Mucocele of the Lung Due to Congenital Obstruction of a Segmental Bronchus: A Case Report; Relationship to Congenital Cystic Disease of the Lung and to Congenital Bronchiectasis*

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The case of mucocele of the lung herein reported is of interest primarily because of its probable relationship to bronchogenic cyst on the one hand, and congenital bronchiectasis on the other. The author suspects that it is, in fact, the same condition as the former, but in an early phase, and that either is antecedent to true congenital bronchiectasis.

Case Report

Miss M.S., Orange County General Hospital. This 15 year old Mexican female was admitted to the hospital March 21, 1949, with symptoms of weakness, easy fatigue, and feeling feverish. There was weight loss of 10 pounds over the previous two months and history of close contact with active tuberculosis in the preceding two years. Physical examination was not remarkable; there were slightly increased breath sounds at the right apex. X-ray films of the chest showed bilateral infiltrations of moderate degree and a circumscribed lesion in the left lung. She reacted to tuberculin. The sputum was negative, but a gastric culture was positive, establishing definitely the diagnosis of pulmonary tuberculosis.

On a regimen of bed rest in the hospital, the pulmonary infiltrations decreased markedly, with the exception of the rounded density in the region of the apex of the lower lobe on the left side. X-ray films taken at several distinct intervals show this to persist (Figures 1 and 2). It was featured by an air-fluid level.

Because of this, the patient was seen in consultation, and it was considered that the rounded lesion should be removed surgically, the tentative diagnosis being either a congenital bronchial cyst with associated tuberculosis, or pulmonary tuberculosis with cavitation in the left lower lobe. On July 22, 1950 findings at operation were as follows:

There were filmy adhesions between the superior and posterior segments and the adjacent chest wall. The arrangement of the lung was abnormal in the following respects: the basal segments were divided into two equal portions by a long vertical fissure; there was a partial fissure between the superior segment of the lower lobe and the basal segments; the interlobar fissure was incomplete, being totally absent in the posterior one-half; the bronchi of the posterior segment of the upper lobe originated from a subsegmental bronchus (B6b) of the superior segment of the lower lobe, as illustrated, and thus traversed the posterior portion of the usual fissure line.

Dissection revealed a smooth, rounded, pale, semitranslucent cyst-like dilatation of the bronchial system, extending through the superior and posterior segments. When an occasional branch was ruptured during the dissection, it was found to contain a large amount of thick, whitish, mucoid material (Figure 3). There was absolutely no evidence of inflammation or of solid deposit.

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After division of the superior segmental bronchus of the lower lobe just distal to the origin, it was possible to visualize the proximal stump lumen. This was imperforate, resembling a shallow, blind, funnel-shaped pocket, the walls of which were completely smooth as were the walls of all the cystic spaces mentioned above. The entire lung had relatively little carbon deposit, but the superior segment was totally devoid of such pigmentation. All segments of the lung seemed to inflate normally, although the posterior segment deflated somewhat more slowly than the other. The blood supply, artery and veins, and the remaining hilar and pulmonary anatomy were normal. The diaphragm, pericardium, and mediastinal structures also were normal.

There was a 2 cm. focus of induration palpable beneath the surface of the superior lingula segment. On removal this was found to be somewhat similar to the above abnormalities, but the bronchi to this subsegment were not dilated and contained no mucoid material. No obstructing septum was present. The bronchial walls here were thickened, more fibrotic and somewhat nodular, and in one place there was a white amorphous deposit, approximately 1 mm. in diameter. Pathologically this proved to be tuberculosis.

The operation performed was a triple pulmonary segmental resection of the left lung, removing the superior segment of the lower lobe and the posterior segment of the upper lobe together, and a subsegment of the superior lingular segment of the upper lobe separately, because of the tuberculous lesion at that point. Her convalescence was uncomplicated. She is still in the Sanatorium with active tuberculosis.

On November 18, 1950 she was bronchoscoped in an attempt to visualize the superior segmental orifice of the left lower lobe. Such a superior segmental orifice was not visible. The remaining tracheo-bronchial tree was within normal limits. The features of the case are:

**FIGURE 1**

*Figure 1*: Radiograph dated March 22, 1949. In the third left anterior interspace, there is a round lesion containing fluid in the lower one-half and air in the upper half. Note that the wall is extremely thin. There is also associated bilateral pulmonary infiltration. — *Figure 2*: Radiograph taken July 1, 1950. This shows the same round lesion in the third left anterior interspace. The wall is still extremely thin. The relative amounts of air and fluid have changed slightly. The size of the lesion has not changed. The bilateral infiltrations have decreased considerably.
1) The diaphragm-like obstruction at the commencement of the superior segmental bronchial orifice.

2) The distention, with glairy mucus, of the entire bronchial system tributary to this (all three branches of the superior segment of the lower lobe and also the posterior segment of the upper lobe) (Figure 4).

3) The distention with air of the alveoli of the tributary segments, in spite of the filling with mucus of the bronchial air ducts and the complete obstruction of the inlet orifice.

4) The normal size, and inflation-deflation of the alveoli of the tributary segments.

FIGURE 3: Photograph of gross specimen, showing the bronchus of the posterior segment of the upper lobe leading into the bronchus of the superior segment of the lower lobe. The insert explains the photograph: (1) the posterior segment of the upper lobe; (2) the bronchus of the posterior segment of the upper lobe; (3) part of the superior segment of the lower lobe; (4) the main stem bronchus of the superior segment of the lower lobe at the site of amputation.
5) The lack of densities conforming to the pathological findings on x-ray examination, one air-fluid cyst alone being demonstrated.

6) The air component of the air-fluid cyst in spite of a totally blocked bronchial system.

7) The lack of pigmentation in the involved segments.

8) The aberrant drainage of the segmental bronchus from the posterior segment of the left upper lobe into the subsegment bronchus (B6b) of the superior segment of the lower lobe.

9) The use of segmental resection allowed removal of the pathological areas with loss of only two segments, whereas lobectomy surgery would have meant the sacrifice of the entire lung.

Discussion

Sante,2 following Mueller's speculation, has well described the probable method of formation of congenital bronchial atresia, partial or complete. The sequence of events is considered to be that a bronchial bud fails during its growth to have normal tubular development. If the failure is complete, the bronchiole becomes represented only by a cord. If growth is resumed after a time, tubularity is established distal to an area of artesia. If the bronchiole so obstructed develops secretory glands, the fluid so formed will accumulate with the production of a fluid cyst.

FIGURE 4: Diagram representing the essential pathology. Only the bronchial systems of the two affected segments are illustrated. The bronchi of the posterior segment of the upper lobe and the bronchi of the superior segment of the lower lobe have a common obstruction, the septum at the commencement of the superior segmental orifice of the lower lobe. All the bronchi tributary to the obstructing septum are dilated and filled with mucus. One small branch, however, contains air in addition to mucus. The parenchyma of both the affected segments is in a normal state of inflation, despite the bronchial obstruction.
The thin, smooth diaphragm at the orifice of the superior segmental bronchus was not removed for obvious reasons, but from bronchoscopic study and the appearance at surgery when it was exposed from the pulmonary side, there was no evidence suggesting an inflammatory or neoplastic origin. Thoracic surgeons know well that single, fluid-filled intrapulmonary bronchogenic cysts nearly always have such smooth, thin septa at the point of origin from a bronchus. Almost certainly these septa are congenital in origin.

Had the specimen herein described been examined by multiple section technique, the true situation might have been lost in a picture of multiple cysts of varying size. The pathological diagnosis would then have been multiple bronchogenic cysts, implying both a separation of the elements and also multiple bronchial obstructions. It is suggested that perhaps most, if not all, so-called multiple, intrapulmonary bronchial cysts are examples of bronchial mucoceles, and that if such diseased portions of lungs were examined by tracing the bronchial systems in a retrograde manner, the true situation would be recognized.

Every other bronchial cyst of the lung removed by the author has shown several small outpouchings in the large cyst wall. These are almost certainly dilated bronchial branches partly taken up in, and partly compressed by, the major cyst mass. It is probable that such an intrapulmonary bronchial cyst is no more than an advanced stage of a "mucocele of the lung."

True congenital bronchiectasis probably exists as another variant of a mucocele, originating by rupture of the obstructing septum after dilatation of the tributary bronchial system has taken place.

A study of the literature fails to reveal an instance where such obstruction of the bronchial system, and coexistent distention with secreted mucus, existed along with inflation-deflation phenomenon of the dependent alveoli, as was discovered in the case under discussion. To produce this combination of circumstance, either multiple obstructions were present at each alveolar duct entrance to prevent entry of mucus into the alveoli, or else (and more likely) the mucus was too viscous to pass through the minute openings. The air passage must have been transalveolar and transsegmental. The experiments of Loosli are of pertinent interest in this regard. He produced bronchial obstruction in dogs by painting the bronchus with 35 per cent silver nitrate. As a result of this, a non-inflammatory atelectasis developed. The following observations were made: "In thick sections which afford a surface view of the alveolar walls, numerous openings or pores are seen in the intercapillary spaces. In thin sections, these appear as partial or complete interruptions in the alveolar septa." — "The small and terminal bronchioles, which do not possess cartilage in their walls, do not contain mucus and are collapsed. The cuboidal cell membrane is intact and thrown into folds. Some contain collections of macrophages, but usually they are empty." — "The alveolar ducts and alveoli appear as slits and crevices in what might be called 'solid tissue.' " These experiments in dogs produced a situation very similar to that of the lesion described. There was bronchial obstruction with the larger bronchi distended with mucus,
whereas the terminal bronchioles and alveoli were collapsed and free from mucus. It is probable that if the experiment were repeated in such a manner as to produce obstruction with the silver nitrate in a segmental bronchus, the larger bronchi would become filled with mucus as described above, but the alveolar portions and the terminal bronchioles would distend with air by way of collateral ventilation from the adjacent segments, thus representing an exact analogy to the case reported herein.

Thoracic surgeons are well aware that when segmental bronchi are clamped shut at surgery, positive pressure intratracheally, by the anesthetist, almost routinely produces inflation of the alveolar system of the occluded segment. This does not usually occur if there exists significant parenchymatous disease of that segment.

Baarsma, Dirken and Huizinga have carried out experiments in man which well demonstrate the transsegmental, transalveolar respiration which exists in the absence of parenchymatous disease. They obstructed either the entire lower lobe bronchus, or the lower lobe bronchus just distal to one or more of the lobar segmental bronchi, resulting in obstruction of only some of the segmental bronchi of the lower lobe. They found that when the bronchus was completely obstructed, atelectasis occurred and there was no evidence of collateral ventilation; whereas, if only a side branch were obstructed leaving at least one of the lobar segmental bronchi unobstructed, then collateral ventilation did occur and atelectasis did not supervene.

The failure of the distended mucus-filled tributary bronchi to give significant x-ray shadows is of great interest. Presumably the failure is due to an unfavorable density contrast by the scattering of the pathology throughout distended lung of considerable depth. It indicates that an

<table>
<thead>
<tr>
<th>TABLE I: PULMONARY CYSTS</th>
</tr>
</thead>
<tbody>
<tr>
<td>1. MECHANICAL</td>
</tr>
<tr>
<td>BRONCHOGENIC</td>
</tr>
<tr>
<td>— Variants</td>
</tr>
<tr>
<td>— Mucocele</td>
</tr>
<tr>
<td>— Cyst</td>
</tr>
<tr>
<td>— Congenital Bronchiectasis</td>
</tr>
<tr>
<td>PARENCHYMAL</td>
</tr>
<tr>
<td>— Variants</td>
</tr>
<tr>
<td>— Blebs</td>
</tr>
<tr>
<td>— Bullae</td>
</tr>
<tr>
<td>— Pneumatoceles</td>
</tr>
<tr>
<td>2. INFLAMMATORY</td>
</tr>
<tr>
<td>Bacterial</td>
</tr>
<tr>
<td>Fungal</td>
</tr>
<tr>
<td>Parasitic</td>
</tr>
<tr>
<td>3. NEOPLASTIC</td>
</tr>
<tr>
<td>Benign</td>
</tr>
<tr>
<td>Malignant</td>
</tr>
<tr>
<td>4. ABERRANT</td>
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<tr>
<td>Gastro-enterogenous, etc.</td>
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</table>
individual may have an apparently normal chest roentgenogram followed 
subsequently by the appearance of a fluid-filled or air-filled cyst (as the 
process continues or results in rupture) and yet have a truly congenital 
disease. In other words, the late appearance of a cyst by x-ray does not 
necessarily mean the cyst is an acquired one.

Presumably the lack of pigmentation typical of congenital bronchogenic 
cysts is due to the small size of the intersegmental and transalveolar 
communications and not to the failure of air to enter the affected tissue. The 
tiny communications probably act as a selective filter, removing carbon 
particles. It is a common finding that the intersegmental planes bordering 
such an obstructed segment have an extraordinary deposit of pigment.

The aberrant drainage of a segmental bronchus from one lobe into a 
tributary of a segment of another lobe is of interest.

The above simple classification is offered because of its simplicity and 
its correlation with the clinical aspects of diagnosis. All subdivisions are 
self-explanatory, excepting the mechanical group. It is considered that 
there are two major varieties of mechanical cyst; (a) bronchogenic, and 
(b) parenchymal. Bronchogenic cysts arise by obstruction to a portion of 
the bronchial tree, usually because of a congenital septum or portion of 
atelesia, but possibly also, on occasion, from an acquired obstruction. Three 
variants of the same fundamental lesion are: (1) mucocele, (2) cyst, and 
(3) congenital bronchiectasis, as discussed previously. Parenchymal cysts, 
likewise, are thought to arise from a mechanical interference with collateral 
ventilation. Such interference could be temporary or permanent, and prob-
ably results from pulmonary edema, inflammation, or fibrosis. The variants 
of the same pathology include blebs, bullae, and pneumatoceles, which are 
probably nothing more than different degrees of the same process.

A similar interference with collateral ventilation, but in such a manner 
as to produce the reverse effect; namely, the loss of air-filling rather than 
an increase in the amount of air trapped, might explain the areas of plate-
like atelectasis frequently found during minor inflammation and/or edema 
of lower lobes.

SUMMARY

A case of mucocele of the lung has been described. It appears to provide 
the “missing link” between bronchial cysts and congenital bronchiectasis. 
It suggests that pulmonary blebs, bullae, pneumatoceles, and perhaps em-
physema arise from disturbances in the collateral ventilation of the lungs.

RESUMEN

Se describe un caso de mucocele del pulmón. Parece que proporciona el 
“eslabón faltante” entre los quistes bronquiales y la bronquiectasia congé-
nita. Esto sugiere que las ampollas, bullas, pneumatoceles y tal vez el enfisema 
emanan de trastornos de la ventilación colateral de los pulmones.

RESUME

L’auteur décrit un cas de mycocèle du poumon. Cette affection semble 
tenir le juste milieu entre les kystes bronchiques et la bronchectasie con-
génitale. Elle amène à penser que les kystes aériques, les pneumatocèles et peut-être l'émphyème pourraient venir de perturbations de la ventilation collatérale des poumons.

REFERENCES