Intralobar Pulmonary Sequestration*

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Pulmonary displacement associated with an anomalous artery has been frequently described since the advent of much chest surgery. The earliest adequate report of such a case is generally attributed to Tissler in his Düsseldorf thesis, unpublished but quoted by Müller.1 Earlier classical descriptions by Rektorzek2 (1861), and Humphrey3 (1884) and later Simpson4 (1908) are quoted by McCotter.5 These cases reviewed by McCotter were all autopsy cases in children under two years of age. In over 6,000 post-mortems at the Hospital for Sick Children in the past 25 years, there was no case. It would therefore appear a rare pathological finding after death and not a killing disease of the young, albeit it may be a severely crippling one. Extralobar sequestration on the other hand is usually an autopsy diagnosis, and rather likely to remain one.

Forty-two cases of intralobar sequestration have been reported in recent years, up to November 1951 (Cole6). Since then Kergin7 has published the record of five cases and Bruzzone8 of one. In this paper six further cases are described. They were all seen at the Sick Children's Hospital, and were diagnosed in part clinically preoperatively and the others by examination of the specimen after excision and by the finding of an aberrant artery when the portion of lung was removed.

The first of these cases diagnosed at the time of operation was in 1948. The resected lung was being discussed at a pathological conference as something of a problem. The surgeon, Dr. F. R. Wilkinson, arrived late, but having read Pryce's9 paper the night before, was able to distinguish himself as a diagnostician. Some of the cases reported in this paper were seen prior to this time, but diagnosed in retrospect. The surgeon had however, noted the systemic artery when he removed the mass. Pryce10 had reported the condition earlier but little attention appeared paid to the first paper until the second one was published. Brief mention of two cases was made by Haight11 in 1941. The term intralobar sequestration has been applied to these cases since Pryce's paper in 1948.

The purpose in reporting the following cases is to add to the list of published cases, and perhaps make some suggestions whereby clinical diagnoses can be made. Awareness of the anomaly and its clinical course make preoperative diagnosis easy. It has been made in all those seen since 1948.

Cases with pulmonary agenesis seen at the hospital have not been included in this paper despite the fact that most observers now consider them to be due to the same embryonic accident produced earlier. Brief mention should be made of three cases in which pneumonectomies were done. We would not subscribe to a theory attributing all kinds and degrees

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of cystic lung to amputated lung buds caused by aberrant arteries. Probably Kirkland's estimate of 5 per cent due to this etiology is right. The three cases alluded to would appear to belong in this minority group. They were in young children. No ordinary infection causing bronchiectasis could be inculpated. The changes involved the whole lung. Large cysts and honeycombed areas of tissue associated with alveolar hypoplasia were found. There was little or no shift of the mediastinum such as would be expected when such general involvement with an acquired case was found. The surgeon noted in each case the lack of disturbance to the child when the lung was removed. No anomalous vessels were found. This does not preclude their having been the teratogenic factor according to Rosenthal. He states that an anomalous artery may cause the severing of the lung bud, and then itself become separated from the aorta. If these cases may be considered as sequestrations, the amputation of the lung bud must have occurred between the 4 mm. and 14 mm. embryo. Bruwer states that the accident at the 4 mm. stage results in agenesis, and at the latter of sequestration of only a portion of lung. The only other teratological explanation of these lungs would be that there was interference with the growth of ducts and alveoli which according to Strukow normally continue to grow up to the seventh year. Müller states such faulty growth may be due to prenatal or postnatal inflammatory changes. This explanation would appear more applicable to those cases of cystic disease developing after birth and not confined to one lung.

**Etiology**

The production of a dislocated portion of lung with a systemic arterial supply is now considered by most writers to be produced by the amputation of a primitive lung bud by an anomalous vessel. This was pointed out by Cockayne in 1917. Berry demonstrated the same thing in 1941. The theory was supported by Pryce. Earlier explanation of such distortions of the lung as advanced by Eppinger and others are not acceptable now. They believed them due to an accessory anlage developed from the stomach. The age of the embryo at which the amputation occurs determines the extent of the damage. Thus early separation results in agenesis, or if our conception of the etiology of the pneumonectomy cases alluded to be correct, to extensive involvement of the whole of one lung. Later disruption involves only a portion of lung. Rusby and Sellors consider extralobar sequestration is due to still later disruption. The latter is therefore more likely to be associated with other congenital anomalies.

**Pathology**

Essentially these displaced masses are characterized by polycystic disease with alveolar hypoplasia. An aberrant artery enters this mass and sometimes supplies part of the adjacent normally developed lung. The cysts may be large blebs or small, giving the lung a honeycomb appearance. The mass may be within a lobe of lung or completely separate from it. The contiguous lung may be involved in the infection and show bronchi-
ectasis. Normal bronchi sometimes enter the mass. The bronchi however are usually represented only by blind pouches. Endarteritis is common. This was pointed out by Betts.21 Such changes might be due to increased pulmonary tension from the systemic blood supply. Bruwer considers them dependent on sepsis and the age of patient. They were not very marked in our cases.

Fatty changes of some degree are often seen. In one case they were of sufficient magnitude to suggest lipoid pneumonia. The case was similar to the one reported by Clagett et al.22 These writers attempted to relate the changes to the increased blood supply from the aberrant vessel, but ruled it out. They analyzed the fat and from their results decided the changes were due to endogenous tissue breakdown. In our case, four large vessels entered the mass, but a similar area appeared in another child’s lung with no anomalous vessel. It is strange if sepsis is the cause, that the changes are not more commonly seen in bronchectasis.

Kirklin12 pointed out that tuberculosis occurs as associated infection in not more than 5 per cent of lung cysts. Bruwer14 (1950) said no cases of associated sequestration and tuberculosis had been reported. Two cases in this series had tuberculosis. The sequestrated masses were formed by large cysts containing tubercle bacilli but in no wise resembling tuberculous cavities.

One case in this series showed calcareous plaques and cartilaginous tissue. Cartilage without bone was present in one other. Pryce reported similar changes in some of his patients.

The pleura is always thickened and becomes a shaggy surface to the resected specimen.

The abnormal artery most frequently comes from the abdominal aorta, but may arise from the intercostals. It may form a large leach of vessels or be a single large artery 0.5 to 0.7 cm. in diameter. Its systemic nature is indicated by its origin and the nature of its walls. These are thicker and more elastic than vessels of the lesser circulation.

Clinical Findings and Symptoms

Intralobar sequestration may be readily diagnosed clinically if such a possibility is borne in mind. The fact that no case was found at autopsy would suggest that such lesions may be quiescent until later in life, unless discovered by x-ray surveys. Symptoms develop when infection occurs. This may result from hematogenous spread of infection or from severe disease in the adjacent lung. The usual causative infections producing bronchiectasis in children are lacking.

The clinical history is fairly uniform. Some cough may be reported as occurring from birth. It is not usually serious and only becomes so when infection of the mass develops. Subsequently general deterioration in health is noted. There are recurrent bouts of fever. Cough is worse. There is usually little or no sputum. Offensive odour to the breath or sputum is infrequent. Severe hemoptysis may occur.

Several findings obtained on physical examination are significant. No
clubbing has been seen in our cases. The mediastinal shift compared to the size of the mass and apparent pulmonary collapse, is slight if any. This fixity is commonly noted at operation. The physical signs are usually greater and more permanent than in ordinary bronchiectasis.

Radiology

The x-ray film demonstrates the true nature of the case. The mass itself is often of considerable density, at times homogenous. Cystic areas are usually visualized in it or in nearby portions of the lung. The mediastinum is mid-line.

The bronchogram usually shows a mass into which no lipiodol penetrates. Occasionally, it may enter for a short distance. The demonstration of a complete bronchial tree, diseased or otherwise, extraneous to the mass, is important. Careful attention to this detail will prevent removal of a mass and leaving behind bronchiectastic portions of lung with the false assumption that disease is all excised.

Treatment and Prognosis

The only effective therapy is surgical removal of the abnormal lung plus any infected normally developed lung. The results are unusually good.

Case Reports

Case 1: A.B. (male) ae. 7, 1930. Child was admitted at age six because of mediastinal tuberculosis. In addition to the hilar lymph nodes, the radiogram showed a large homogenous mass in the right lower chest. The tuberculin test was positive.

FIGURE 1: A.P. of Chest showing cystic areas.
There was a large tuberculous ulcer in the right main bronchus. Five attempts were made to obtain a good bronchogram of this area without success. Lobectomy was done October 1932 after preliminary phrenicotomy. The mass was separated with great difficulty from the diaphragm and the adjacent pulmonary lobe. An intercostal artery entered that mass.

Specimen: There were many tough pleural adhesions. There was a large cystic cavity in the apex of this specimen. The posterior portion of the specimen showed practically no normal lung tissue. The great bulk of the lobe was filled by a large irregular cavity traversed by numerous partitions. The largest cavity measured 6 x 2 cms. There were many smaller ones. The cavities contained small amounts of pus. Microscopically the whole lung presented a similar picture. There were a few alveoli lined by cuboidal epithelium. There was little or no elastic tissue. Cilia were rare. The degree of destruction was out of all proportion to the fibrosis seen.

Case 2: R.A. (male) a.e. 5. Child had not been well for six months when he had an acute attack of bronchitis. Since that time he coughed a great deal, tired easily, and had bouts of high fever. His tuberculin test was positive as was also his sputum.

Right pneumonectomy was supposedly done in 1940 but eight years later his upper and middle lobes were found undisturbed at a subsequent operation.

At operation dense adhesions bound the lung to the chest wall and diaphragm. It was necessary to remove a portion of the latter to free the lung. There were also many adhesions between the mediastinum and the edge of the lung. In one of these was a fair sized blood vessel which came through the diaphragm from the aorta.

Specimen: The surface was ragged. There was a cavity 6 x 5 cm. in the lower portion. The medial portion of this extended toward the mediastinal border in a sort of bay 3 cm. long filled with yellow caseous material. No dilated bronchi were found. There was a large thrombosed vascular channel 6 mm. in diameter filled.
with thrombus. There was a large fibrosed vessel on the base. There were elastic fibers in its wall.

**Case 3:** N.G. (female) ae. 4. 1939. This child was in hospital in infancy with pneumonia. She was well after this until a few months before admission, when she complained of weight loss, lassitude, and later anorexia and cough. Subsequently she had frequent bouts of fever. The x-ray film presented is typical of the disease showing the left lower mass. Many attempts were made to fill this with lipiodol without success.

Left lower lobectomy was done in 1948. The lobe appeared grossly diseased. There was no air containing lung except at the upper part. The remainder of the lobe was firm on palpation. There were many adhesions in mediastinal and diaphragmatic areas. There was a large leash of vessels coming from the aorta four inches below the arch to the postero lateral aspect of the lobe about one and one half inches from the diaphragmatic surface. Three of these were as large as the radial artery.

**FIGURE 3:** Specimen of sequestrated mass. Note emphysematous blebs. Large artery entering the mass. Cut sections of mass.
Specimen: The lobe was or normal size. A consolidating process involved the whole of the postero-lateral portion. There were fibrous adhesions. The remainder of the lung was deep bluish color while the affected part was pale greyish yellow. Four large arteries entered the posterior aspect of lobe 2 cm. from the diaphragmatic surface. These arose directly from the aorta and broke into ramifications through the parenchyma (See celloidin injected lung picture). Water forced through with considerable pressure entered the pulmonary artery. The vessels appeared to be large bronchial arteries with a large anastomosis with the pulmonary arteries. The posterior basic and the middle half of the adjacent middle basic appeared to be the only parts involved. The posterior basic division was constricted about 2 cm. from the hilum. From this point on the bronchial divisions appeared as greatly dilated cystic spaces and fibrous tissue. They were between 1 and 1.5 cm. in diameter. Yellow granules were imbedded in both spaces and the fibrous tissue. They were filled with pus.

Microscopically all the bronchi and bronchioles were dilated cystic spaces filled with pus. Most were lined with columnar epithelium but some small spaces showed cuboidal epithelium. The cysts were separated by oedematous fibrous tissue with extensive lymphocytic infiltration, and great vascularity. There were some phago-

**FIGURE 4:** Artist's drawing of lung showing systemic vessel and blind bronchus.
cytes containing fatty material. The blood vessels were large and thick walled and had organized well developed elastic tissue. There was little elastic tissue in the walls of the bronchi. The remainder of the specimen showed the usual type of bronchiectasis and did not communicate with this portion.

Case 4: A.S. (female) ae. 13, 1940. This child had acrodynia at three years of age with cough ever since. One year ago she had pneumonia, since then cough has been much worse and of late has been accompanied by haemoptysis. Physical signs were all in the left chest. In this same area, there was a rounded mass behind the heart. It was impossible to fill this area with lipiodol but above it, the bronchogram showed definite bronchiectasis. Left lower lobectomy was done supposedly but later x-ray films showed the bronchiectasis still present and only the rounded shadow removed. At 18 this diseased area was removed at hospital for adults. Death followed two days later.

The first operation showed dense pleural adhesions. There was much haemorrhage from a vessel of fair size in the vicinity of the pulmonary ligament. Its origin was not determined. The lung was removed by excising proximally to a noose.

The specimen consisted of an irregular portion of lung. There were no normal landmarks. The pleural surface was shaggy and haemorrhagic. Palpation showed irregular consolidated areas. The cut surface showed large vessels and bronchi. The interventing lung tissue was fibrotic and consolidated. The bronchi as shown in the bronchogram were not found.

Sections showed general bronchial dilatation. The bronchi were lined with stratified columnar epithelium and were greatly thickened. The thickening was for the most part due to increased lymphoid tissue and infiltration with chronic inflammatory cells. The muscle coats were gone. There were a few islands of cartilage. There were also spicula of bone in one fibrotic area. Many alveoli were hypoplastic and lined with cuboidal epithelium.

Case 5: J.L. (female) ae. 9 1/2 yrs., 1949. This child has never been really well, but worse since pneumonia at two years. She has had persistent cough without much sputum since. During this period she had five bouts of fever, 103 to 105 degrees F., and pneumonia again nine months ago. There has been general deterioration in health since that time.

Operation: The whole left pleural cavity was obliterated by adhesions. These were more marked along the main fissure between the upper and lower lobe. On palpation, the lower lobe was firm and rubbery, with slight crepitations. At the base of the lung, on the lateral portion, there was a large artery coming from below the diaphragm but it was impossible to determine its origin.

The specimen showed an enlarged and firm lower lobe and lingula. The aberrant artery entered 3 cm. from the posterior tip. The cut surface showed numerous cavities filled with pus.

Microscopically all the bronchi and bronchiolae were dilated and their walls infiltrated with inflammatory cells. The muscle tissue, glands, and elastic tissue were replaced by fibrous tissue. The abnormal vessel was a systemic artery.

Case 6: B.N. (male) ae. 6, 1952. Child was well until five years of age he had pneumonia, following which his general health deteriorated. He had many bouts of fever. Cough developed, without sputum and became progressively worse. Both his radiograph and bronchogram showed the changes characteristic of cystic fibrosis. Lobectomy was done in 1952.

At operation many cysts were seen over the left lower lobe. There was a large artery in the pulmonary ligament running from the aorta below the diaphragm.

Pathology: The left lower lobe measured 14-9 x 3 cm. The dorsal segment was the only portion that inflated well. The large cysts over the base could not be filled by inflation. On the cut surface the dorsal segment appeared normal. The
posterior basic segment was composed of many multiloculated, thin wall cysts containing clear fluid but no air. There was no communication between these cysts. There was some alveolar tissue between them. The anterior and middle segments were compressed to about 1.5 cm. in thickness. The dorsal bronchus was normal. The middle and anterior bronchi were displaced superiorly and fanned out over the cystic regions. The bronchus to the posterior basal segment was defective, being represented only by a few short bronchioles descending just under the pleura on the mediastinal surface.

The pulmonary artery measured 0.3 cm. in diameter while the aberrant vessel was 0.7 cm. across. It entered the lung at its posterior surface about 1 cm. above the inferior border at the end of the pulmonary ligament. It penetrated the lung about 0.8 cm. and broke into four branches which arborized into all of the cystic portion (see drawing). At the bifurcation, there was a small linear area, yellow and slightly thickened. Two of the branches accompanied a bronchus that did not communicate with the exterior. The vessels were large for only 2 cm. before they arborized. The isolated bronchus was probably a sequestrated posterior basal bronchus. It was about 0.4 cm. wide. It branched into bronchioles when the artery divided. Some of the branches entered into small centrally placed cysts by small atria but never connected with any normal airway.

Microscopic examination showed the walls of the cysts to be composed of columnar ciliated epithelium lying on a smooth muscle wall. In places there was continuity between the cysts and normal alveoli and ducts. The wall of the blind bronchus was composed of ciliated columnar epithelium, smooth muscle, cartilage, collagen, glands, vessels and nerves. The intervening parenchyma appeared normal. The walls of the main bronchi were heavily infiltrated with chronic inflammatory cells. The cyst wall showed little inflammation.

The walls of the aberrant artery were composed of alternating layers of elastic tissue and smooth muscle fibre. There were approximately 14 wide elastic lamina as well as numerous finer elastic fibres.

**SUMMARY**

1) Six further cases of a congenital lung anomaly of both teratological and clinical interest are described.

2) The anomaly consists of a portion of displaced lung tissue in the chest cavity with hypoplastic alveoli and dilated bronchi which give the lung its honeycombed or cystic appearance. This displaced lung is the recipient of a systemic vascular supply usually from the aorta. It has come to be called intralobar sequestration since Pryce's paper was published.

3) It is probably due to the amputation of a primordial lung bud by an aberrant artery, occurring in the 4 to 14 mm. embryo.

4) The history, physical findings, and radiographic changes which make its diagnosis easy, are discussed. The clinical importance of its correct diagnosis is indicated.

5) Three cases are mentioned in which the whole lung was involved in the process and the question raised if such a result could not ensue rather than agenesis when earlier more massive portions of the bronchial tree are amputated.

**RESUMEN**

1) Se describen seis casos más de una anomalía congénita pulmonar de interés tanto clínico como teratológico.

2) La anomalía consiste en una porción de tejido pulmonar desplazada
en la cavidad torácica con alveolos hipoplásicos y bronquios dilatados que dieron al pulmón apariencia de panal o cística. Este pulmón displásico tenía un sistema vascular emergiendo generalmente de la sorta. Se ha llamado secuestración intralobar desde que se publicó el trabajo de Pryce.

3) Probablemente se debe a la amputación de una lema primitiva del pulmón por una arteria aberrante ocurriendo en el embrión de 4 a 14 mm.

4) La historia, los hallazgos físicos y los cambios radiográficos que hacen fácil su diagnóstico, se discuten. La importancia clínica de su correcto diagnóstico se indica.

5) Se mencionan tres casos en los que todo el pulmón estaba comprometido por el proceso y suscitaron el problema de si tal resultado podrían ocurrir mas que la agenesia cuando mas tempramente porciones mas voluminosas del arbol bronquial son amputadas.

RESUME

1) L'auteur décrit six cas d'anomalie congénitale du poumon, qui présen-
tent un intéressent clinique et tératologique.

2) L'anomalie consiste en une portion de tissu pulmonaire déplacé dans
la cavité thoracique avec alvéoles hypoplastiques et bronches dilatées, qui
donne au poumon un aspect kystique ou en "rayon de miel." Ce poumon
anormal reçoit un système vasculaire de suppléance, provenant générale-
ment de l'artère. Il est passé dans les habitudes de l'appeler "séquestration
intralobaire" depuis la publication de l'article de Pryce.

3) L'anomalie est due probablement à la séparation d'un bourgeois primiti-
tif par une artère aberrante, survenant chez l'embryon de 4 à 14 mm.

4) L'auteur discute l'histoire du malade, les éléments de l'examen physi-
que et des altérations radiologiques qui en facilitent le diagnostic, et men-
tionne l'importance clinique d'un diagnostic correct.

5) L'auteur rapporte trois cas dans lesquels la totalité du poumon était
intéressée par ce processus. Il pose la question de savoir si un tel résultat
ne pourrait pas être plutôt en cause que l'agénésie lorsque des portions
plus importantes de l'arbre bronchique sont amputées.

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