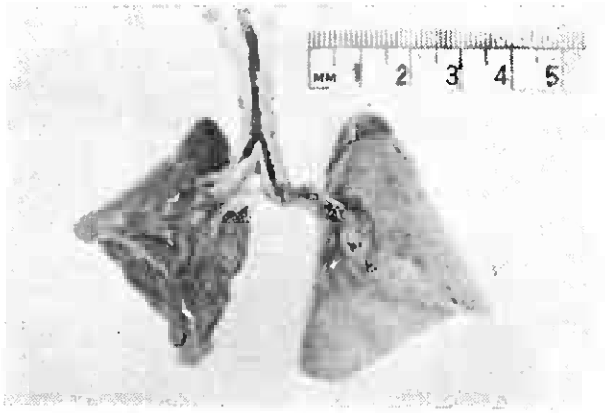

Fig. 3. Atresia of the right main bronchus. The right main bronchus ends blindly about one cm. from the carina. The bronchial stump is connected to the hilar region of a large monolobed right lung by loose connective tissue.

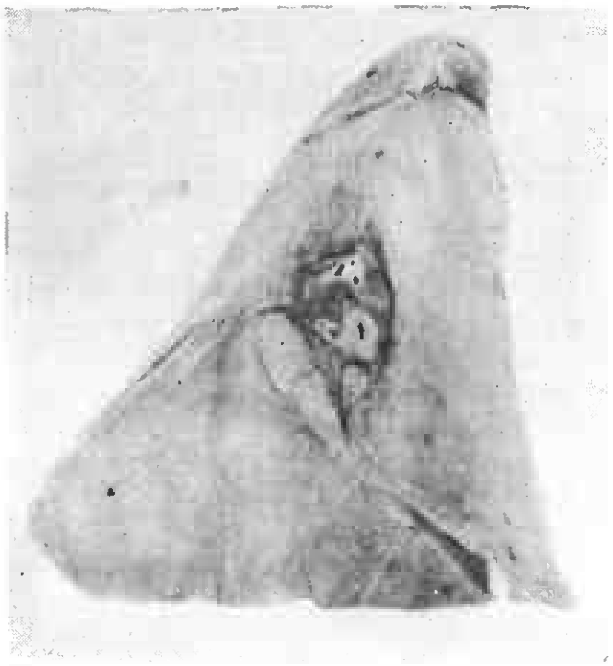

Fig. 4. Hilar region of the right lung cut to expose the pulmonary artery and vein and the bronchus which arises blindly.

The left bronchus was normal. The bilobed left lung was completely atelectatic.

Other gross findings were bilateral adrenal hypoplasia and unicornuate uterus.

Histology

The trachea and main bronchi were lined by ciliated columnar epithelium. Sections of the proximal right main bronchial stump showed that it ended by dividing into two branches. The lower branch bifurcated and led to two small masses of hypoplastic lung tissues. The upper branch appeared to be blocked by cartilege and ended blindly (Fig. 6 & 7). The proximal bronchial stump was connected to the hilar region of the large monolobed right lung by loose connective tissue in which no epithelial cells or glandular structures were seen.

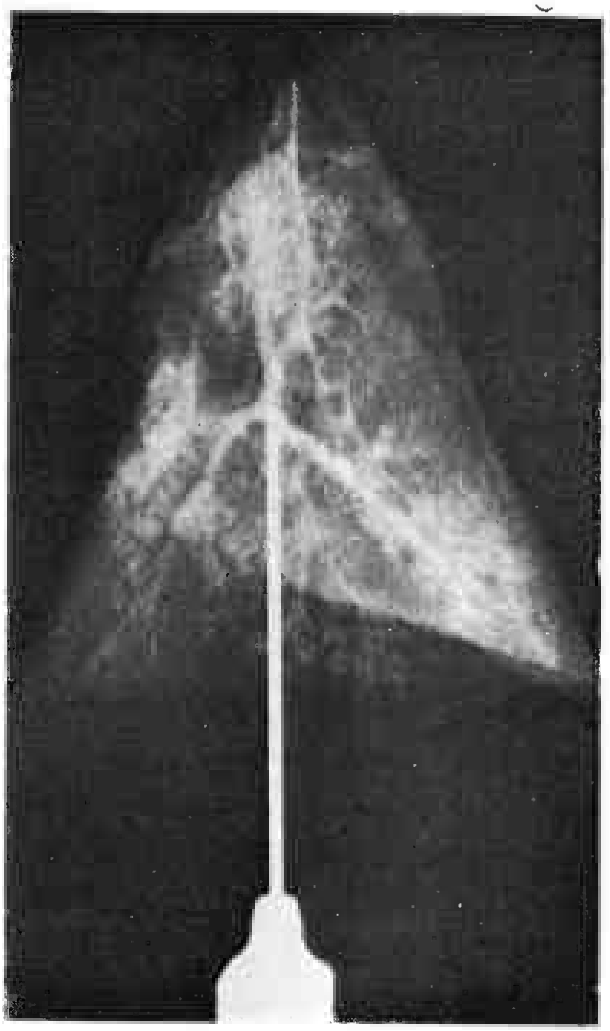

Fig. 5. X ray of the right lung after injecting urografin into the bronchus. The bronchus ends by dividing into 3 branches and resembles the distribution of normal right upper lobe bronchus.

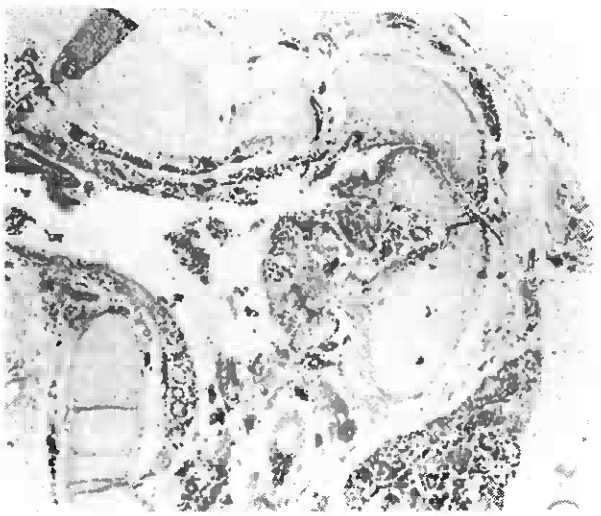

Fig. 6. Section of the right proximal bronchial stump. It ends by dividing into two branches. The upper branch appears to be blocked by cartilege H. & E. X40.

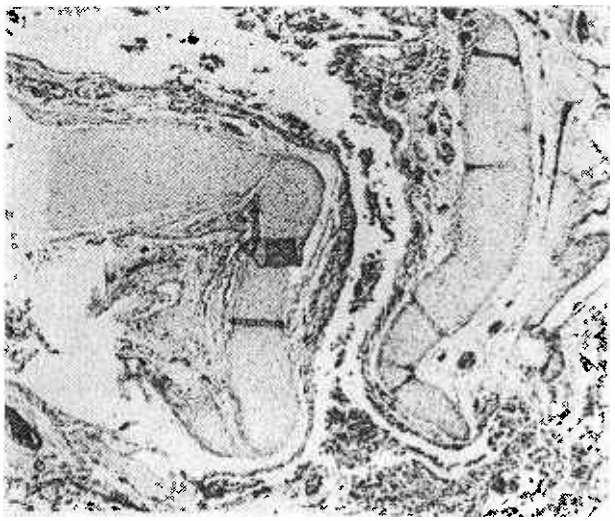

Fig. 7. Section of the right proximal bronchial stump. The lower branch bifurcates and leads to two small masses of hypoplastic lung tissues. H. & E. X40.

The large bronchus arising blindly in the right lung was lined by stratified ciliated epithelium and tall columnar epithelium with basal nuclei and clear cytoplasm. The alveoli were distended and their walls were thin. Some alveoli contained squames.

Sections of the left lung showed non-expansion of alveolar ducts.

DISCUSSIONS

The respiratory system arises as a median ventral diverticulum of the foregut in embryos at an ovulation age of about 24 days (Streeter's Horizon XI-XII). This diverticulum gives rise to right and left lung buds which elongate into the primary bronchi. The main lobar bronchi appear as monopodial outgrowths of the primary bronchi (Streeter's Horizon XIII-XV). Subsequent development consists of the repeated branching of the tubular system.

Disturbance of the developing pulmonary anlage in early embryonic life may give rise to two types of malformations:

(1) Complete arrest of development distally. Depending on the age of embryo when the disturbance occurs, various kinds of anomalies may result *e.g.* complete agenesis of tracheobronchial tract, bilateral or unilateral pulmonary agenesis. Complete agenesis of pulmonary tissue is extremely rare (Potter, 1961); Claireaux and Ferreira (1958) described a case of bilateral pulmonary agenesis and cited three previous cases.

Unilateral pulmonary agenesis, however, occurs more commonly. A review of 119 cases plus one additional original case was made by Valle (1955). Schneider (1912) divided pulmonary agenesis into three main groups: (a) Cases in which there is a complete absence of bronchus, alveolar tissue or its vascular supply. (b) Cases in which there is a rudimentary bronchus but no lung tissue. (c) Cases in which there is a poorly developed main bronchus invested in an ill developed fleshy lung tissue. The present Case 1 belongs to the first group.

(2) Atresia of a segment of the respiratory tract. These are rare anomalies. The mode of genesis of this type of anomaly is uncertain and appears to differ from that of the first type. A case of complete absence of trachea with well developed bronchi and lungs was described by Milles and Dorsey (1950). Waddell et al (1965) reported two cases and cited 3 similar cases of atresia involving a small bronchus of the left upper lobe associated with a localised area of emphysema.

Atresia of a main bronchus, as present in Case 2 has not been previously described. In the present Case 2, the right "lung" though not communicating with the exterior, is fully developed and is more voluminous than the left lung. The alveoli are thin walled and distended. The alveolar distension distal to the atresia is probably due to active secretion of fluid by the developing lung tissue; this has been shown to occur experimentally (Carmel et al 1965).

SUMMARY

A case of pulmonary agenesis and a case of bronchial atresia involving the right major bronchus, both occurring in stillborn are presented. Atresia of a main bronchus presented in Case 2 has not been previously described.

ACKNOWLEDGEMENT

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