A REPORT ON AN EXAMPLE OF CONGENITAL BRONCHIECTASIS WITH THE RESULTS OF POST-MORTEM AND PATHOLOGICAL INVESTIGATION.

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GEORGE L., aged 18, was admitted to the Victoria Park Chest Hospital at the request of Dr. Fell, of Colchester, on February 27, 1934, complaining of cough accompanied by profuse and foul sputum.

The patient's mother said that he had been "a bit wheezy on the chest" since infancy, this symptom having been first noticed when he was fourteen months old. There was no history of pleurisy, pneumonia or pulmonary inflammation, and he had enjoyed good health till the onset of his present illness and had had no difficulty in following his occupation as a milk roundsman. In January, 1934, he complained of pain in the right side of his chest. At the same time the breath became offensive and he began to cough up large amounts of purulent sputum. The onset of these symptoms was evidently acute.

The patient was a pale-faced, poorly nourished and under-developed lad, looking two or three years younger than his age. The breath was fœtid. There was advanced clubbing of fingers and toes. On physical examination of the chest the whole of the right side was dull to percussion and loud bronchial breathing was heard all over. Despite the bronchial secretion, which the expectoration proved to be profuse, very few adventitious sounds could be heard. The heart was displaced to the right.

At the time of admission to hospital, the patient said he was expectorating about 4 oz. of sputum daily. This was confirmed during the first twelve hours when 2 oz. were brought up, but on the second day the amount rose to 7 oz. and on the third day to 25 oz. During the next few days the amount varied between 30 and 60 oz. X-ray examination in the anterior-posterior position showed in the upper third of the right lung field numerous annular shadows, the rings overlapping and evidently representing a cystic or saccular condition of the upper lobe, no evidence of normal pulmonary tissue being seen. The lower two-thirds of the lung fields on this side was clouded, apparently as the result of pleural thickening, but annular shadows obscurely visible indicated that the condition present in the upper lobe extended throughout the lung. On the left side no abnormal shadows were seen, but the wide intercostal spaces and elevated ribs of a compensatory emphysema were noted, and this condition was later found at autopsy. The heart and mediastinum were displaced to the right.

The sputum examination supplied evidence of the mixed infection commonly associated with gross pulmonary infection, the Pathological Department reporting *M. catarrhalis*, fusiform bacilli, spirochetes and Gram-negative bacilli. No tubercle bacilli were found.

The patient was acutely ill at the time of his admission. During the first week the temperature was of the remittent type ranging from 99° F. in the morning to 100·6° F. in the evening. The pulse was rapid (100 to 136). Respiration was embarrassed (26 to 32).

The prognosis was evidently grave but artificial pneumothorax seemed to offer a slender chance of diminishing toxæmia from the diseased and safe-guarding the opposite lung. A preliminary bronchoscopy was suggested with the object of cleaning the diseased lung before attempting to collapse it, and this was kindly undertaken by Dr. Scott Pinchin on March 10. The patient did not seem to be adversely affected by this procedure at the time but towards the end of the afternoon he sank into a state of coma and died.

AUTOPSY.

The post-mortem examination was made on March 12, and Dr. D. S. Page reported as follows :—

External Signs.—Underdeveloped youth of 18; extremely thin.

Thorax.—Pleuræ: Right cavity obliterated, but layers separated without much difficulty. Left normal, except for some adhesions towards the base.

Lungs: Right; the lung is slightly less than the normal size. The pleural surface is of a pinkish colour and entirely devoid of carbon pigment, contrasting sharply with that of the left lung, which is almost black. It feels cystic.

On section a large number of cavities full of pus are exposed. Many are about ½ in. in diameter, others are smaller. The lining membrane is smooth and thin, and is red in colour. At this level (i.e., after the first cut through the posterior part of the lung) only a small quantity of solid tissue intervenes between the cavities. A deeper section made after hardening in formalin and spirit exposed large areas of dense tissue reddish-brown in colour. Nothing resembling normal pulmonary tissue was seen, and no carbon pigment. Where they are exposed the bronchi appear normal, apart from inflammation. The left lung shows hypertrophic emphysema and small patches of purulent pneumonia.

Glands: There is a chain of enlarged inflamed glands in the right posterior mediastinum; these also are devoid of carbon in marked contrast with a small bronchial gland on the left side, which is deeply pigmented.

Pericardium and heart normal. No lesions were found elsewhere.

Microscopical Appearances.—The cavities are lined with ciliated columnar epithelium continuous with that of the bronchi which are seen to open into them; in some instances the muscle and elastic fibres can also be traced into the cavity walls. The intervening tissue consists largely of fibrous and fibro-cellular tissue with some collapsed and inflamed pulmonary tissue, but in some sections a few almost normal alveoli are seen. A striking feature is the large amount of unstriated muscle present; a thick layer is seen in the bronchi and scattered fibres around some of the cavities; in some sections the muscle is very abundant and has no obvious connection with bronchi.

COMMENTARY.

The case of bronchiectasis just described differs from those commonly encountered in certain important respects. An advanced degree of bronchiectasis was discovered when the patient first came under observation, although there was no antecedent history of chest disease other than asthma. The onset of symptoms of pulmonary sepsis was acute and led to the patient's death within a period of five weeks. X-ray examination indicated that the whole of the affected lung was cystic. At autopsy there was an entire absence of pigment and of normal pulmonary tissue in the affected lung, the tissues being replaced by innumerable cavities filled with pus. Microscopical examination proved that the cavities were formed from dilated and degenerate bronchioles.

The term congenital cystic disease of the lung has been used to include a variety of conditions in which a saccular state of the lung has been assumed to have been present from birth. The term cystic disease should, however, not be applied to conditions in which the "cysts" are formed from dilated bronchioles. The latter are examples of bronchiectasis and there is no valid reason for calling a dilated and saccular bronchiole a cyst even though the condition is due to congenital malformation. Congenital cystic disease of the lungs and congenital bronchiectasis are rare conditions, but there is sufficient evidence that both occur. An example of the former is described by Collins [1] who discovered a diffuse honeycombing of both lungs in the body of an infant who died of pneumonia when fifteen months old. The cysts which were filled with pus were lined with flattened epithelium, no columnar epithelium was evident in their walls and there was nowhere evidence of any connection between the bronchi and the cysts. A similar condition was found by Sydney Smith [2] in the lungs of an infant found dead. Its development indicated an age of about eight months of intrauterine existence and the condition of the lungs showed that breathing had not occurred. Both lungs showed advanced cystic degeneration, the cysts, which varied in size from that of a pin's head to a pea, being distributed throughout the whole of the lungs. The cyst walls were lined with flattened cells and no trace of cubical epithelium, of muscle or of elastic fibres was found. Such examples of cystic disease should obviously not be confused with the bronchiectatic dilatations which may so closely simulate the former condition. Koontz [3] has recorded the post-mortem findings in a small infant, dying twelve days after birth, whose lungs contained numerous small cysts (up to 3 mm. in diameter). In this case, although the "cysts" were lined by flattened epithelial cells, communication with the bronchi was evident, a constriction in the walls of these being apparently responsible for the dilatation of their distal extremities.

When an infant dies at or shortly after birth and pulmonary cysts or bronchiectatic sacs are discovered, we may assume a congenital defect as the cause, though in an infant of a few months old a bronchiectasis or a bronchiolectasis might perhaps be due to post uterine inflammation. When we encounter a cystic condition of the lungs in later life, though we may in certain instances have good reason to suspect a congenital origin, some doubts as to the ætiology may well be entertained. Yet sometimes there is valid reason for concluding that a terminal infection of

a congenitally deformed lung has been responsible for the fatal issue. Though, as Hart and Meyer [4] maintain, we can only be certain of a congenital abnormality when we discover it in the fœtus or the new born babe, the evidence that such a condition was present before a terminal infection is sometimes convincing.

What is the evidence for a congenital defect in cases similar to that described above? The history may throw some light on the problem. A history of bronchopneumonia or "congestion of the lungs" might well suggest that bronchiectasis was the sequel of an unresolved pneumonia. Cylindrical bronchiectasis may, as the result of endogenous or exogenous reinfection of a damaged lung, become rapidly converted into saccular bronchiectasis involving a whole lung. But the absence of any history of preceding pulmonary inflammation in the present instance is striking. Such an event is not likely to be forgotten by a mother, and there is no reason to doubt the accuracy of the history. The patient had had no acute pulmonary illness before that which caused his death. As Eloesser [5] has pointed out, the presence of gross sepsis and acute disease does not necessarily imply a post-natal origin. "Respiratory infection may not be a cause, but merely the factor which, changing an aseptic and symptomless cystic lung into an infected and suppurating one, makes disease manifest." The absence of pulmonary pigmentation has been regarded as evidence of a congenital defect. The term "pale or cystic degeneration" (Hart and Meyer), which has been applied to it, emphasises one of the most striking characteristics of the group. The lack of pigment was in the present instance also noted in the hilar glands of the affected side. Such an absence of pigment suggests that the lung had never functioned effectively. Koontz has emphasised the importance of this evidence in drawing attention to the contrast between the pigment-free portions of the lungs containing congenital defects and adjacent portions exhibiting acquired pathological lesions of a similar nature which are often deeply pigmented.

Our hope that the microscope might supply proof of a congenital abnormality was disappointed. Communication between bronchioles and cysts was evident and the character of the walls of the latter proved that they were in fact bronchial dilatations. An excess of unstriated muscle fibres scattered around the cavities was perhaps suggestive of disordered growth. Nevertheless though a diagnosis of infantile bronchiectasis cannot be entirely ruled out the evidence taken as a whole seems to justify a diagnosis of congenital defect.

My acknowledgements are due to Mr. Smart who kindly photographed the excised lungs, to Dr. J. V. Sparks who made the X-ray examination, to Dr. S. Roodhouse Gloyne for helpful criticism, and last but not least to Dr. D. S. Page who conducted the post-mortem examination and also supplied the pathological reports on the lung sections.

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[2] SMITH, SYDNEY. *Brit. Med. Journ.*, 1925, **1**, 1005.
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[4] HART and MEYER. Handbuch d. spez. Path., Anat. u. Hist., iii, 469.
[5] ELOESSER, L. *Journ. Surg., Gynæcol., and Obstet.*, 1931, **52**, 747.

PLATE I.

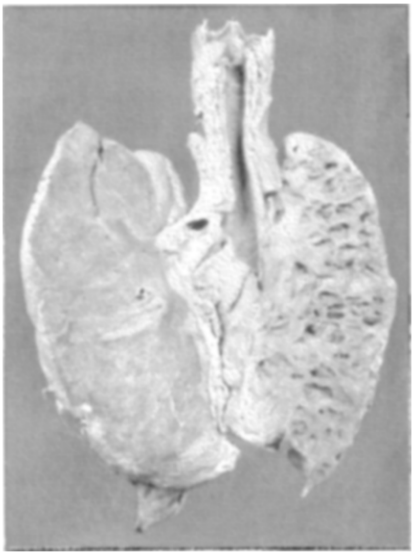

FIG. 1.

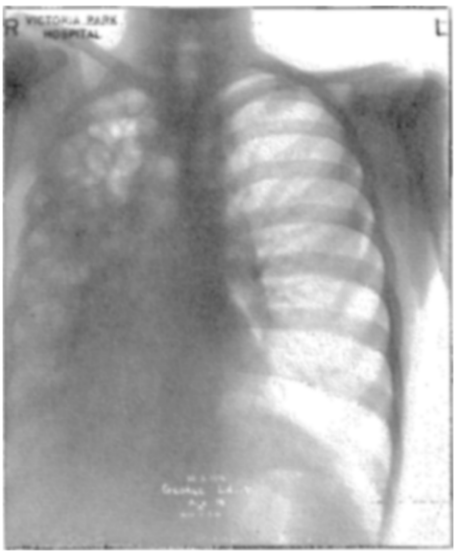

FIG. 2.

To illustrate article, "A Report of an Example of Congenital Bronchiectasis, with the Results of Post-mortem and Pathological Investigation," by W. BURTON WOOD, M.A., M.D.Cantab., M.R.C.P.Lond.

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