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Congenital Adenomatoid Malformation of the Lung

(An Autopsy Case Report)

Jc G. Chi, M.D., Young Kook Ahn, M.D.*

Department of Pathology, College of Medicine, Seoul national University

Congenital adenomatoid malformation of the lung was acknowledged as a separate entity of cystic diseases of the lung and was introduced in the English literature by Ch'In and Tang in 1949. This condition is also known as congenital cystic adenomatoid malformation of the lung, and is a rare condition. According to Kwittken and Reiner (1962) a total of 32 cases were reported till 1962.

In all reported cases, congenital adenomatoid malformation(CAM) involves one lobe or one lung. And CAM occurred in premature infants or stillborns in many instances.

The purpose of this communication is to report a rare case of CAM which involved the entire lung in a new born infant who died of acute respiratory distress soon after birth.

Case Report(RCM# 45)

This female infant was delivered at full term to a 32 year old para 1-0-1-1 woman, after normal prenatal course, weighing 2700gm. at birth. Soon after the initial cry cyanosis and respiratory difficulty were noted. Intercostal retraction was marked. Clear frothy fluid came out from her nose and mouth, and the material was sucked out. However, respiratory distress was not improved. She was intubated because of persistent severe cyanosis. However, the lungs could not be expanded, and the patient died at 7 hours of age.

At autopsy (12 hours postmortem) the pleural cavities were free of fluid and adhesions. Entire lobes of the lungs were excessively voluminous weighing 70gms. The right lung was trilobed and the left lung was bilobed, and they maintained normal contour. The pleural surface of the lung was pale pink smooth and glistening. Both lungs had a consistency of liver. The lungs were dissected after formalin fixation. The trachea was in midline and patent. All lobar and segmental divisions of bronchial trees were widely patent. No cystic areas were noted throughout. Cut sections showed a diffusely consolidated pale grayish appearance with patent bronchial trees. No tumefaction was seen anywhere.

Microscopically, sections from entire lobes of the lungs showed essentially same findings that were characterized by diffuse adenomatous or microcystic structures. These microcysts were lined by prominent cylindrical or cuboidal epithelium displaying no papillary infoldings. These small uniform tubular structure could easily be distinguished from bronchioles. Cartilage was not present in any of these tubular structures. These structures did not show cystic dilatation although the spaces vary considerably in size. The transition from the columnar epithelium of the bronchiolar segments to the alveolar epithelium was not easily identified.

*Dr. Ahn's Obstetrics and Gynecologic Clinic, Shingil-Dong, Seoul 151, Korea. This study was supported in part by the China Medical Board research grant #74-732-11-5.

No evidence of hyaline membrane formation or inflammation was present. Interstitial connective tissue was separating these tubular or microcystic structures diffusely, and this interstitium was quite prominent in some areas of the lungs (Fig. 3). Dilated capillaries were seen intermixed with edematous and less cellular interstitium. Smooth muscle and elastic fibers were occasionally noted in interstitium. No evidence of inflammation was present. The bronchial and bronchiolar developments appeared normal. Cartilage rings in small bronchioles were well preserved.

The remainder of the organs including central nervous system showed no gross or microscopical abnormalities.

Discussion

Congenital adenomatoid malformation of the lung is sometimes interpreted as a hamartoma and is usually of a localized disease of the lung. In fact Halloran et al (1972) described that CAM is a surgically correctable cause of neonatal respiratory distress and death. However, if one reviews the cases described under the name of CAM one finds cystic and adenomatous elements vary considerably by different case. However, diffuse adenomatoid change of the entire lobes of the lungs was not described. Many reports (Craig et al, Goodyear et al, Spector et al, Thomas) state that the characteristics of CAM are (1) an adenomatoid increase of terminal respiratory structure (2) polypoid growth of the mucosa, lined by respiratory or cuboidal epithelium (3) absence of cartilage, apart from remnants of pre-existing cartilage and (4) absence of inflammation.

Above descriptions does not applicable to this case in two respects. First of all there was no cystic dilatations in any portion of the lung, and secondly the lesion was not a localized lesion but a diffuse anomaly throughout the entire lungs both right and left. Neverthless, the nature of lining epithelium, intimate relationship with surrounding connective tissue and absence of cartilage, etc are certainly in favor of adenomatoid malformation particulary as it occurred in a newborn infant. Our case is very similar microscopically to the cases reported by Dempster (1969) and Dijk and Wagenvoort (the third case), 1972, in which they used the term adenomatoid hamartoma or adenomatoid malformation. In these respects, our case might be called a "diffuse adenomatoid malformation".

We are reluctant to use the term hamartoma for the lesion we have in this case, because the pathologic process is a diffusely scattered involving entire lungs. The lobar architecture of the lung was well preserved and so was the tracheobronchial trees. There was no tumefaction anywhere and therefore no evidence of compression, which was so common in other cases of CAM, was present. In fact at fresh state of the lung we thought grossly that this probably represented massive aspiration pneumonia.

The pathogenesis of CAM is not known. The concept of hamartoma certainly could well be supported by many cases of localized type forming a mass. However, that concept cannot be adequately applicable for the widespread lesion like our case. As one reviews the slides of lung tissue of this case one gets an impression that adenomatoid structure does actually represents an overgrowth of distal bronchiolar tissue. Cuboidal to cylindrical epithelial lining, intimate association with mesenchymal elements among adenomatoid structures, absence of cartilage around these tubular structure and complete lack of atrial or alveolar differentiation are all inclined toward the view of the above contention.

Summary

An autopsy case of congenital adenomatoid malformation in a newborn female infant who was born after 38 weeks gestational period was reported.

This patient had marked cyanosis and respiratory distress immediately after birth, and was dead in 7 hours of age. Adenomatoid malformation was diffuse throughout the entire lungs, and there was no mass effect anywhere. Microscopically myriads of tubular structures lined by cuboidal or cylindrical epithelium were seen, embedded in fibrous stroma in which some smooth muscle and elastic fibers were present. No other gross or microscopic anomalies were observed.

≫國文抄錄≪

肺의 先天性 腺腫樣 畸形

(1剖檢症例 報告)

池 堤 根·安 榮 國

서울大學校 醫科大學 病理學校室

著者들은 38週後에 出生한 新生兒에서 出生直後부터 나타난 甚한 青色症과 呼吸困難으로 말미암아 生後 7時間만에 死亡한 女兒의 剖檢에서 稀貴한 肺畸形을 觀察하고 이를 記述하였다.

本 腺腫樣 畸形은 兩肺가 모두 같은 程度로 侵犯되었으며 肉眼的으로는 그 硬度가 肝臟과 같았고 氣管 및 氣管支등은 正常이었다. 이 患者의 畸形이나 異常은 肺에만 局限되어 있었다. 肺組織은 顯微鏡的으로 無數한 비슷한 크기의 管性 或은 腺性 構造로 되었으며 그 內腔은 圓柱狀 上皮細胞로 피복되어 있었다. 嚢性構造는 아무데서도 없었다. 間質에는 약간의 평활근섬유가 觀察되었다.

본 病變은 肺의 瀰蔓性 腺腫性 畸形이라 부름이 合理的이라 思料되었으며 아마도 最終 細氣管支 發育의 先天性 異常이라고 생각되었다.

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Legends for Figures

Fig. 1. Gross specimen of the lungs together with tracheobronchial trees. The picture was taken after formalin fixation of the tissue. Note uniform involvement of the entire lobes of the lungs as well as intact tracheobronchial trees.

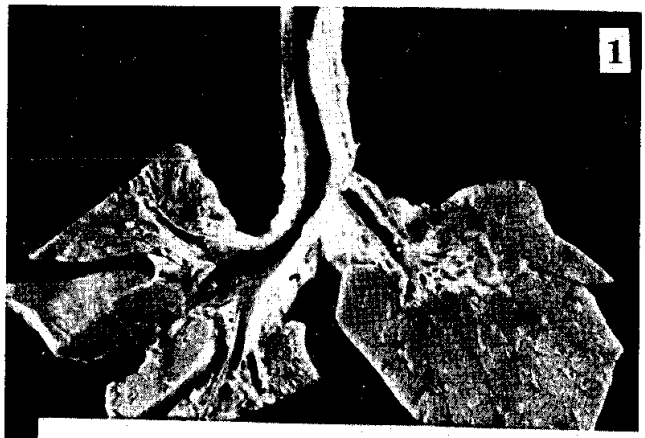
Fig. 2. Cut sections of the entire lobes of the lungs, showing remarkably same gross appearance characterized by a diffuse adenomatoid pattern with patent bronchial lumina.

Fig. 3. Photomicrograph of the lung. Note irregular tubular structures separated by various amount of stromal connective tissue. (H&E×20)

Fig. 4. Two bronchioles are seen in right corners and the remaining adenomatoid structure is seen.(H&E×40)

Fig. 5. Another area of the lung, showing adenomatoid malformation. (H&E×40)

Fig. 6. These adenomatoid structures are lined by columnar or cuboidal epithelium resting on vascular loose connective tissue stroma. (H&E×100).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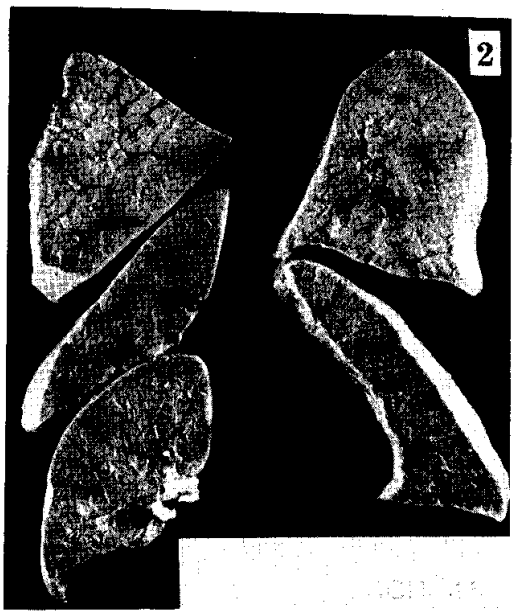
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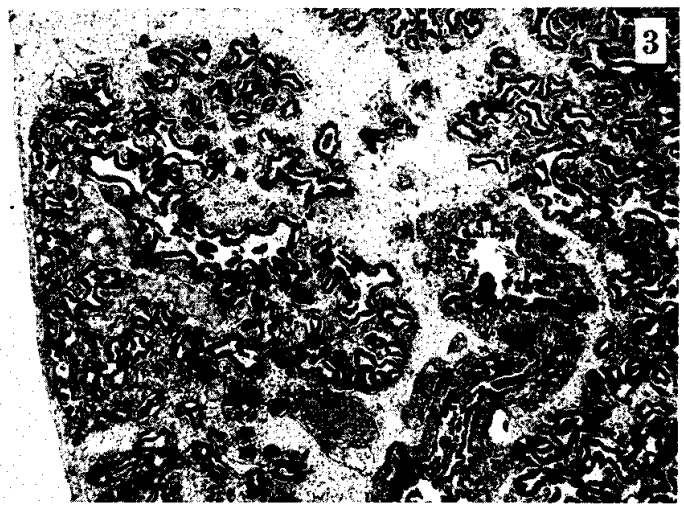
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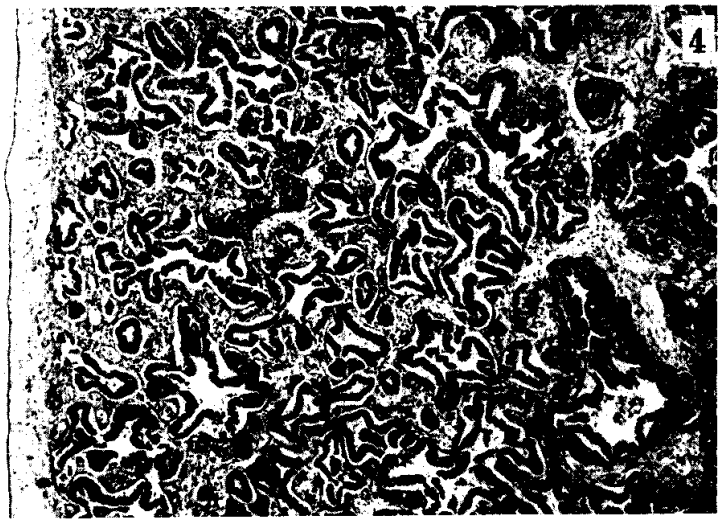
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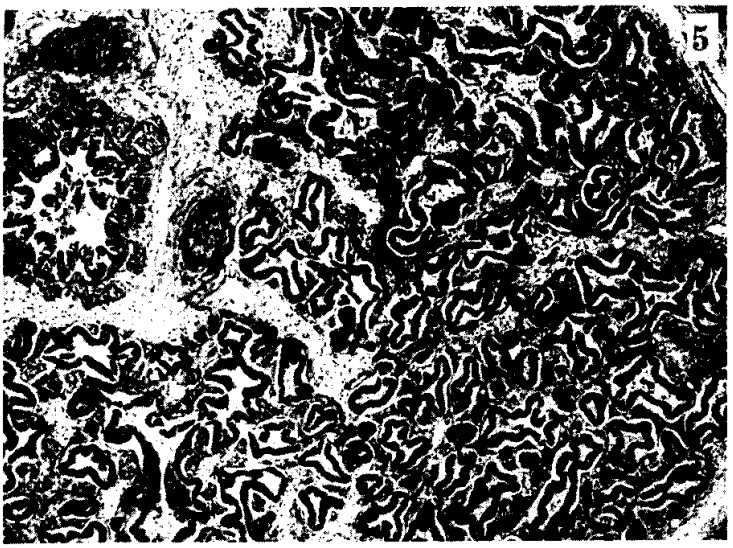
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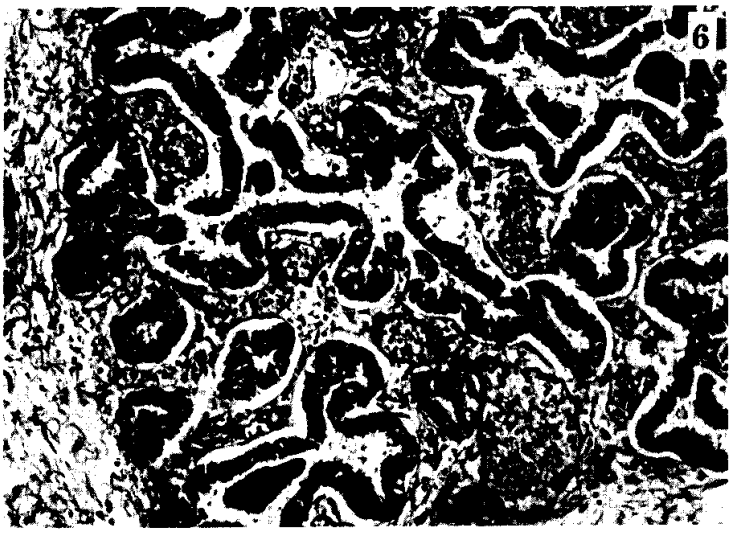
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