The Syndrome of Bronchial Mucocele and Regional Hyperinflation of the Lung
Report of Four Cases*

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Studies of four cases of bronchial mucocele with regional hyperinflation are reported. Three are female patients. The first indication of a pathologic condition in three patients is assumed to have been in early childhood, although an exact diagnosis was determined much later by operation. Regional emphysema was noted in all of them, extending nearly to the entire lobe. The mechanism of this emphysema is discussed briefly, and a congenital defect of the bronchi of involved lobes is postulated to be the cause of the emphysema and formation of the bronchocele.

When the lumen of the bronchus becomes occluded, either congenitally or later, the dilatation of a distal portion may develop, with gradual mucous accumulation. This dilated bronchial structure is referred to as "the bronchial mucocele [bronchocele]." It is of interest that if the segmental bronchus is occluded there is no atelectasis, but a striking degree of focal hyperinflation occurs in the involved segment. This complex of bronchial atresia and focal emphysema was first reviewed in 1966 by Curry and Curry,1 who added two cases to the six they found previously reported. Talner et al10 in 1970 added 11 other cases along with their own two examples. They summarized clinical, roentgenologic and pathologic findings in these 21 patients, although there was no description of focal emphysema in three cases. They referred to this pathologic condition as "the syndrome of bronchial mucocele and regional hyperinflation of the lung."

We have observed four cases of this entity. In three of them the onset can be traced back to early childhood and in another case an early onset is strongly suspected. Thus, these four cases should be postulated as congenital.

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Case Reports

Case 1

The patient is a nine-year-old boy. At ten months of age the boy was examined because his grandfather had tuberculosis. A chest roentgenogram (Fig 1) showed a consolidated shadow in the left hilus, with a high radiolucency in the left upper lung field. Since then, he has occasionally had fever, with production of sputum. At two years of age, a consolidated shadow measuring 2 x 2 cm and containing fluid appeard in the left hilum.

Figure 1. Case 1, ten months after birth. Consolidated shadow at left hilum.
peared within the lung. At three years of age the patient was admitted to the Kyoto Municipal Hospital because of a "fever attack." X-ray film findings on admission showed a pneumonialike infiltration, in addition to a cystic shadow containing fluid. The erythrocyte sedimentation rate was 32 cu/mm per hour, with a leukocyte count of 10,300 cu/mm. His temperature was 37.5°C. The temperature fell rapidly after administration of antibiotics, and the pneumonialike shadow decreased profoundly. He left the hospital three weeks later. Since that time there were no clinical symptoms until the patient was eight years old, and it was recommended that an operation be performed. On admission, there were no abnormal physical signs. The urine and blood were normal. A tuberculin reaction was negative. Ventilatory and blood gas values were within the normal range. The tubercle bacilli in sputum was negative. A roentgenogram (Fig 2) showed a 4.0 x 3.0 cm cuboidal shadow containing fluid, with apparent hyperradiolucency in the left upper and middle lung field. The border of the left lung expanded to the right edge of the spine. Bronchography (Fig 3) showed that the bronchial trees of the left upper and lower lobes were pressed down to the lower half of the left lung field, and B1 + 2 was not demonstrated. Pulmonary angiography (Fig 4) showed that the origins of left A3 and A1 + 2 were in normal localization but their distal branches were very fine showing immature arteries.

Left thoracotomy was performed. It was noted that the whole left upper lobe was inflated with air S1 + 2 and S3 including the cyst were resected. After resection, the cyst was incised, revealing yellowish, green mucous fluid. No open connection between the cyst and the proximal bronchus was noted. Three slightly ectatic bronchi could be observed in a distal direction from the lumen of the cyst. Histologic examination revealed the inner surface of the cyst to be almost entirely covered with ciliated epithelium, with small areas of connective tissue having the appearance of lesions. In the pulmonary parenchyma surrounding the cyst, there were dense areas of bronchioli within which mucous plugs containing cell debris were noted. Alveoli were leaking in air space, and thickened interstitial tissues with lymphocytes and plasmocytes were seen. These findings may show that there was repeated inflammation due to the accumulation of mucous secretion surrounding the cyst. On the other hand, in the distant peripheral region from the cyst, pulmonary emphysema was noted almost everywhere, with the exception of occasional small atelectatic areas with pro-

FIGURE 2. Case 1, on admission (patient nine years old). Thin-walled cystic shadow (4 x 3 cm) with fluid level, hyperradiolucency in left upper and middle lung field. Expansion of left lung reaches right edge of spine.

FIGURE 3. Case 1, (bronchography). Left upper and lower lobe bronchi are pressed down to lower half of left lung. B1 + 2 is not demonstrated.

FIGURE 4. Case 1 (angiography). Origins of left A3 and A1 + 2 are in normal positions. Their peripheral branches are very fine.
and ventilatory capacity findings were normal. The tuberculin reaction was positive. Her chest x-ray film (Fig 6) findings were almost the same as in the past several years, showing a hyperradiolucency in the right upper lung field, with a thin-walled cystic shadow of 2 cm in diameter and containing fluid. There was a large (8 x 6 cm) consolidated shadow with an equivocal fluid level superimposed on the right mediastinal shadow. During bronchography it was noted that the right upper lobe bronchi were pressed down to the position below the second intercostal space.

Right thoracotomy was performed. The right upper lobe was overinflated with air, and did not deflate on thoracotomy. A large cyst located within the mediastinum and a small portion of the emphysematous lobe were resected. Unfortunately, the surgeon could not find any abnormal structure by palpation of the smaller cyst in the upper lobe, and it was left unresected. Histologically, the wall of the cyst was covered with one layer of epithelium cells and small cartilages, with mucous glands and muscle bundles noted beneath the mucous membrane. These findings showed that the cyst was of bronchogenic origin. Histology of the emphysematous portion was almost the same as described in Case 1.

This patient had a bronchogenic cyst and a mucoid bronchocele surrounded by lobar emphysema.

**Case 3**

The patient is a 21-year-old girl who was told of an abnormality in the chest x-ray film when she was six years of age. A tuberculin reaction was positive at that time, but she received no treatment. For about 20 days she complained of general malaise. Roentgenographic findings revealed a shadow in the upper chest area.

On admission, no abnormality was noted in her blood, serum, or urine. The ECG and ventilatory capacity findings were normal. A chest roentgenogram showed a thin-walled cystic shadow measuring 3 x 2 cm, with fluid in the left upper hilar field, and a slight hyperradiolucency with decreased vessel shadows in the left upper lung field. Bronchography revealed that the left upper lobe bronchi were pressed down below the second intercostal space, showing hyperinfiltration of the left upper lobe.

A left thoracotomy was performed. S1 + 2 and S3 were resected. Histologic findings of the resected cyst and the pulmonary tissue were almost identical to those in cases 1 and 2.

**Case 4**

The patient is a 30-year-old housewife who suffered from nephritis at eight years of age and pneumonia at 15 years of age. Two weeks before admission she had had a baby girl. Because of atomic bleeding and subsequent fever, she was examined roentgenologically. A shadow was noted in the left upper lung field. She had never been examined roentgenologically until then. She was advised to come to our hospital.

On admission, she was well nourished. No abnormal findings were noted in her blood and urine examination. Results of various serum tests were normal. The ECG and ventilatory capacity findings were normal. A chest roentgenogram showed apparent hyperradiolucency with decreased vessel shadows in the left upper lung field and a cystic shadow measuring 1.3 x 1.5 cm and containing fluid within the hyperlucent lung field. Bronchography showed apparent dislocation of the upper lobe bronchi to the position below the third intercostal space. Left thoracotomy was performed, and S1 + 2 was resected. Histologic findings in the resected cyst and the pulmonary tissue were almost the same as those described above.
SYNDROME OF BRONCHIAL MUCOCELE

DISCUSSION

The bronchocele was first reported by Ramsay et al in 1953. Ramsay thought that the pathogenesis was the same as the induction and development of bronchogenic cysts and congenital bronchiectasis, differing only in their time of onset. It may be hard to believe that the bronchogenic cyst, which is of ectopic origin, is the same entity as the bronchocele, which is originally orthotopic. But, the finding in Case 2, in which a bronchogenic cyst coexisted with a bronchocele with emphysema (unfortunately, this bronchocele was not resected) might support Ramsay's theory.

In three out of four of our cases, the first indication can be traced back to early childhood (ten months, six years and six years of age) either anamnestically or roentgenologically. The remaining patient had never been examined roentgenologically; therefore, the onset is undetermined. The early onset of mucoceles suggests that they are usually congenital.

The most frequent site of a bronchocele seems to be the left upper lobe. In three of our four cases, bronchocele and regional emphysema were localized in the left upper lobe. The same observation was made by Talner et al whose studies showed that in 14 of 21 patients the cyst was located in the left upper lobe.

The most interesting phenomenon is the regional emphysema. All four of our cases showed this change. Among 21 cases of Talner et al. 16 showed emphysema. Bronchocele undoubtedly occurs as the result of bronchial atresia. The cause of this atresia is unknown. In spite of bronchial atresia, the distal portion of the involved segment is frequently hyperinflated. It should be postulated that intersegmental air drift remains. This passage might be the pores of Kohn. The relationship between atresia of the segmental bronchus and emphysema of the same segment was investigated clinically and experimentally by Culiner et al. Martin and Reich et al. It was also discussed thoroughly by Talner et al.

On the contrary, if the atresia occurs in the lobar bronchus, atelectasis is the common result.

As shown in our cases, regional emphysema frequently extends over the involved segment to the neighboring segments or to the whole lobe. This extension of emphysema beyond the involved segment cannot be explained solely by the interalveolar air passage. There might be some check value mechanisms in other bronchi than the atretic bronchus, produced by the pressure of the inflated segment. In this condition congenital immaturity of other segmental bronchi, as shown in our cases, may play a role in the air-trapping phenomenon. Similar regional emphysema was observed in the bronchogenic cyst, in which incomplete obstruction of bronchi due to the pressure of the cyst was postulated to be the cause of air-trapping.

Thus, from clinical and pathologic experience with our cases, we believe that the formation of the bronchocele and regional emphysema might be based on a congenital developmental defect of bronchi in the involved lobes.

REFERENCES