Case Report Section

Congenital Adenomatoid Malformation of the Lung

GORDON O. BAIN, M.D.
Edmonton, Alberta, Canada

In recent years the diagnosis of congenital cystic disease of the lung has been clarified and restricted as a result of the recognition of a variety of emphysematous and cystic lesions of diverse etiologies. True cystic disease is rare.\textsuperscript{1} Rarer still is the closely related condition first reported in 1897 by Stoerk as “cystic fetal bronchial adenoma,”\textsuperscript{2} and introduced in the English language medical literature in 1949 by Ch’In and Tang of Peking as “congenital adenomatoid malformation.”\textsuperscript{3} That the adenomatoid malformation is a variant of congenital cystic disease was acknowledged by Ch’In and Tang, but it is sufficiently distinctive to warrant recognition. Examination of published photographs indicates that the lesion in different cases varies from mainly solid, enlarged, tumor-like lobes to multicystic lesions. Moreover, as Craig, et al.,\textsuperscript{4} point out, there are cases of true congenital cystic disease in which areas resembling the adenomatoid malformation are found upon pathological examination. It would appear possible to assemble a series of transitional cases linking true congenital cystic disease and the adenomatoid malformation. The following case is typical of the adenomatoid malformation and represents, we believe, the 28th reported case. Previously reported cases have been tabulated by Gottschalk and Abramson.\textsuperscript{5}

\textit{Case Report}

\textit{Case:} The mother, aged 23 years, para I, gravida II, first visited her doctor in the sixth month of pregnancy. Examination revealed no abnormal findings. Her first child, aged 13 months, was reported to be alive and well. The mother returned to the office six weeks later with an extremely distended abdomen, the uterus being tense and tender. The blood pressure was normal. The fetal heart could not be heard. A

\textit{FIGURE 1:} Cut surface of adenomatoid malformation. Fixed tissue.
FIGURE 2: Normally developed, unexpanded left lung above, and fetal right lower lobe below.

FIGURE 3: Peripheral portion of adenomatoid lobe with alveolar differentiation at top right and adenomatoid tissue at bottom left. Insets show under higher magnification alveolar development at top left and adenomatoid tissue at bottom right.
roentgenogram revealed a single vertex presentation. She was admitted to the hospital and the membranes were ruptured. Over 8 liters of amniotic fluid was released, and a stillborn girl was soon delivered. The placenta was normal.

Autopsy: (Relevant findings only). External examination revealed a 1,530 gram premature infant with crown-rump measurement of 25 cm. The chest appeared somewhat increased in anteroposterior diameter. The 10 gram left lung was grey-purple and atelectatic. Mediastinal structures were displaced toward the left by the massive, tumor-like, 100 gram right lung. Closer inspection revealed that the large, smooth, rounded, pink-grey mass represented the upper lobe of the right lung, the lower lobe consisting of a small, inconspicuous, grey-purple structure. No middle lobe was identified. The cut surface of the massive right upper lobe was solid, homogeneous, grey and fleshy with a few groups of irregular and deformed bronchial channels (Figure 1).

Histological Examination: The liver was the site of pronounced hemopoietic activity, in keeping with prematurity. The left lung was normal in structure and unexpanded (primary atelectasis). The small right lower lobe consisted of lobulated, fetal lung, but aside from retardation in development, was not malformed (Figure 2). The massive right upper lobe presented a striking adenomatous appearance characterized by great proliferation of bronchiolar structures without alveolar differentiation except in a narrow subpleural zone (Figure 3). In none of the many sections examined did bronchiolar structures have cartilage or tubular mucous glands in their walls. Many of them, however, were supported by narrow bands of smooth muscle. Some of the larger bronchi were irregular and tortuous with papillary infolding of their walls. An interesting finding was small groups of bronchioles lined by tall columnar epithelium with clear cytoplasm giving a strongly positive PAS reaction for epithelial mucin (Figure 4). Finally, it was noted that capillary blood vessels were inconspicuous in the supporting mesenchyme of the non-alveolated tissue, but comparatively abundant in the narrow subpleural zone in which alveoli were present.

DISCUSSION

The adenomatoid lobe bears little resemblance grossly to the usual form of congenital cystic disease. Five main features characterize the adenomatoid malformation and

![FIGURE 4: Structure of central portion of adenomatoid lobe is shown at the left. At top right are dilated subpleural malformed bronchi and a focus of columnar mucinous epithelium. Lower right shows mucinous epithelium under high magnification.](http://journal.publications.chestnet.org/pdfaccess.ashx?url=/data/journals/chest/21330/)
Vol. xxxvi  
CONGENITAL ADENOMATOID MALFORMATION  
433

differentiate it from cystic disease: (1) absence of bronchial cartilage (2) absence of bronchial tubular glands (3) presence of foci of tall columnar mucinous epithelium (4) marked overproduction of terminal bronchiolar structures without alveolar differentiation except subpleurally (5) massive enlargement of the affected lobe. Areas of adenomatoid structure may be present in true cystic disease, but in the latter the cystic bronchi usually have cartilage or tubular glands in their walls.

Superficially, the adenomatoid malformation resembles fetal lung, except that it is not lobulated. The presence of alveoli at the periphery of the lobe is a puzzling, yet characteristic, finding. Craig et al.1 by increasing the intrabronchial pressure at operation, were able to demonstrate the inflatability of the malformed lobe in one case. The published photomicrographs from their four viable cases treated surgically show much more alveolar structure than did the case described in this report. In the present case alveoli were absent in some parts of the periphery of the malformed lobe; here the bronchial structures were dilated (Figure 4).

Characteristic of the adenomatoid malformation is the massive size of the affected lobe, which displaces other thoracic structures. This evidence of excessive growth led to the suggestion that the lesion may be a form of hamartoma, although it has little in common with the cartilaginous lesion usually designated by that term. In true cystic disease the affected lobe may occasionally be enlarged, but here it is a result of hyperdistention due to a check-valve mechanism.7

The reported cases have been mostly premature stillborn infants, but a few have been born alive at term. In a few instances the abnormal lobe has been resected surgically, with survival and cure. The untoward effects of the disorder result from intrathoracic pressure effects with circulatory and respiratory embarrassment. Anasarca, a feature of 13 of the 27 reported cases, is attributable to such pressure effects. Polyhydramnios, a feature of four previous cases, was in each instance associated with anasarca. Polyhydramnios was present in our case, but not anasarca. Obstruction of venous return from the placenta has been considered the cause of the polyhydramnios.8


REFERENCES