CYSTIC DISEASE OF LUNG — REPORT OF A CASE TREATED BY LOBECTOMY

E. A. CACHIA
M.B., B.Ch. (Manch.), D.C.H.

and

J. A. MUSCAT
M.D., F.R.C.S.

St. Luke's Hospital

A cyst or cysts of the lung may be a manifestation of disease of a widely varying nature ranging from congenital lobar emphysema to hydatid disease. An exhaustive list of pathological states in which a cystic condition of the lung may be present would also include such conditions as cavitational cancer of the lung, pulmonary tuberculosis and lung abscess. However, the term as used here is limited to a congenital condition of the lung in which a single cyst or multiple cysts are present, scattered throughout the parenchyma of a lung or confined to a lobe or to part of a lobe. Thus bronchogenic cysts of developmental origin but situated outside the substance of the lung, in the para-hilar region or in a fissure would be excluded Congenital lobar emphysema being considered as developing as a result of an inherent weakness of a lobar bronchus and lacking the appropriate epithelial lining is also excluded.

Lindskog describes the condition as resulting "from error in development of the bronchial buds. Such cysts are typically lined by ciliated mucus-producing epithelium, usually not communicating with the bronchial tree, and having muscle and cartilage within their walls". Occasionaly they are found to communicate with the respiratory tract.

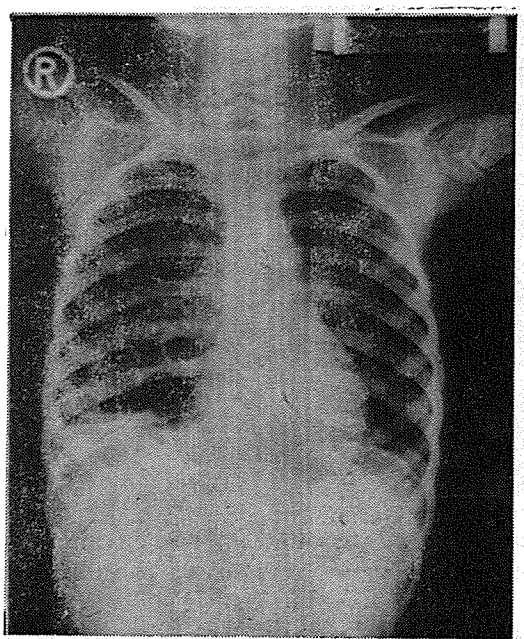
Other terms that have been used are: "pulmonary cysts, congenital cystic disease of the lung, pneumocysts, solitary and balloon cysts of the lung". The term "congenital bronchiectasis" is particularly confusing, but true bronchiectasis is said to be occasionally present at birth.

The matter of the lining is an important distinguishing point, but as Lindskog, Potts and others state, in the event of supervening infection it may be difficult or impossible to make out and to distinguish hence from an abscess of the lung.

The earliest mention of cystic lungs is probably by Thomas Bartholinus in 1687, (Leyden edition of Malpighius). H. Meyer published the first clinical report on this condition in 1859. By 1925 Koontz could collect 108 cases from the literature and add one of his own. In an early paper on Pulmonary Lobectomy by Robert and Nelson in 1933 one of the ten cases described had a pathological diagnosis of "Congenital cystic type of bronchiectasis" with "the lobe showing two large cysts and several small ones, and microscopic section showing that the epithelium was similar to that found in the trachea.

Case Report

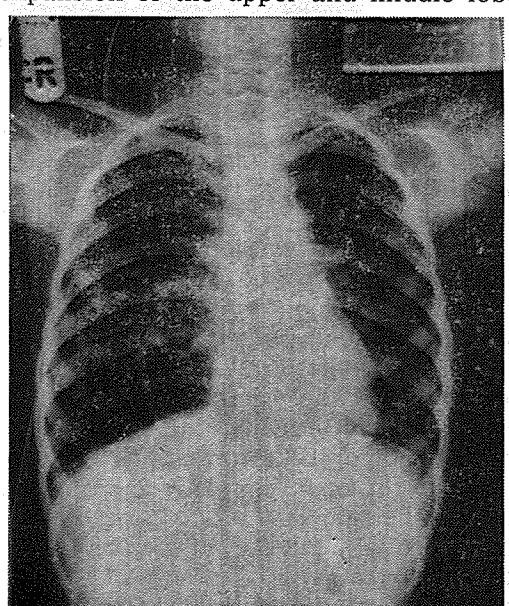
A girl aged 4 was referred to .St Luke's Hospital urgently on the 27th December, 1964, for "acute tonsillitis and severe arthralgia of the dorsolumbar spine". She had had a 3-day history of fever, pain in the back and vomiting. On examination she looked flushed and ill, her fauces being intensely injected; her chest was clear. She was put on 1 million units of Penicillin and 0.5 g. of Streptomycin daily. By the 29th she was showing no improvement; her temperature rose to 104 F. She complained of pain in the right hypochondrium and examination of the chest revealed diminished air entry to the right base. Investigations gave the following results:Hgb. 68%; W.B.C. 10,900 (Poly: 65%, Lymph. 20%; Mono: 15%); E.S.R. 40 mm. 1st. hr.; Mantoux and Serum



10. 3. 65: Showing cystic changes rt. lower lobe with adjoining areas of consolidation.

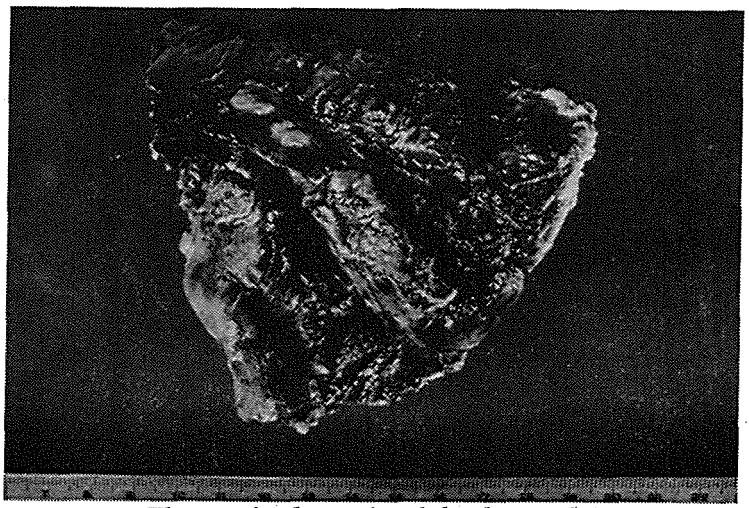
agglutination tests: negative. No protein and no casts were found in the urine. Throat and nasal swabs and a stool culture showed no pathogenic organisms. Examination of gastric contents showed no *Myco. tuberculosis* to be present. X-ray examination of the chest showed 3 cavities with fluid levels to be present in the right lung. There was a mottled consolidation of the right hilar region. On 21. 1. 65 the child was put on 750 mgm. chloramphenicol daily, and for the next 5 months she was put on erythromycin or chloramphenicol ,or penicillin and streptomycin. On the 4th January, 1965, she was given a blood transfusion (150 ml.). A bronchogram was performed on 6. 6. 65, which failed to show any filling of the right, middle and lower lobes.

Operation was performed on 3. 8. 65 under general anaesthesia (Dr. C. Podestà). The chest was opened through a right postero-lateral thoracotomy incision along the upper border of the sixth rib. The right lower lobe was found to be partially collapsed and adherent to parietal pleura over its mediastinal and diaphragmatic aspects. These adhesions were dense and vascular and their separation caused fairly severe bleeding ,not easily controlled. She was given 250 ml whole blood and 300 ml plasma over the operation. The right lower lobe was removed and the upper and middle lobes, which appeared normal to sight and palpation, could be made to expand quite readily. The chest was closed with one intrapleural drain. Post-operative progress was satisfactory. The drain was removed on the third day and the sutures on the tenth after operation. Chest X-ray (16. 9. 65) showed some residual pneumothorax. This cleared up slowly with full expansion of the upper and middle lobes.



21. 5. 65: Cysts still evident, the surrounding pneumonitis is less marked.

The Histological Report (Professor G. P. Xuereb) was as follows: Received in formol saline the lower lobe of the right lung measuring 10.4 × 9.7 × 5.5 cm. Part of the lobe is well aerated; its border is distinct. There are however, fibrous, haemorrhagic pleural adhesions over an area of lung 9 cm. from above down and 9 cm. from side to side; the pulmonary tissue at this side is replaced by cystic collapsed tissue. The specimen has been preserved whole. Microscopical examination shows haemorrhage and oedema within alveoli; there are areas of pneumonic consolidation with fibrosis, focal aggregates of lymphocytes, and eosinophilic infiltration. Large mononuclear cells are also present. There is no bronchi-



The excised cystic right lower lobe.

ectatic dilatation. Cystic lobe right lung; haemorrhage and oedema.

Discussion

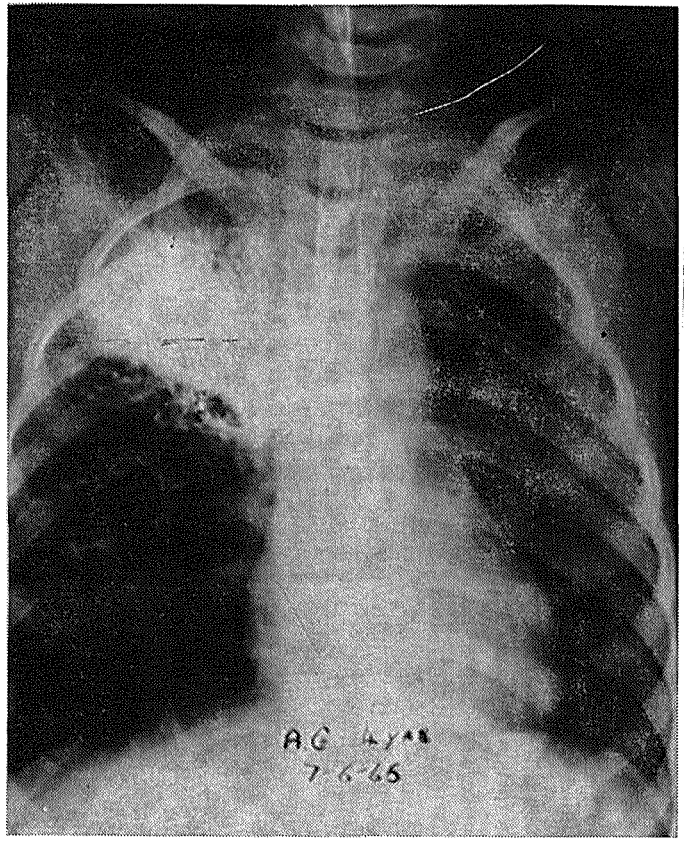
The true incidence of congenital cystic disease of the lung is difficult to determine. In a recent report on 372 lung resections in children and adolescents from H. Bruegger in Allgau, Germany, the presence of lung cysts or cystlike formations was the indication for operation only in 5 of the cases. In the same series there are 4 cases of congenital pulmonary emphysema. Moersch and Claget (1947) described a series of 44 cases seen over a period of 10 years at the Mayo Clinic. Most authors are in fact agreed that the lesion is a rare one but do not give figures.

The cysts commonly have a bronchial communication which predisposes them to the development of complications. These are mainly of two kinds. In the first place they may suddenly enlarge as a result of a sharp rise of tension within them and cause acute sypmtoms from the considerable compression and displacement of surrounding healthy lung and mediastinum. This may happen in very young children and may call for urgent and radical surgery. Gross (1946) describes a case where pneumonectomy was performed with success on a three week infant which developed this type of complication. They may secondly, become acutely infected giving rise to a severe pulmonary infection. This is the commoner event and occurred in 26 of the 92 cases studied by Lichtenstein (1953). In this latter series no fewer than 30 of the cases were symptomless and discovered on routine investigation.

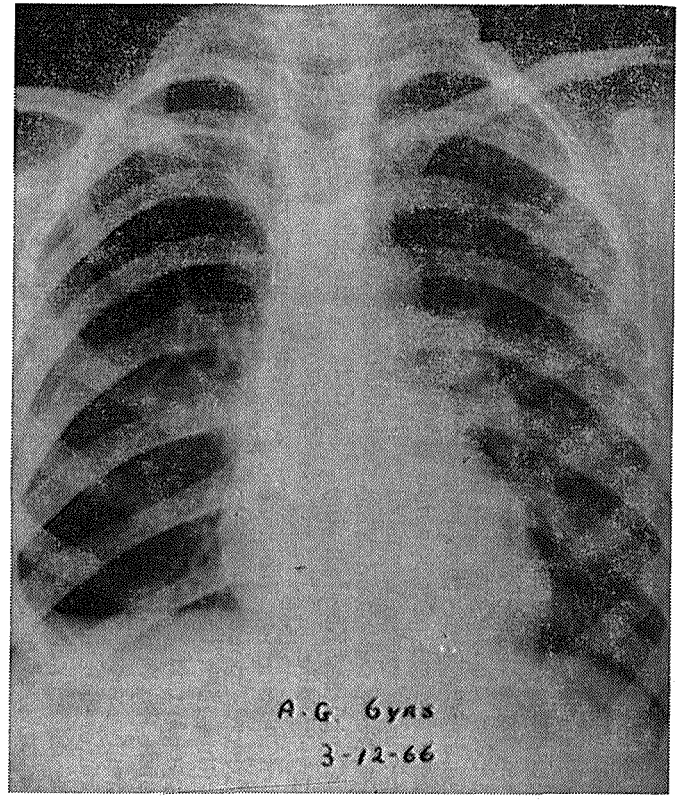
The case reported here presented as a severe pulmonary infection requiring parenteral antibiotic therapy and blood transfusion. Although the brunt of the infection was brought under control in a matter of 12 days it went on smouldering for 8 months until the diseased tissue was removed surgically.

A third group of cases may come to light as a result of investigations prompted by the presence of symptoms in patients who are not actually ill. Haemoptysis is one such symptom. This was present in 16 out of 22 cases studied by Dickson, Clagett and McDonald (1946). Other symptoms are cough and expectoration, pain and soreness of the chest and shortness of breath.

Although pre-operative diagnosis is often possible a definitive diagnosis may have to wait on an examination of resected tissue by the pathologist. The history is often suggestive and clinical examination will focus scrutiny on the region of the lung field involved. It is however on X-ray studies that a diagnosis is based. Plain X-rays in postero-anterior and lateral erect and supine projections will show up the cysts and provide information as to their contents and the presence and degree



Bronchogram — showing absence of filling of right middle and lower lobes.



3. 12. 66: The middle lobe is fully expanded.

of peribronchial inflammation.

Congenital cysts as Flavell (1957) underlines are generally large and more uniform than those seen in congenital bronchiectasis, and the degree of peribronchial inflammation is much less.

In infants and children a real difficulty exists in distinguishing congenital cysts from the alveolar cysts that develop on staphylococcal pneumonia. A thorough search for pathogenic strains of staphylococci from the respiratory and gastrointestinal tracts and from the blood is necessary. In the case reported here all such investigations were negative.

Other points of distinction from secondary staphylococcal pneumatocoeles are the rapidity with which they develop often in the late pneumonic phase of the illness, and the quick fluctuations in size they not uncommonly show (Potts and Riker, 1951). The differentiation is important because of the difference in management required for the two conditions; staphylococcal pneumatocoeles are often multiple and regress gradually. They may take a year to do so (Swenson, 1959). Other conditions which may have to be differentiated in children are fibrocystic disease of the pancreas, and Letterer-Siwe disease. In adults pulmonary tuberculosis with cyst formation, has to be excluded. In Moersch and Clagett's series six cases were being treated for pulmonary tuberculosis before diagnosis was established.

Pulmonary abscess, empyema, carcinoma and mediastinal tumour have also to be differentiated. Maier and Haight (1940) have underlined the difficulty of differentiating from empyema. Lindskog, Liebow and Glenn (1962) point out that when infection supervenes it may be difficult or impossible to distinguish an infected intraparenchymal cyst from abscess of the lung, and Sellors has written thus: "the treatment of cystic disease of the lung does not make satisfactory reading and the number of cases that have been aspirated or drained without further attempt at cure shows how little the condition is understood."

The treatment of choice is surgical excision of the diseased tissue, the preferred operation is lobectomy. In infected cases the management rests on appropriate antibiotic therapy coupled with supportive measures to control and contain the pneumonitis followed by bronchography in order accurately to localize the disease, followed by operation. Bronchoscopy is of little value and is not advised. More limited resection is advised by some but is seldom practicable. As eventual infection of the cyst is the rule surgery is mandatory in localised disease however mild the symptoms. This is generally regarded justifiable "in this era of safe thoracotomy".

The bronchogram of the case reported here showed normal filling of the right upper lobe but failed to show proper filling of the middle or lower lobes. At operation the middle lobe was found to be collapsed but expanded readily on removal of the diseased lower lobe. Thus the true extent of the disease may only be determined at operation.

Cysts that enlarge suddenly and cause acute respiratory embarrassment by displacement and compression may require urgent and radical surgery. Simple decompression by aspiration does not work and predisposes to pulmonary infection. It is generally regarded inadvisable. Infants and children stand pulmonary resection well. No adverse effect on growth and development has been noted following pneumonectomy in infants.

Summary

A case of a four year old girl presenting as an acute pulmonary infection and found to have cystic disease of the right lower lobe is here reported. A successful result followed lobectomy.

Some aspects of cystic disease (Congenital) of the lung are discussed.

References

Burch, B.H. and Miller, A.C. (1961) Surgical Diseases of the lung.

Dickson, J.A., Clagett, O.T. and Macdonald, J.R. (1946), J. Thor. Surg., *15*, 196.

Flavell, G. (1957). An introduction to Chest Surgery. Oxford University Press.

FLEMING (1934). Arch. Dis. Child., 9, 201.
GROSS, R. (1946). Ann. Surg. 123, 229.
KOONTZ, A.R. (1935). Bull. Johns Hopkins Hosp., *57*, 247.
LICHTENSTEIN, H. (1953). Dis. Chest, *24*, 646.
LINDSKOG, G.E., LIEBOW, A.A. and GLENN, W.W.L. (1962). Thoracic and Cardio-Vascular Surgery with related pathology. Appleton-Century-Crofts.
MAIER, H.C. and HAIGHT, C. (1940). J. Thor. Surg., *9*, 471.
MILLER, A.C. (1926). Arch. Surg., *12*, 392.
MOERSCH, H.J. and CLAGETT, O.T. (1947). J. Thor. Surg., *16*, 179.
POTTS, W.J. and RIKER, W.L. (1951). Arch. Surg., Surg., *61*, 684.
SELLORS, T.H. (1936). Tub., *20*, 49.
SWENSON, O. (1959). Paediatric Surgery. Staples. Appleton-Century-Crofts.