MORPHOLOGY OF PULMONARY EXTRALOBAR SEQUESTRATION IN NEONATAL DEATH BY HYALINE MEMBRANE DISEASE

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□ *We present an unusual case of extralobar pulmonary sequestration associated with hyaline membrane disease (HMD) that caused the death of a premature baby in the first day of life. The sequestered parenchyma was nourished by an aberrant aortic vessel. Notable was the presence of typical HMD in all the lung parenchyma perfused by the pulmonary artery; the sequestered lung tissue presented a dysplastic structure compatible with CCAM. A few similar cases have been found in the literature. In all of the reported cases there are morphologic aspects characteristic of HMD in the portions normally receiving blood from the pulmonary artery. These findings suggest the importance of the blood pulmonary circulation in the pathogenesis of HMD, whose exact causes are not fully known.*

Keywords *hyaline membrane disease, pulmonary extralobar sequestration*

Although the mortality caused by hyaline membrane disease (HMD) has been decreasing and many associated factors have been identified, its pathogenesis and some causative associated agents have not yet been completely elucidated.

We present the autopsy data for a newborn with extralobar pulmonary sequestration (EPS) in which the morphologic aspects of HMD were observed in both lungs, except in the sequestered parenchyma, which was irrigated by an anomalous vessel from the aorta. To our knowledge there are only a few similar case reports [1–5] of HMD involving the whole of the lung perfused by pulmonary arteries except for the extralobar parenchyma irrigated by the systemic circulation.

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CASE REPORT

The mother was a 20-year-old woman in her third pregnancy, gestation 3, abortus 1. In the 18th week of the present pregnancy a cerclage was made because of the risk of premature delivery. Blood examination revealed blood type A, Rh positive; VDRL negative; immunoglobulin G (IgG) positive to rubella and herpes simplex; and IgG and IgM positive for cytomegalovirus. In the third trimester polyhydramnios was detected. The delivery was normal (37 weeks, 6 days), 2500 mL of amniotic fluid being collected. The female infant weighed 2080 g, adequate for gestational age. Apgar score at 1 min was 2 and after 5 min was 4. Respiratory distress with gasping was present immediately after birth and accentuated until the infant died at 15 h. Chest X-rays demonstrated a diffuse hypotransparency, impeding cardiopulmonary individualization. A hemogram evinced a total count of 17,000 leucocytes, 54% of erythroblasts and a normal differential.

PATHOLOGIC FINDINGS

Autopsy was performed the day after death. Gross examination disclosed a white female infant without external anomalies, weighing 2180 g, with a total length of 47 cm, head 31 cm, and foot length 6 cm, compatible with 31 weeks. There was moderate hydrothorax, small dark red lungs, and a shift of the mediastinum to the right; the right lung weighed 38.2 g. The left lung was compressed by a soft, pink mass, measuring 6 × 5 cm, located in the left posterior costodiaphragmatic angle; it was not weighed. It was well encapsulated, presenting a spongy pink uniform parenchyma resembling normal lung tissue. This mass had no communication with the tracheobronchial tree or esophagus, receiving blood supply from the abdominal aorta through a vessel that crossed a normal diaphragm (Figures 1 and 2).

MICROSCOPIC EXAMINATION

A hematogenous placentitis was evidenced. It was represented mainly by vascular lesions involving all the fetal vascular circuit, villous dysmaturity, necrotic villitis, intervillitis, and lymphoplasmatic deciduitis. In the chorium and in the umbilical cord some fibroblasts presented nuclear aspects compatible with cytomegalic degeneration (by hematoxylin-eosin and in situ hybridization).

Both lungs exhibited morphological aspects that permitted the characterization of pulmonary dysplasia. The entire right and left lung showed resorption atelectasis and hyaline membranes lining the peripheral airways allied to venous and capillar congestion (Figure 3). A mild mononuclear focal

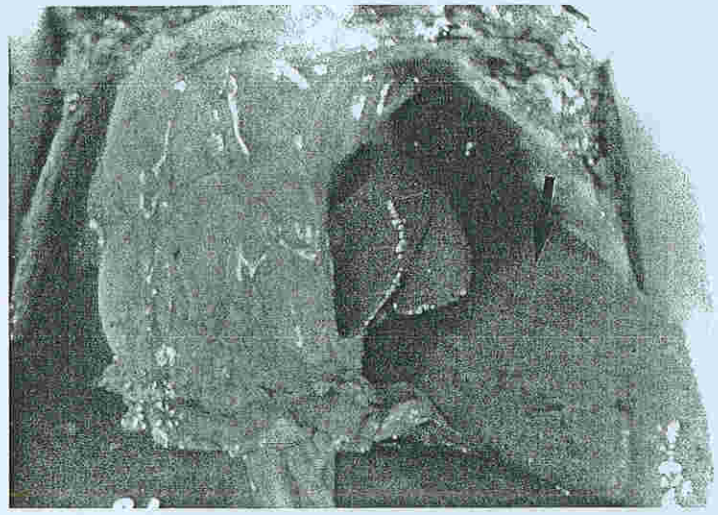

Figure 1. Thoracic cavity, showing shift of the mediastinum to the right. Compression of the left lung by a mass of extrapulmonary tissue (arrow) located in the costodiaphragmatic angle.

infiltrate was present in the alveolar septa and in the walls of the bronchi, bronchioli, and vessels. The acinar arterioles presented a thick medial wall. In the sequestered extralobar lung tissue no secondary atelectasis and no hyaline membranes were seen, but interstitial pneumonitis was observed. The parenchyma was similar to the canalicular phase of development and exhibited morphological aspects compatible with adenomatoid cystic malformation (CCAM type II) (Figure 4). A chronic leptomeningitis and ependimitis were evidenced, as well as the presence of hematoxynophilic mineralized plates randomly distributed or located around the vessels.

Degeneration of the internal elastic fibers of the abdominal aorta, of the vessel that irrigated the sequestration, and of the great pulmonary arteries was also present.

DISCUSSION

We report a case of EPS in a neonate who died in the first day of life of respiratory distress syndrome. The complete autopsy showed near the left lower lobe of the lung an EPS completely separated from the tracheobronchial tree and irrigated by an anomalous abdominal aortic vessel. In the areas of the normal lung perfused by the pulmonary artery, typical lesions of hyaline membrane disease were present, whereas the EPS perfused by systemic

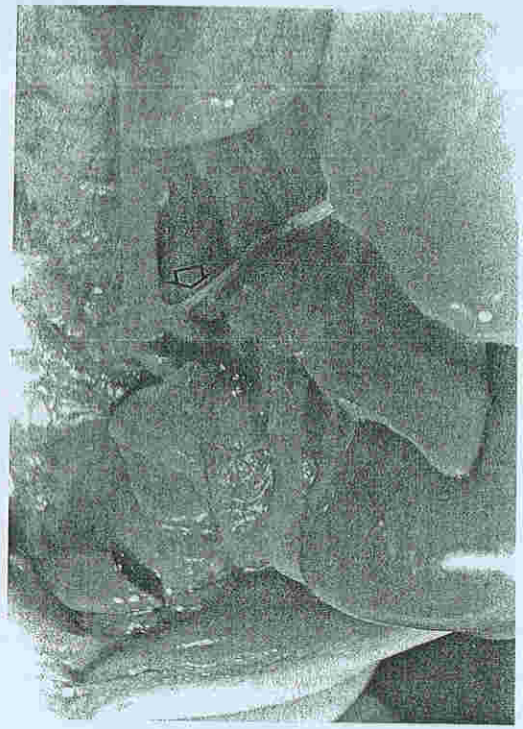

Figure 2. Thoracoabdominal cavity: an abnormal aortic vessel (arrow) supplies the EPS (upper right).

circulation displayed no HMD. To our knowledge, only a few similar cases have been reported [1–5] in which HMD was present in the normal lung tissue and absent in the sequestered lung tissue. The absence of morphologic aspects of HMD in the EPS was attributed to the importance of the circulation in the pathogenesis of the disease. The regions perfused by systemic circulation were better oxygenated, and it is suggested that the inequalities in oxygenation of the lung play a role in the localization of the disease. Lindgren [6], on the basis of stereomicroangiographic studies, emphasized the existence of a vascular component in the pathogenesis of HMD. In the sequestered lung tissue morphologic aspects compatible with CCAM were present, a finding not rarely observed [4, 7–9]. Hruban et al. [10], Gerle et al. [11], and Choplin and Siegel [12] stress that foregut malformations and associated congenital anomalies are generally believed to be restricted to EPS, but in the present case no other malformation was detected. Noteworthy was the presence of interstitial lung pneumonitis represented by a chronic inflamma-

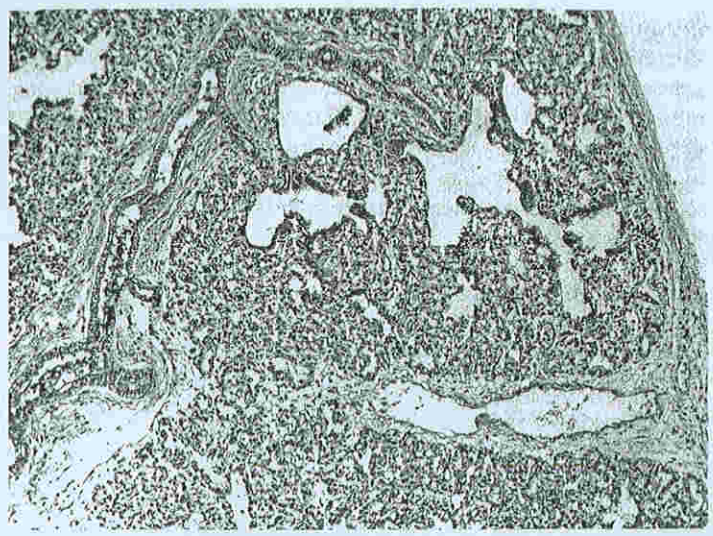

Figure 3. In a peripheral lobule hyaline membranes line terminal bronchioles and alveolar ducts. ×560.

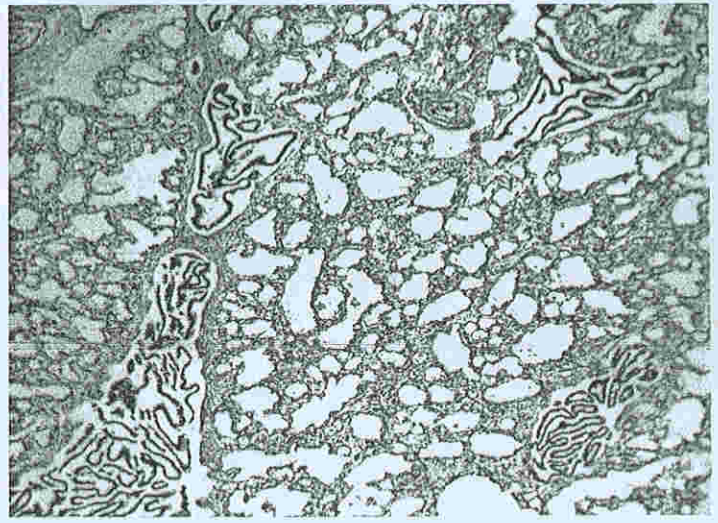

Figure 4. Extralobar sequestration with morphologic aspects of congenital cystic adenomatoid malformation, type 2. ×125.

tory interstitial process. As the placenta and several other fetal organs exhibited a chronic inflammatory process allied to vascular lesions (aorta, pulmonary arteries, aberrant aortic vessel) identical to those described in congenital viral infections [13, 14], we suggest a maternal infection, probably of viral etiology (cytomegalovirus?), as the cause of vascular and lung anomalies.

Our case also shows that a sequestered lung, not connected to the tracheobronchial tree, is protected against HMD present in all other parts of the pulmonary parenchyma.

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