INTRALOBAR PULMONARY SEQUESTRATION

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The sequestration within the lung of a mass of abnormal pulmonary tissue, the blood supply to which is systemic and not pulmonary, is an uncommon but clinically recognizable entity. The clinical and pathological features are reproduced from case to case with remarkable consistency, and it is important to recognize the condition because it produces symptoms, is amenable to surgical correction, and does not respond to management other than surgical. There are reasonable grounds for regarding the lesion as a congenital abnormality of vascular supply to the lung ; the sexes are equally affected, and the left lower lobe is the most frequent site in which the abnormality occurs.

Eight previously unreported examples of intralobar pulmonary sequestration are recorded, and the clinical and radiographic features and the pathogenesis are briefly discussed.

CASE REPORTS

CASE 1.—A 12-year-old girl without complaints was submitted to routine chest radiography and found to have an air-containing cyst lying posteriorly in the right lower lobe (Fig. 1). As part of her investigation a bronchogram was made and the cyst did not fill with radio-opaque oil (Fig. 2). In 1957 at right thoracotomy she was found to have a cyst 4 cm. in diameter in the right lower lobe, and the cyst was dissected from the lobe with the division of one small bronchus. A pulmonary artery supplying the cyst was not identified. After its dissection from the lung the cyst was left pedicled on an artery 6 mm. in external diameter arising from the descending thoracic aorta and traversing the right pulmonary ligament. The artery was controlled, the ligament divided, and the cyst amputated. The defect in the lower lobe was closed by the approximation of the visceral pleural edges. Recovery was uneventful and the girl is well three years later. Histologically the cyst wall was composed of fibrous tissue without an epithelial lining and the wall was thickened in some places, where it contained calcified plaques.

CASE 2.— A 6-year-old girl was investigated because of recurring respiratory illnesses and because of an opacity in the left lower lobe which increased in size with each respiratory illness and failed to disappear completely between the illnesses. The opacity had first been noticed when the girl was 4 years old. The appearances on postero-anterior and lateral views of the chest (Figs. 3 and 4) and on bronchograms (Fig. 5) were typical of those of left lower intrapulmonary sequestration. In 1959 at thoracotomy the part of the left lower lobe normally occupied by the basal segments was of rubbery consistence and dark grey in appearance. It was clearly demarcated from the normally aerated apical segment of the lower lobe and from a 2 cm. fringe of normal lung along the oblique fissure, which was complete. The upper lobe looked and felt normal but was unusually small. Two systemic vessels, one 3 mm. and one 7 mm. in diameter, originated from the descending thoracic aorta close to the diaphragm and traversed the pulmonary ligament to enter the abnormal area of pulmonary tissue. These vessels were divided between ligatures, and the abnormal lung was then cut off the aerated parts of the lower lobe by sharp dissection. without the sacrifice of any normal pulmonary tissue and without residual blood or air leak from the normal parts of the lower lobe. A vessel or bronchus of such size as to require control was not encountered during this dissection. The visceral pleural edges were approximated to close the defect in the lower lobe. Recovery was uneventful and the child is well nearly two years later.

The resected specimen measured $9 \times 6 \times 4$ cm. and its lower surface was concave to accommodate the dome of the diaphragm. On section it was shown to be composed of numerous cavities containing pus or thick mucus. Histologically, fibrosis and lymphocytic infiltration of the interstitial tissues were seen. The cysts were lined by columnar epithelium. in places ciliated. No alveolar tissue or cartilage was found in the specimen. The arteries entering the specimen were said to resemble pulmonary rather than systemic arteries, on the ground of their content of elastic tissue.

CASE 3.—A 55-year-old man, a distillery worker, was investigated because of recurring respiratory illnesses over three years. A left lower pulmonary opacity (Figs. 6 and 7) persisted during a period of observation. The bronchi were normal bronchoscopically ; a bronchogram was not made. In the
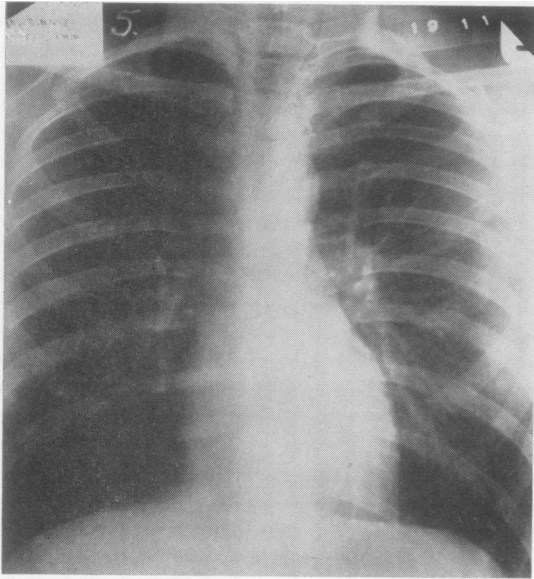
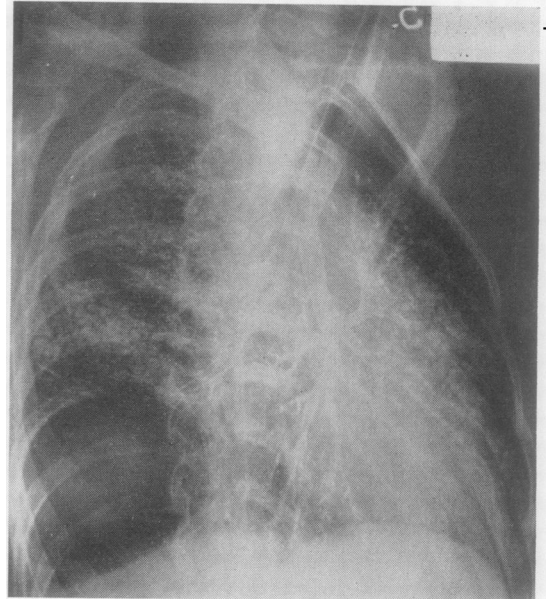FIG. 1.—Case 1: Postero-anterior view of the chest showing the cyst above the right dome of the diaphragm.

FIG. 2.—Case 1: The cyst is most clearly seen in the left oblique bronchogram. The left bronchi are normal; radio-opaque oil does not enter the right lower cyst.
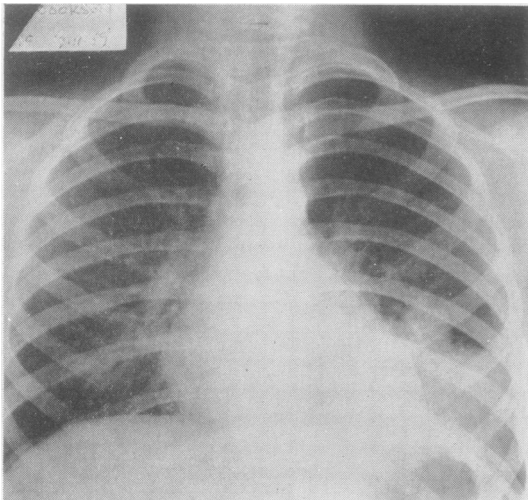

FIG. 3.—Case 2: Postero-anterior view showing the opacity in the left lower lobe.
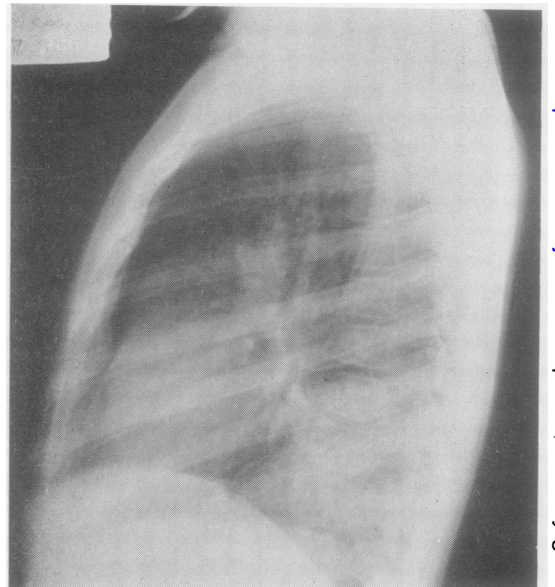

FIG. 4.—Case 2: In the left lateral view the opacity is seen to lie posteriorly in the lower lobe.

belief that the opacity might represent an empyema, thoracentesis was undertaken and 10 ml. of thick pus was aspirated. At the time of aspiration this was accepted as confirmatory evidence of the diagnosis of an empyema, but subsequent films showed a left pneumothorax without a fluid level, and the opacity still in the left lower lobe. The pus was now recognized as having come from the lung, and because of the chronicity of his symptoms and the position of the opacity, the diagnosis of intralobar pulmonary
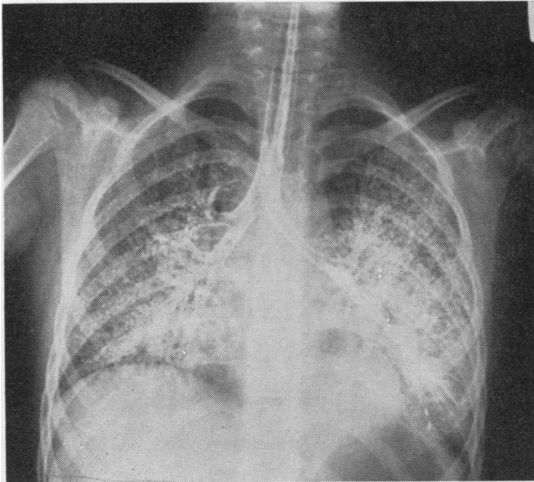
FIG. 5.—Case 2: The opacity in the left lower lobe does not fill with radio-opaque oil, and the left basal bronchi are displaced.

sequestration was made. The findings at left thoracotomy in 1957 were typical of this lesion. The systemic arterial supply to the sequestered mass measured 9 mm. in diameter and traversed the pulmonary ligament from its origin from the descending thoracic aorta. Left lower lobectomy was undertaken. The histological report on the resected specimen was similar to that reported in Case 2, but in this instance cartilage was recognized.

CASE 4.—A 28-year-old woman was radiographed as a contact of a relative found to have pulmonary
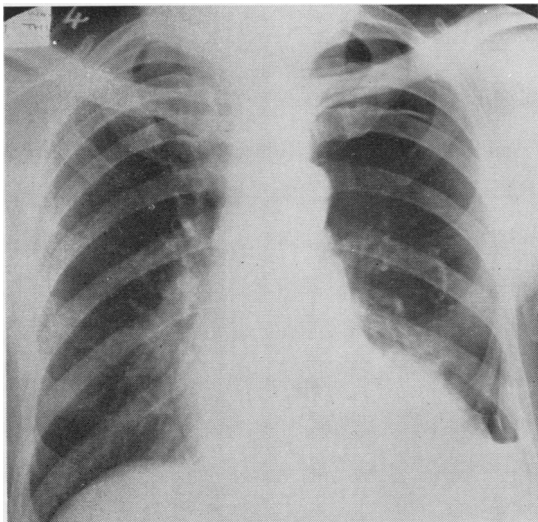
tuberculosis. She was shown to have a left basal pulmonary opacity. She denied symptoms, but the opacity persisted during a period of observation ; tubercle bacilli were not found in the sputum, other routine investigations were normal, and she was therefore, in 1957, submitted to left thoracotomy. The operative findings were similar to those described in the preceding case, except that the apical lower segment was large. Left basal segmental resection was undertaken ; the pulmonary arterial supply to the basal segments was very small, and the systemic artery in the pulmonary ligament measured 8 mm. in diameter and arose from the descending thoracic aorta. The histological findings were similar to those in the two preceding cases. The patient is well nearly four years later.

CASE 5.—A 17-year-old boy was investigated because of pneumonic illnesses recurring over five years. The radiographic and bronchographic appearances (Figs. 8 and 9) suggested the diagnosis of right lower intralobar pulmonary sequestration. In 1955 at right thoracotomy findings similar to those described in the preceding cases were managed by right basal segmental resection ; the pulmonary arterial supply to the basal segments was again unusually small, and the systemic artery supplying the sequestration measured 8 mm. in diameter. This systemic artery was traced across the pulmonary ligament to the oesophagus, alongside which it passed through the hiatus oesophageus, where it was ligated. Its subdiaphragmatic origin was not ascertained. Recovery from thoracotomy was uneventful ; the

FIG. 6.—Case 3: Postero-anterior view showing the opacity in the left lower lobe and a small residual pneumothorax.
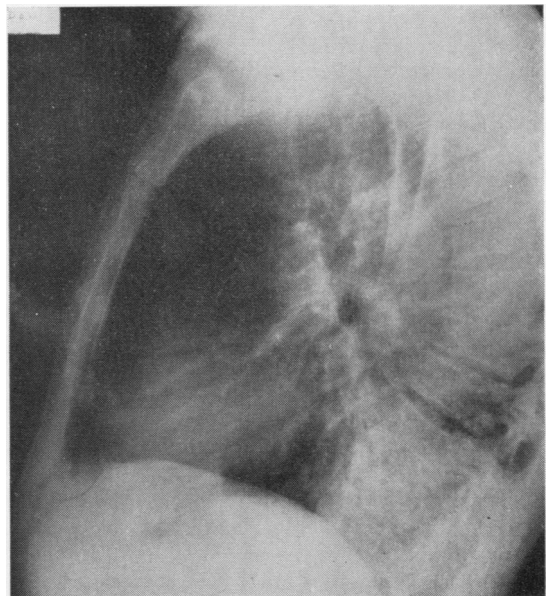

FIG. 7.—Case 3: Left lateral view showing the opacity lying posteriorly in the lower lobe.
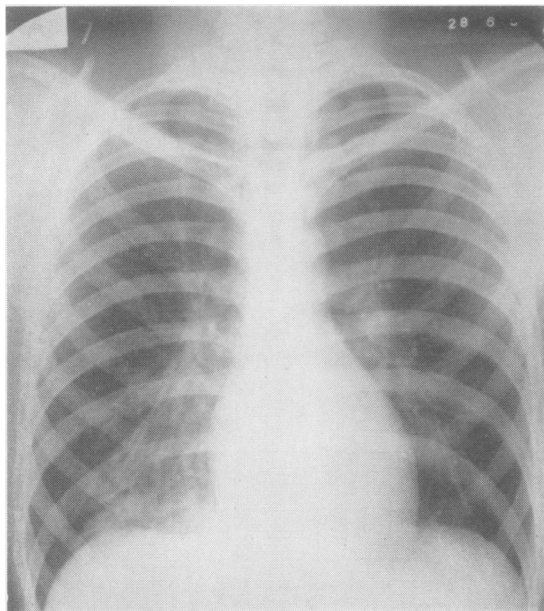
FIG. 8.—Case_5: Postero-anterior view showing the opacity in the right lower lobe.
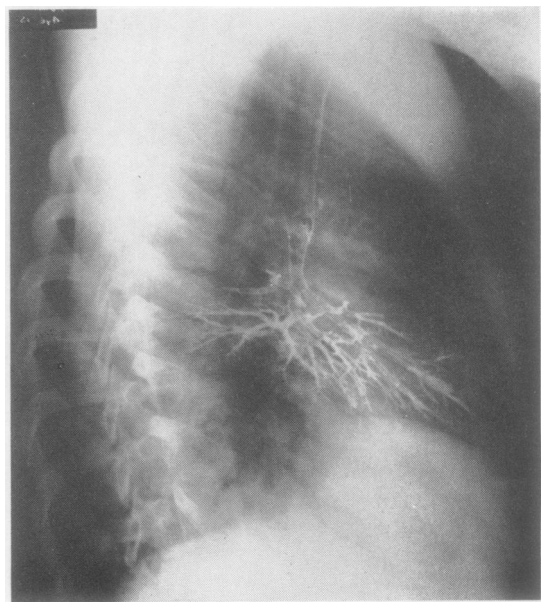

FIG. 9.—Case 5: The opacity in the right lower lobe does not fill with radio-opaque oil, and the right basal bronchi are displaced.

histological appearances were typical and the boy is well five years later.

CASE 6.—An 18-year-old boy was investigated because of recurring respiratory illnesses over three years. He was shown to have, radiographically (Fig. 10), and, in 1955, at thoracotomy, a typical left lower pulmonary sequestration of intralobar type. The systemic arterial supply in this patient measured 1 cm. in diameter. Left basal segmental resection was undertaken, recovery was uneventful, and the boy is well nearly six years later.

CASE 7.—A 45-year-old housewife developed a respiratory illness after a gynaecological procedure. A left lower pulmonary opacity persisted for some weeks after symptoms had been relieved, and further investigations, including bronchography, suggested that she might have a left lower intralobar pulmonary sequestration (Figs. 11 and 12). In 1949 left lower lobectomy was undertaken. The systemic arterial supply to the sequestered mass measured 7 mm. in diameter, and within the sequestration there was a pus-containing loculus 5 cm. in diameter. Recovery was uneventful ; from the respiratory point of view the patient was well in 1959, but required treatment for systemic hypertension.

CASE 8.—In 1951 an 8-year-old boy was admitted acutely ill with respiratory symptoms and high fever, and an opacity in the right hemithorax compatible with the diagnosis of either a lung abscess or an empyema (Figs. 13 and 14). Pus was obtained on thoracentesis, and the loculus of pus was drained by

rib resection. The appearances at rib resection were those of lung abscess rather than of empyema. Drainage was undertaken under local anaesthesia, and when the boy coughed the walls of the cavity closed round a finger introduced into it through the defect in the chest wall. The pleural space was obliterated. On serial radiographs over several weeks the drained
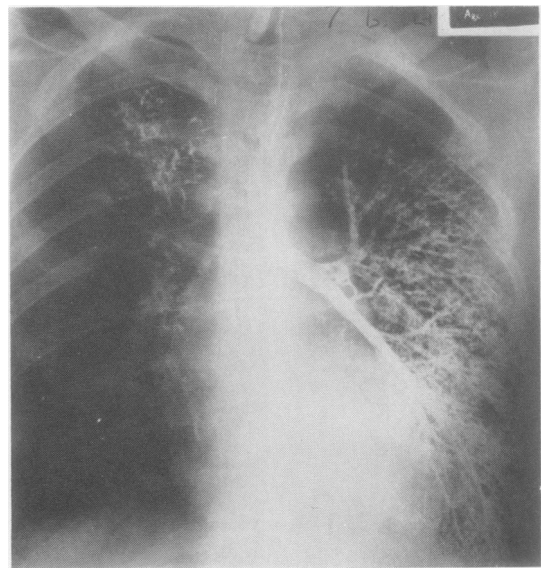

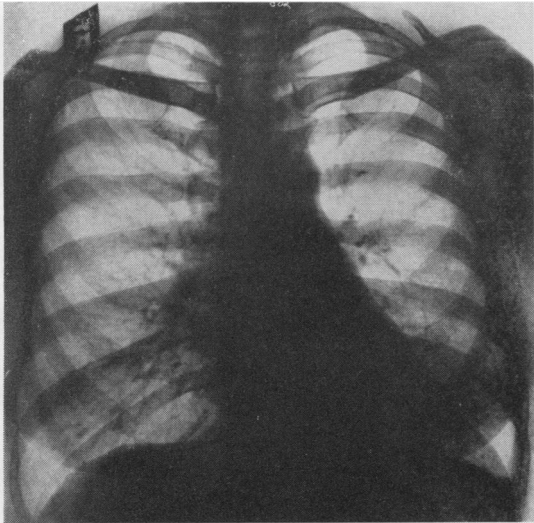FIG. 10.—Case 6: A bronchogram typical of a left basal sequestration.

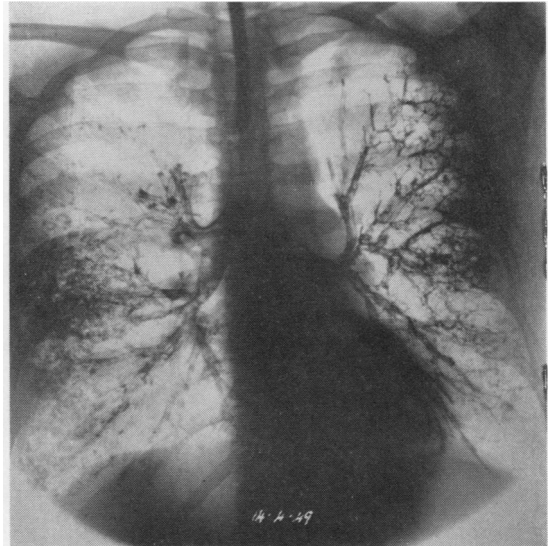FIG. 11.—Case 7: In the postero-anterior view of the chest the left basal opacity is difficult to see.

FIG. 12.—Case 7: The displacement and crowding of left basal bronchi are typical of a space-occupying lesion in the left lower lobe.
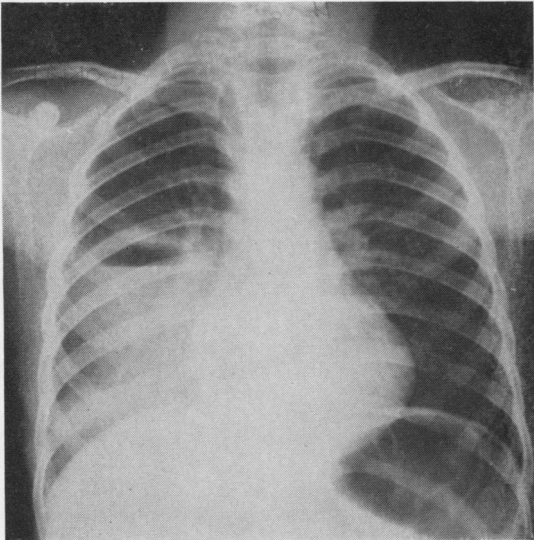
FIG. 13.—Case 8: Postero-anterior view showing the pulmonary or pleural abscess containing a fluid level.
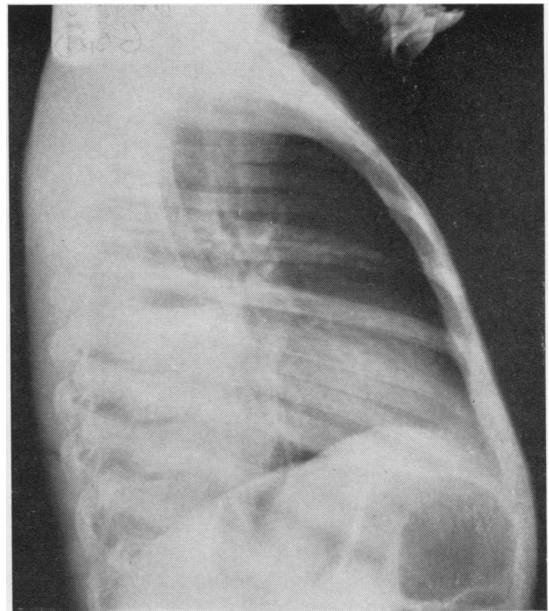
FIG. 14.—Case 8: In the lateral view the opacity lies sufficiently far posteriorly to make differentiation between lung abscess and empyema difficult.

abscess did not change in size, and when the cavity was filled with radio-opaque oil to demonstrate more clearly its outline, an incomplete retrograde broncho-gram was made. It was decided to submit the child to right thoracotomy, and he was found to have a pulmonary sequestration in the right lower lobe, an abscess in which had been drained. Right lower lobectomy was undertaken, and the boy is well nine years later. The systemic arterial supply to the sequestered part of the lobe entered the hemithorax

through the right dome of the diaphragm, at the level of which it measured 8 mm. in diameter.

DISCUSSION

There are several theories regarding the aetiology of intralobar pulmonary sequestration. Gallagher, Lynch, and Christian (1957) have summarized them and, together with most recent authors, accept the theory of Smith (1956) as explaining most of the features of the disease. The primary fault is probably a failure of the pulmonary artery to develop sufficiently to supply the furthest parts of the lung. The arterial supply to the lung from the dorsal aorta, a supply that normally regresses during embryological development, persists to that part of the lung inadequately supplied by pulmonary arterial blood. The pulmonary loculus, sequestered from its normal blood supply, develops abnormally, perhaps because it is subjected to systemic arterial pressure.

The basal segmental areas of the lower lobes, particularly the left lower lobe, are furthest from the pulmonary hilum and the origin of the pulmonary artery, and this might account for the fact that the lesion is commonest in the left lower lobe. The pulmonary artery to the lobe harbouring the sequestration is always smaller than normal (Smith, 1956). Lemmon, Kirklin, and Dockerty (1954) have shown, by the use of coloured vinyl acetate injection techniques, that the anomalous systemic artery is distributed only to the sequestration, and the pulmonary artery supplying the lobe in which the sequestered segment lies is distributed only to normal, aerated parts of that lobe. The vessels in the sequestration are systemic in type and thick-walled, and lie in the centre of the abnormal area where disorganization is most advanced, perhaps suggesting a relationship. There is no significant anastomotic circulation between the systemic arteries supplying the sequestered segment and the pulmonary arterial supply to the parent lobe.

More proximal deficiency of development of the pulmonary artery may result in hypoplasia of the subtended lung (Maier, 1954). The systemic arterial supply to such a hypoplastic lung is abnormally large, and yet the clinical features of pulmonary sequestration do not develop. Such a hypoplastic lung is, however, particularly prone to infection. The typical lesion, found apparently exclusively on the right, is a small unilobar lung. The bronchial distribution to this may be either totally anomalous or anatomically recognizable: for example, the bronchial anatomy may be that of a normal right upper lobe; the pulmonary arterial supply is through a small pulmonary artery of normal origin; and the systemic arterial supply is often in the distribution of normal bronchial arteries, but of abnormally large size. This lesion occurs in the absence of congenital cardiac anomalies such as the tetralogy of Fallot, in which the bronchial arterial supply to the lungs is also abnormally large.

CASE 9.—The bronchogram from a 4-year-old boy is shown (Fig. 15). The bronchial anatomy outlined is that of only the upper lobe on the right side. The boy was submitted to right thoracotomy in 1950 because it was thought that his recurring respiratory infections were probably a consequence of bronchiectasis in an undemonstrated shrunken middle and lower lobe. At thoracotomy the pulmonary artery to the unilobar right lung was almost threadlike, although patent. No remnant resembling the middle and lower lobe was found, and the systemic arteries to the right lung were four in number, two in the pulmonary ligament, one along the stem bronchus, and one from the superior intercostal artery crossing lateral to the azygos vein. The systemic arteries measured 4–6 mm. in diameter. No resection was undertaken because the unilobar lung was regarded as valuable packing, preventing mediastinal shift and overdistension of the left lung. The boy is well 10 years later without respiratory symptoms.

Although all the earlier recorded cases were in adults, intralobar pulmonary sequestration has
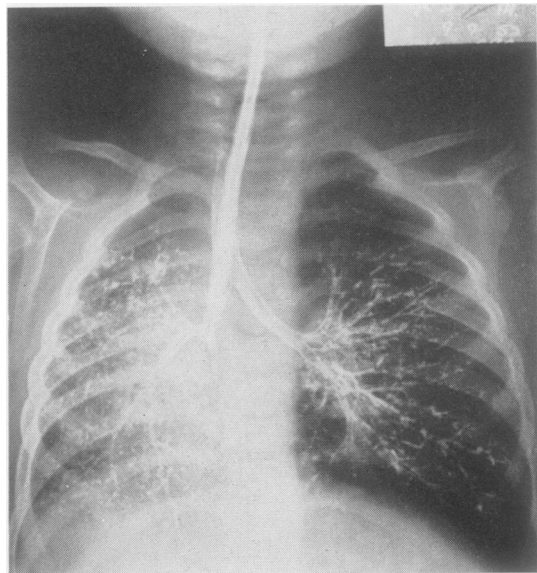

FIG. 15.—Case 9: A bronchogram showing, on the left, the normal disposition of bronchi, and, on the right, bronchi conforming with the normal anatomy of only the right upper lobe.

from its first recognition (Huber, 1777) been regarded as probably of congenital origin, because of the associated abnormalities of systemic arterial supply to the abnormal area. Gebauer and Mason (1959) believe that the primary lesion is an inflammatory one, and that the arterial supply is nothing more than an inflated bronchial artery. It is true that the usual course of the systemic arterial supply to the common left lower sequestration is from the descending thoracic aorta, traversing the pulmonary ligament, that there is normally in the pulmonary ligament a bronchial artery, and that in left lower lobar bronchiectasis, especially in children, this bronchial artery is large. There are, however, many recorded cases of pulmonary sequestration, the systemic arterial supply to which has arisen from below the diaphragm (Batts, 1939; Pryce, Sellors, and Blair, 1947; Findlay and Maier, 1951; Bergmann and Flance, 1956). These systemic arteries may traverse the cupola of the diaphragm (Case 8) or may enter the thoracic cavity through the hiatus oesophageus (Case 5). It is difficult to envisage the migration of systemic arteries from below the diaphragm to supply pulmonary tissue at the site of an acquired inflammatory lesion, whereas it is acceptable for a congenitally anomalous part of the lung to retain its arterial supply from the dorsal aorta; and for this supply to traverse the diaphragm is in no way remarkable if the systemic arterial supply to the lung is established before the development of the central part of the diaphragm. Although most cases are still recognized in adults or adolescents, three cases in the present series were of children under 12 years, and a case in infancy has been recorded (Simopoulos, Rosenblum, Mazumdar, and Kiely, 1959), in which the first radiographic abnormality was detected in the first few days of life, the typical lesion being observed to develop over the subsequent years.

The constant abnormality is an artery of considerable calibre entering the lung from the systemic circulation. The commonest site of intralobar pulmonary sequestration is the left lower lobe, in the vicinity of the posterior basal segment, and the abnormal artery usually arises from the descending thoracic aorta, entering the lung by traversing the pulmonary ligament. Right lower lobar intrapulmonary sequestration is less common than left lower sequestration (the proportion is about 2:1 in the reported cases) and there are rare examples of sequestration elsewhere in the lung (Cohn and Hopeman, 1955). The abnormal systemic arterial supply may arise from an intercostal artery or from a subdiaphragmatic source.

The abnormal pulmonary tissue usually occupies about two-thirds of the lower lobe. In more than half the reported cases the mass is polycystic, the cysts containing glairy mucus or pus (Cases 2–6); in some of the remaining cases most of the sequestration is represented by a single large cyst, sometimes containing air (Case 1) and sometimes pus (Case 8), and in either instance the single cavity is surrounded by compressed and fibrotic lung. A polycystic mass with one large and several smaller cysts may also be found (Case 7). Occasionally the sequestered mass contains recognizable bronchi branching parallel to the ramifications of the systemic arterial supply. A sharp line of demarcation between the polycystic variety of sequestration and the rest of the lobe may be found, and demarcation may be so clearly defined that the abnormal lung can be cut cleanly from the normal (Case 2). The absence of pigment in the sequestered loculus is evidence of absence of function in common with the rest of the lung (Pryce *et al.*, 1947). In 10 to 15% of cases a bronchogram has outlined bronchi within the sequestration (Lalli, Carlson, and Adams, 1954). In the remainder, bronchial tissue, although sometimes recognizable histologically within the abnormal mass, does not communicate with normal lung, and significant air leaks do not result when the sequestration is amputated from the rest of the lobe. For this reason lobectomy is probably an extravagant resection, justifiable only in an adult when the aerated pulmonary tissue in the affected lobe is little more than a shell about the sequestration, and particularly to be avoided in a child in whom the remaining part of the lobe is likely to grow. Alveoli are not recognizable histologically within a sequestration. The anomalous systemic artery has elastic rather than muscular walls and is said to resemble a conducting vessel such as a large pulmonary artery rather than a muscular systemic artery (Smith, 1955; Gerard and Lyons, 1958); arteriosclerotic changes are often present in this artery. Venous drainage is pulmonary, and not to the azygos or hemiazygos systems. In the rare condition of extrapulmonary sequestration, in which pulmonary tissue in addition to the normal complement is found in the mediastinum separate from the normal lung, the blood supply may be both systemic and pulmonary, but venous drainage is systemic. In the 12-year period during which the eight cases of intrapulmonary sequestration were encountered one so-called extrapulmonary

sequestration or accessory lung was seen (Schofield, 1955).

The route whereby the sequestered segment becomes infected is evident in those cases in which communication with the bronchial tree can be demonstrated ; the remainder probably become infected by contiguity of tissue, by lymphatic extension, or by haematogenous dissemination. Infection is an almost constant feature. Exceptionally an air-containing cyst in a lower lobe may be found by chance in a symptomless patient. Communication, however indirect, with the bronchi of the parent lobe must be present in such cases, but may not be bronchographically demonstrable.

As a corollary of the constancy with which infection is found pathologically, the clinical manifestations of recurring pulmonary infection are the most constant presenting features. The usual radiographic opacity is in a lower lobe, lying posteriorly against the mediastinum. On serial films the opacity may alter in extent, but does not disappear completely. Fluid levels may appear during episodes of acute infection, and may later disappear. When the opacity is a solid lesion it is usually ill-defined ; when it is a solitary air-containing cyst, or a cyst containing a fluid level, the definition is usually sharp. Visual examination of the bronchi is usually uninformative, apart from the demonstration of pus within the subtended segmental orifices and the absence of another cause for bronchial obstruction. Bronchography usually provides evidence of a space-occupying lesion (Figs. 5, 7, and 9), that is, one which displaces neighbouring bronchi ; in a small percentage of cases bronchi may be outlined within the opacity ; the number of the displaced bronchi in the remainder of the lobe is usually smaller than in the normal anatomical arrangement of bronchi in that lobe.

The diagnosis is most consistently achieved before operation when the possibility of encountering the lesion is kept in mind. A history of recurring pulmonary infections, the site and nature of the pulmonary opacity, the bronchographic findings, and awareness of the clinical entity will usually enable an accurate diagnosis to be made. Additional evidence of diagnostic value may be obtained by aortography (Kenney and Eyler, 1956) or by angiography (Gerard and Lyons, 1958), but these diagnostic adjuncts are not necessary and carry some risk to the patient. Where, however, the condition of pulmonary hypoplasia related to a deficiency of pulmonary

arterial supply is suspected from plain films and bronchograms (Fig. 15), angiography will probably clarify the diagnosis and thoracotomy may thus be avoided. Aspiration in the belief that the lesion may be an empyema is probably undesirable (see Case 3), unless the patient is acutely ill from infection (see Case 8). Pus may be aspirated from within the sequestration, so that the demonstration of pus does not confirm the diagnosis of empyema, and the accidental production of a pneumothorax, while of some diagnostic value, exposes the patient to the risk of pleural infection. Similarly, should the lesion prove to be a pulmonary tumour, diagnostic thoracentesis is equally undesirable, because of the concomitant danger of the dissemination of tumour in the pleural space and in the chest wall. On principle a patient with a persisting pulmonary opacity of unknown nature requires diagnostic thoracotomy, and this is a safe procedure. The diagnostic procedures outlined, without aspiration, aortography, and angiography, will have given sufficient lead to the diagnosis to avoid accidental rupture of the abnormal systemic artery to the sequestration, an event recorded in earlier cases as a cause of death. When the lesion is recognized, the resection of choice should be the most conservative possible.

SUMMARY

Eight previously unreported examples of intralobar pulmonary sequestration are recorded. The clinical and radiographic features and the pathogenesis are briefly discussed. The examples presented include all the common pathological variants. It is suggested that a resection more conservative than lobectomy is usually possible.

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