CLINICAL FEATURES OF INTRALOBAR SEQUESTRATION OF LUNG

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Intralobar bronchopulmonary sequestration is a condition that has received general recognition only in the past decade.

The lesion is congenital in origin and consists of a portion of lung which is not in communication with the main bronchial tree and is supplied by an artery other than the pulmonary artery. This vessel usually arises from the lower thoracic or upper abdominal aorta, and passes, directly or through the diaphragm, to the sequestered lung; it is elastic in type and often undergoes atheromatous change. The segment itself usually lies in the postero-medial part of one or other lower lobe. The mass is usually cystic, with evidence of bronchial structures in the cyst walls, shows inflammatory changes, and is devoid of visible carbon pigmentation.

Such segments are peculiarly liable to infection, and in the past have usually come to light as a result of recurrent pulmonary sepsis, although the correct diagnosis has often not been apparent until the aberrant artery has been discovered at operation. It is now evident that features exist which allow a strong presumptive diagnosis to be made even when recurrent infections have not occurred.

Ten patients, whose cases are reported below, were treated at the Connaught Hospital (Army Chest Centre) during a period of four years. The first two cases have been briefly reported previously (Davies, 1957).

Case 1

A signalman aged 19 was referred for investigation of malaise, productive cough, and right-sided pleuritic pain of four weeks' duration. Fifteen months earlier he had been treated for a right-lower-lobe pneumonia. During this illness there was radiological evidence of a large thin-walled lung cavity, which disappeared after treatment with antibiotics and postural drainage. On discharge from hospital his chest x-ray film had been reported to show no residual abnormality. After this illness he had continued to have a slight productive cough but had felt fit and led an active life.

The only abnormal physical findings on admission were diminution of breath sounds and a few rhonchi at the right base.

Investigations.—Postero-anterior (P.A.) and lateral chest radiographs showed a cyst 8 cm. in diameter, with a fluid level, situated in the posterior basal region of the right lower lobe. Lateral tomograms showed a multicystic mass with several fluid levels. A bronchogram showed a small communication between the largest cyst and the posterior basal segmental bronchus. Screening gave no further information. The sputum contained only normal respiratory flora, and cultures for *Mycobacterium tuberculosis* were negative. The Mantoux test was negative (1/100 O.T.); both the E.S.R. and blood count were normal.

At operation (right lower lobectomy) a large cystic mass was found in the postero-lateral part of the right lower lobe. The mass was supplied by a large thin-walled artery which came up through the diaphragm alongside the aorta to enter the lung in the inferior pulmonary ligament. The cysts contained gelatinous material, and the presence of a small communication with the normal bronchial tree was confirmed.

Case 2

A major aged 45 was admitted to hospital having suddenly coughed up about half a pint (280 ml.) of blood. He said he had a "smoker's cough" and had been producing about a teaspoonful of purulent sputum daily for the past four years. He had no other symptoms, and, apart from some crepitations audible at the left base, there were no other abnormal findings on examination.

Investigations.—The P.A. chest radiograph showed a cystic opacity in the left lower zone partially obscured by the heart shadow. The lateral radiograph showed the opacity to be located in the posterior part of the lower lobe. Tomography showed a cyst about 1 in. (2.5 cm.) in diameter. Screening gave no additional information. Bronchography showed a normal bronchial tree, but a small amount of dye reached the cyst through a minute communication with a subsegmental bronchus. A second bronchogram a few weeks later failed to show the communication. Bronchoscopy was normal. The sputum contained only normal respiratory flora, and cultures for *M. tuberculosis* were negative. The E.S.R. and blood count were normal.

At operation (resection of basal segments of left lower lobe) an ill-defined cystic mass was palpable in the posterior part of the lower lobe. The mass was supplied by a large artery arising from the aorta just above the diaphragm. Several cysts were present in the specimen, but no communication between them and the bronchial tree could be found.

Case 3

A guardsman aged 22 had a short influenza-like illness, after which he developed a cough producing small amounts of mucoid sputum, and a low-grade fever which persisted despite a course of penicillin. A chest radiograph showed an opacity in the right lower zone, and he was referred to the Connaught Hospital for investigation. There was no history of previous chest illness, and a mass miniature radiograph three years earlier had been passed as normal. On admission he did not look ill, and there were no abnormal physical findings.

Investigations.—The P.A. chest radiograph showed a group of five or six spaces, with fluid levels, and each about 1 in. (2.5 cm.) in diameter, lying medially in the right lower zone. The lateral film showed this polycystic mass to be situated posteriorly in the right lower lobe. Tomography and screening gave no further information. Bronchography showed a normal bronchial tree with no evidence of communication with the cysts. The sputum contained only normal respiratory flora, and cultures for *M. tuberculosis* were negative. The Mantoux test was positive (1/100 O.T.); the E.S.R. was 75 mm. in one hour (Westergren) but the blood count was normal. The diagnosis remained in doubt and the patient was treated conservatively, with antibiotics and postural drainage, for three months prior to operation.

During this time there was considerable change in the radiological appearances, which suggested the diagnosis. At first the cystic spaces became less apparent and the whole opacity appeared more dense. Later one large cyst, 8 cm. in diameter and containing a fluid level, appeared; subsequently the fluid level disappeared but the "hair-line" cyst wall remained just discernible. During this time the patient appeared fit, but intermittent blood-streaking of his sputum occurred.

At operation (right lower lobectomy) a large cystic mass was found in the position of the posterior and lateral basal

segments of the right lower lobe. This was supplied by several small arteries which came through the diaphragm from the abdominal aorta.

The cysts contained clear gelatinous material, and no communication with the bronchial tree could be demonstrated.

Case 4

An officer cadet aged 24 was treated for a left lower lobe pneumonia. Sputum cultures at that time produced a profuse growth of pneumococci. There was a good initial response to penicillin, but after one week his fever returned, he had several haemoptyses, and a chest radiograph showed a cavity to be present. He was then given chlortetracycline, to which there was a good clinical response. On discharge

FIG. 1.—Case 4. Postero-anterior radiograph of chest.

FIG. 2.—Case 4. Lateral tomograph.

from hospital he felt fit, and his chest radiographs showed only a small residual shadow in the posterior part of the left lower lobe. At a routine out-patient follow-up six weeks later a chest radiograph showed a cavitated opacity, and the patient was referred to the Connaught Hospital for investigation.

On admission he did not look well but was symptom-free, and there were no significant abnormal signs. There was no history of previous chest illness, and three routine mass miniature radiographs taken in the previous four years had been reported to be normal. As a child he had been in hospital for 10 months with suspected tuberculosis of the hip.

Investigations.—Chest radiographs showed an opacity, containing a thin-walled cyst 2 in. (5 cm.) in diameter, with a fluid level, apparently located in the posterior basal segment of the left lower lobe (Figs. 1 and 2). Tomography confirmed the presence of the cavity and surrounding opacity but gave no further information. Screening was normal, and bronchography showed a normal bronchial tree with no evidence of communication with the cavity. The sputum contained normal respiratory flora only. The E.S.R. and white-cell count were normal, and the Mantoux test was positive (1/1,000 O.T.). Bronchoscopy revealed no abnormality.

A provisional diagnosis of bronchopulmonary sequestration was made and thoracotomy arranged, but a few days before this was due to be performed sputum smears contained many acid-fast bacilli on three consecutive days. This coincided with the onset of cough and the disappearance of the fluid level.

On this account treatment with streptomycin and isoniazid was instituted and continued for three months. Serial chest radiographs showed the intermittent presence of fluid in the cavity and diminution of the surrounding opacity. No subsequent specimens of sputum contained acid-fast bacilli, and all cultures proved negative for *M. tuberculosis*.

At operation (left lower lobectomy) a cystic mass was present in the posterior basal region of the left lower lobe and was supplied by two large arteries which arose from the thoracic aorta just above the diaphragm. The mass contained two cysts, one filled with air and the other with gelatinous material; between the cysts there was a "honey-combed" area. There was no evidence of communication with the bronchial tree and no histological evidence of tuberculosis, and specimen cultures and guinea-pig inoculation were also negative.

Case 5

A major aged 37 was perfectly well until March, 1957, when he developed a productive cough and experienced several small haemoptyses. He was treated with penicillin and postural drainage, and soon became symptom-free. He was referred for further investigation as his chest radiographs showed a persisting opacity in the right lower lobe. There were no abnormal physical signs. He had a history of pneumonia at 3 years and of being "chesty" until 7; otherwise he had always been fit and athletic. Routine chest radiographs in 1953 and 1954 had been passed as normal.

Investigations.—The P.A. chest radiograph showed a rounded mass projecting beyond the right border of the cardiac shadow into the right lower zone. Lateral radiographs and tomograms showed a rounded mass about 6 cm. in diameter, with a small fluid level, in the postero-medial part of the right lower lobe. Screening and barium swallow gave no further information. Bronchography showed a normal bronchial tree with no evidence of communication with the lesion. Bronchoscopy was normal. The sputum contained only normal respiratory flora, and cultures for *M. tuberculosis* were negative. The E.S.R. and white-cell count were normal and the Mantoux test was positive (1/10,000 O.T.).

At operation (right lower lobectomy) a cystic mass was present in the postero-medial part of the right lower lobe; this was supplied by a large artery which passed up through the diaphragm. There were also several small arteries which came from the thoracic aorta. The cyst contained purulent material, and no communication with the bronchial tree could be demonstrated.

Case 6

A signalman aged 19 gave a history of three attacks of pneumonia affecting the right lung in childhood. An empyema occurred on one occasion, and was treated by rib resection and drainage. Subsequently he was fit and symptom-free for 10 years. Two months prior to his admission he had an influenza-like illness which had been complicated by a productive cough and one small haemoptysis. The cough persisted, and he continued to produce small amounts of sputum.

There were no abnormal physical signs beyond the scar from the rib resection in childhood and some limitation of movement at the right base.

Investigations.—The P.A. chest radiograph showed a rounded opacity about 1 in. (2.5 cm.) in diameter, with a fluid level, in the right lower zone. Tomography demonstrated a cystic mass situated postero-medially in the right lower lobe. Screening gave no further information. A bronchogram showed a normal bronchial tree with no evidence of communication with the cyst. The sputum contained only normal respiratory flora, and cultures for *M. tuberculosis* were negative. The E.S.R. and white-cell count were normal.

At operation (right lower lobectomy) a cystic mass was found in the postero-medial part of the right lower lobe which was supplied by an artery arising from the aorta just above the diaphragm. The cyst contained mucoid material. A small communication between it and the normal bronchial tree was present.

Case 7

A lance-corporal aged 38 had no chest symptoms until he developed an intermittently productive cough after an influenza-like illness. There were no abnormal physical findings. His only previous illness was due to a duodenal ulcer, for which a partial gastrectomy was performed in 1954—that is, four years earlier; his chest radiograph at that time was reported to be normal.

Investigations.—The P.A. chest radiograph showed an oval (2.5 by 5 cm.) cystic lesion in the right lower zone adjacent to the mediastinal shadow (Fig. 3). Tomography showed multiple cystic spaces apparently in the posterior basal segment of the right lower lobe (Fig. 4). Screening gave no further information. Bronchography revealed a communication between the posterior basal segmental bronchus and the cysts. Sputum cultures produced only normal respiratory flora; the E.S.R. was 30 mm. in one hour (Westergren); the blood count was normal and the Heaf test positive (grade III).

At operation (right lower lobectomy) a cystic mass about 5 cm. in diameter was found in the posterior basal region of the right lower lobe. Several small systemic arteries reached the mass via the inferior pulmonary ligament. It was not possible to demonstrate a communication between the mass and the bronchial tree.

Case 8

A trooper aged 36 was admitted to hospital complaining of right-sided pleuritic pain, a productive cough, and several small haemoptyses. He stated that for six years he had experienced recurrent episodes of productive cough and haemoptyses, although he had never produced large amounts of sputum or felt ill. There were no abnormal physical findings.

Investigations.—The P.A. chest radiograph showed an opacity, which contained an air space and a small fluid level, in the right cardiophrenic angle. The lateral film

showed the opacity to be confined to the posterior part of the right lower lobe. Tomography and screening gave no further information. A bronchogram showed a normal bronchial tree with no evidence of communication with the cystic area. Sputum cultures produced only normal respiratory flora. The Heaf test was positive (grade II); the E.S.R. was 60 mm. in one hour (Westergren), but the white-cell count was normal.

At operation (resection of basal segments of the right lower lobe) a cystic mass was found in the postero-medial part of the right lower lobe, and this was supplied by a small artery arising from the aorta. No communication between the bronchial tree and the cyst could be found.

Case 9

A lance-bombardier aged 20 was admitted to hospital for investigation of an abnormal shadow seen on a routine mass miniature radiograph. He was symptom-free on admission,

FIG. 3.—Case 7. Postero-anterior radiograph.

FIG. 4.—Case 7. Lateral tomograph.

but three months earlier had experienced right-sided pleuritic pain which lasted for five days. At the age of 10 he had been investigated for suspected pulmonary tuberculosis with reported negative findings; no details of this episode were available. There were no abnormal physical findings.

Investigations.—Chest radiographs showed a rounded shadow about 4 cm. in diameter in the right lower lobe. Tomograms showed this to be located in the region of the medial basal segment. Bronchography showed a normal bronchial tree; the lower lobe bronchi on the right side were displaced laterally by the mass. Sputum cultures produced only normal respiratory flora; the E.S.R. and blood count were normal and the Heaf test was negative.

At operation (resection of the basal segments of the right lower lobe) a mass 3.5 cm. in diameter was found in the medial part of the right lower lobe. This was supplied by an artery arising from the lower thoracic aorta. The mass felt firm and contained no cystic spaces on macroscopic examination. Histological examination showed the mass to consist of an outer wall of fibrous tissue and smooth muscle enclosing bronchioles lined by ciliated columnar epithelium and alveoli lined with cuboidal epithelium.

Case 10

This patient, a private aged 20, had no symptoms and there was no history of previous chest illness. An opacity had been noticed in the right lower zone on a routine mass miniature radiograph, and he was referred to the Connaught Hospital for further investigation. There was a congenital deformity of the thorax but no other abnormal signs.

Investigations.—Chest radiographs showed a cystic opacity about 4 cm. in diameter lying posteriorly in the right lower lobe. Tomography and screening gave no further information. Bronchography outlined a normal bronchial tree and failed to show any communication with the cystic mass. Sputum cultures produced only normal respiratory flora; the Heaf test was positive (grade III); the Casoni test was negative; and the E.S.R. and blood count were normal.

Thoracotomy was advised, but the patient refused this. He has since defaulted from the local chest clinic, but is known to have remained well for six months after discharge from hospital.

The radiological and bronchographic findings, normal E.S.R. and white blood count, with the absence of chest symptoms at any time, are considered to justify the presumptive diagnosis of bronchopulmonary sequestration.

Discussion

It is generally accepted that bronchopulmonary sequestration is congenital in origin, but there is no agreement concerning the mode of development of the condition. Several hypotheses have been put forward, and most of these presume the vascular anomaly to be the cause of the bronchopulmonary abnormality. The main views are: (a) that traction on the embryonic lung bud by the abnormal artery gives rise to the sequestration (Pryce, 1946); and (b) that insufficiency of the pulmonary artery supply to the developing lung bud results in persistence of vascular connexions between the primitive dorsal aorta and the developing lung (Jones, 1955; Smith, 1956). It has often been shown that branches of the pulmonary artery cannot be traced to the bronchopulmonary mass, which appears to be supplied by the aberrant artery alone.

It has been suggested that normal post-natal alveolar development fails because the affected lung tissue is perfused with oxygenated blood, and that the cystic changes occur after birth as a result of the systemic pressure of the blood supply (Smith, 1956).

More recently mechanical explanations of the pathogenesis of the defect have been questioned, and Boyden (1958) has expressed the view that there is no embryological evidence of a causal relationship between the arterial and bronchopulmonary anomalies. The variation in the combination of defects which can occur (Pryce, 1946), and the considerable variation in the size of the anomalous artery and its site of origin, are further factors which tend to suggest that the two components have an independent cause rather than that one causes the other. No satisfactory explanation for the liability of the sequestered lung tissue to become infected has yet been given.

The clinical features described in this condition are variable; typically there is a history of recurrent pneumonia, pleurisy, or empyema (Pryce *et al.*, 1947); the symptoms commonly begin in the second decade (Pryce, 1946); and the lesion is said to be more common on the left than on the right side in the approximate ratio of 3:2 (Smith, 1955).

In the present series only one patient (Case 6) had what is said to be the typical history of recurrent pneumonia and empyema, although two others complained of a chronic cough of some years' duration. Two patients had experienced a previous chest illness—a pneumonia-like illness in Case 1 and pleurisy in Case 9. Of the remaining patients, four (Cases 3, 4, 5, and 7) had no history of previous chest symptoms and one (Case 10) had no symptoms at any time.

The ages of the patients at the time of their first symptoms attributable to the lesion ranged from 5 to 41 years. Several cases have been reported in which symptoms have dated from early infancy (Lalli *et al.*, 1954; McDowell *et al.*, 1955; Jones, 1955), and in others symptoms have not occurred until after the age of 50 years (Pryce *et al.*, 1947). In this series the lesion occurred more commonly on the right side (eight cases) than the left (two cases). Haemoptysis was a frequent symptom and occurred at some time in five of the patients.

Despite the gross radiological abnormalities, usually including cystic spaces with fluid levels, the illness resulting in admission was not severe in most of the patients, and they usually became symptom-free after treatment with antibiotics. The E.S.R. was often within normal limits, and leucocytosis was exceptional. These features are of some value in differentiating the condition from others which might give similar radiological appearances such as bronchiectasis, lung abscess, staphylococcal pneumonia, and tuberculosis.

Postero-anterior and lateral chest radiographs show a cystic or solid mass usually located in the postero-medial part of one or other lower lobe. The bulk and character of the opacities in serial radiographs tend to change frequently (McDowell *et al.*, 1955); this was evident in those cases observed for longer than a few weeks prior to operation.

In this connexion it is of interest that five of the patients had had previous routine chest radiographs which had been reported as normal.

By definition a sequestered lung segment has no communication with the bronchial tree; when such a communication does exist with a lesion which satisfies the other criteria for description as a sequestered segment, it is considered to have resulted from infection (Smith, 1955). These communications are small; their presence may be suggested by the finding of fluid levels

on the radiographs, which were seen at some stage in six of our cases. Sometimes bronchography will show the communication; this occurred in three of the patients, and in two of these fluid levels were not seen on the radiographs.

Despite careful dissection a communication was demonstrated in only two of the specimens. The poor correlation between the radiological and the pathological findings suggests that spontaneous closure of the communication occurs. This may be one reason for the considerable changes in the appearance of serial radiographs.

Tomography accurately demonstrates the situation and extent of the lung lesion but in our experience fails to outline the aberrant artery. This vessel may not be demonstrated even with the aid of angiocardiology (Pinney and Salter, 1957). Anticipation of the presence of the vessel is essential in the event of surgical treatment, which might otherwise lead to catastrophic haemorrhage (Bruwer, 1955); but neither angiocardiology nor aortography appears warranted, since the likely presence of the artery should be inferred from the clinical and radiological characteristics of the condition.

Tuberculosis in association with bronchopulmonary sequestration has been reported on several occasions. The infection may involve another lobe of the lung (Tosatti and Gravel, 1951), the lung tissue adjacent to the defect (Edge *et al.*, 1957), or the sequestered segment itself (Boyd, 1953; Smith, 1955; Johnston, 1956).

In one of our patients (Case 4) thoracotomy was undertaken with a provisional diagnosis of bronchopulmonary sequestration and associated tuberculosis, since acid-fast bacilli had been present in the sputum. Sputum cultures subsequently proved negative for *M. tuberculosis*, and no histological or bacteriological evidence of tuberculosis was found on examination of the resected specimen.

The natural history of bronchopulmonary sequestration is incompletely known. Most of the patients reported have been surgically treated and have had histories of repeated or chronic chest illness; there have been no follow-up reports of patients who have been treated conservatively. Since some patients have no symptoms until later life, intervals of many years between respiratory illnesses (10 years in Case 6) are not uncommon, and some patients become symptom-free after treatment with antibiotics, it is possible that the outlook without surgery is not always one of chronic respiratory ill-health. Resection offers a complete and permanent cure and is bound to commend itself when no general contraindication to surgery exists; nevertheless, there is some justification for regarding the recurrence of infection, rather than its occurrence, as the indication for surgery.

Summary

The clinical, radiological, and gross anatomical features of intralobar sequestration of the lung are described. Case reports of 10 patients are presented, nine of whom were operated upon. It is suggested that in patients with this condition a strong presumptive diagnosis may be made clinically.

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THE PREVALENCE OF ANAEMIA IN THE COMMUNITY

A SURVEY OF A RANDOM SAMPLE OF THE POPULATION

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It is suspected that anaemia is common in Great Britain to-day, but, apart from some surveys of special groups of the community, no attempt has been made to determine its prevalence in the general population since the war.

In an attempt to obtain up-to-date information about the distribution of haemoglobin values, packed cell volumes, serum-iron and vitamin-B₁₂ levels, and mean cell diameters, we have carried out surveys in population samples of two defined communities—one industrial and the other agricultural. We report in this paper the results of our first survey, which was done as a pilot study in a South Wales mining valley—the Rhondda Fach—in association with the Epidemiological Research Unit of the Medical Research Council Pneumoconiosis Research Unit (see Cochrane *et al.*, 1955), who had planned a survey of chronic bronchitis, ischaemic heart disease, and rheumatoid arthritis there during May and June, 1958.

Sample and Procedure

Details of the sample are shown in Table I. During the weeks between the census and the actual start of the survey 21 names had to be withdrawn (8 persons had died, 11 had left the area, and 2 had given erroneous date of birth), leaving an available sample of 779 subjects, of whom 193 were women and 586 men. These 779 subjects were visited and invited to come to a centre in Ferndale, the largest town of the Rhondda Fach. A total of 723 (543 men and 180 women), or 92.8% of this available sample, were examined. Between 30 and 40 people were seen each day and each person was in the centre for about 45 minutes, during which time many tests were made. In view of this fairly full

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