Intralobar Sequestration of the Lingula Pulmonalis

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Intralobular sequestration is a congenital cystic malformation of the lungs associated with an aberrant systemic artery. The term "intralobar" distinguishes the condition from the completely sequestered, vascularity independent, accessory lungs that are found attached to extrapulmonary structures. As the affected area is almost invariably situated in the posterior basal segment of the lower lobe, this anatomic relationship is often included in the definition of the anomaly.

Since Pryce1 advanced his theory on the causal role of the systemic arteries, a number of critical reports have appeared concerning the coincidence of these vessels with the bronchial dislocation and the embryologic relationship between intralobar sequestration and accessory lungs.

The present case, involving the lingula pulmonalis, may be of interest in respect to the unusual site of occurrence and the dual source of the arterial supply, as well as its apparent intermediate position between the intralobar and extralobar conditions.

The cystic mass was noted in the cadaver of a white woman of 62 years, with a history of bronchiectasis. It corresponded in shape, size and position to a normal lingula, but a constriction of the pleura marked its separation from the remainder of the inferior division. In the concavity of the cardiac notch a smaller cystic area, derived from the superior lingular segment, was delineated in the same manner (Fig. 1).

Dissection revealed that the pleural constrictions were caused by the attachment of the pleura to a fibrous septum similar in structure and the ease of its separation to the normal intersegmental planes of the lung. Only branches of pulmonary veins, and small arteries derived from a vessel coursing in the frenum of the upper lobe, entered the lingula just beneath the pleura (Fig. 2). As the mediastinal surface of both

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Figure 1: Photograph of the costal surface of the inferior division of the left upper lobe. The delineation of the sequestered lingula is obvious (A), and the smaller cystic area appears as an elevated mass in the cardiac notch (B). A cyst may be discerned in the inferior aspect of the lingula (C). The strip of adherent pericardium includes the tortuous pericardiophrenic artery (D).
affected areas was adherent to the pericardium, the predominant arterial supply to the lingula and the exclusive supply to the smaller mass was derived from the pericardiophrenic artery.

Further dissection showed the branches of both segmental bronchi to be markedly bronchiecstatic while the superior division was normal in this respect. The bronchial ramifications of the left upper lobe was not of the prevailing pattern but corresponded to a trifurcate form which Boyden and Hartmann described under the designation of “split” anterior bronchus. With the prevalent bifurcate pattern, B1 is the only bronchus supplying the anterior segment, whereas, in the trifurcate patterns B2 becomes subsegmental and an additional bronchus, BX, is interposed between B1 and B2. The extent to which a branch BX appropriates the anterior segment depends on the degree of “descent” of the origin of B1. The present case represents the extreme in this respect and conforms with 7 per cent of the series observed by Boyden and Hartmann in that B1 arises in common with the bronchi of the inferior division.

Histologic sections of the sequestered tissue exposed several epithelium lined cysts, the two largest being pea-sized and filled with a brown gelatinous substance that is frequently found in these structures.

**DISCUSSION**

Following the report by Pryce, the etiology of intralobar sequestration has become a subject of controversy. He claimed that the bronchi were dislocated by the traction of a persistent branch of the embryonic splanchnic plexus. There is no doubt that the aortic derivative to sequestered areas is a remnant of the original arterial network of the foregut and its pulmonary outgrowth, but, as Boyden has well explained, the probability that vessels which are essentially capillary in nature could segment the developing bronchi by traction is inconsistent with the existing knowledge of developmental processes.

A second theory, advanced by Smith, stipulates that the cystic condition is a post-natal development resulting from the subjection of the area served by the aberrant artery to the increased force of the systemic blood pressure.

![Figure 2: Drawing of bronchial pattern of the left upper lobe including the arteries supplying the sequestered tissue.](image-url)
In a large series of human embryos, Boyden found lung cysts developing spontaneously in a 31 mm. specimen, and a systemic artery to the posterior basal segment in another 41 mm. fetus. Because of their independent occurrence and a warranted critical view of the aforementioned theories, he believes the association of the artery and sequestration to be coincidental. However, it is difficult to conceive that two relatively infrequent aberrations would usually occur together without some type of interdependence.

Though Pryce and his associates thought that arterial traction was responsible for both intra- and extralobar sequestration, prevalent opinion holds that accessory lungs and intrapulmonary malformations are embryologically unrelated. Recent writings on this subject maintain that extrapulmonary lung tissue arises from accessory sites of proliferation in the primitive foregut (accessory theory) rather than by segmentation of the developing lung (fraction theory). Smith supports this concept by elaborating several definitive anatomic discrepancies between extralobar sequestration and accessory lung, but a critical analysis indicates that these differences are not incompatible with the fraction theory.

The only valid reason for assuming an extrapulmonary origin of accessory lungs is that a small number have been found attached to the gut. The fact that the normal lung primordium is an outgrowth of the foregut certainly attests to the potentiality of this region to differentiate into respiratory epithelium, and it is probable that masses found adherent to the lower esophagus or stomach may have arisen from this source. However, to presume this to be the origin of extralobar growth which are not found contiguous with the gut, but in proximity to the lung, requires a sequestering agency more remarkable than that needed to separate it from the highly plastic embryonic pulmonary tissue. In addition, the problem of the transfer of the blood supply from splanchnic to parietal vessels would be identical regardless of the derivation.

Diverse anatomic relationships and the concurrence of multiple anomalies are other features that Smith uses to reinforce the concept of separate etiologies. The situation of intralobar sequestration is almost always in the posterior basal segment and found on the left side in only 60 per cent of the cases. Lower accessory lung, on the other hand, is 90 per cent sinistral and is often found with other malformations.

The constant localization of intralobar sequestration is due to the dependency on an access to the aorta. The posterior basal segment of either lung, by virtue of its nearly exclusive relationship with the pulmonary ligament, is almost the only area of the lung where lesions deprived of pulmonary arteries could survive. The one exception is the inferior segment of the lingular division which is attached to the aorta by way of the frenum.

The congenital abnormalities of the diaphragm or pericardium that are often found with accessory lung, but seldom with intralobar lesions, would only require a temporal difference in the origin of the conditions. Smith acknowledged this probability stating: "In accessory lung also an earlier and less tangible origin seems probable in most cases. Obviously if the development is traced sufficiently far back to the somite stage of the fetus, all lung tissue regardless of its situation and connection at birth has a common origin, and whether it arises according to the accessory theory or fraction theory becomes of less importance." It is not the contention here that the probable common origin rests simply in germ layer homology, but that both intra- and extralobar sequestration are the result of a dislocation of the highly plastic developing bronchi, and the factors responsible for accessory lung may be more tangible than those producing intralobar sequestration.

As reported in a previous paper, an attempt was made to determine possible causes of lung agenesis, aplasia, hypoplasia
and accessory lung by investigating the development of the bronchial diverticula consistently found in the lung of the shrew (Blarina brevicauda). The analysis of a series of embryos, ranging from 4 mm. to the 17 mm. fetus, showed that these structures resulted from an interaction between an unusual extent of lung growth and body cavity formation. Because of an excessive degree of dextrocardia in this species, the right pleuroperitoneal opening was larger and remained patent longer than in other mammals. This permitted the evolution of a lung pattern that produced a lobular extension of the right lower lobe. With the expansion of the liver and the consecutive closure of the pleuroperitoneal hiatus and the costophrenic sinus, the overextended lobule suffered compression.

The consequence of this situation illustrated the remarkable plasticity of developing lung tissue. Although the whole appendage showed the potential of becoming a functional lobule, the increasing spatial restriction obliterated all secondary branches leaving but a pair of extrapulmonary diverticula characteristic of pulmonary aplasia (Fig. 3). It is noteworthy that these structures in the adult seldom gave any indication of the extent of differentiation they had achieved, and except for the diverticula the conventional contour of the lobe was restored.

When it is considered that the left pleuroperitoneal hiatus in the human remains patent longer than the right, the unilateral predisposition of accessory lung and phrenic defect is understandable. Failure of the lung to remain clear of the hiatus during its closure would be detrimental to both the caudal segments of the lung and the diaphragm, and the intimate contact of the lung with parietal structures permits the development of an accessory blood supply.

As a similar relationship exists between the growing lung and the ventromedial (pericardial) wall of the early pleural sac, a plausible embryology of the previously described case may be inferred. During the fifth week of development, the fusion of

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**FIGURE 3:** The development of the bronchial diverticula in the shrew. A, Dorsal aspect of the bronchial development and lobulation of both lungs in 4 mm. embryo. B, Lungs of 5 mm. embryo showing the diverticulate bronchi as they grow through the pleuroperitoneal hiatus. C, Bronchial arborization in right posterior lobe at 9 mm. D, Right posterior lobe at 10 mm. showing the repression of the secondary branches and mesenchyme of the diverticula. E, Right posterior lobe at 17 mm. showing vesicular remnants of secondary branches. F, Typical adult diverticula represented as smooth tubular structures.
the pleuropulmonary continuity and the pulmonary system, a transfer of drainage to the superior phrenic vein that courses with the pericardiophrenic artery would have produced a true accessory lung.

SUMMARY

Intralobar sequestration of the lingula pulmonalis was noted in a white woman cadaver of 62 years. As the affected tissue was adherent to the pericardium, it received an extrapulmonary arterial supply from both the pericardiophrenic artery and a systemic vessel derived from the aorta. This case was of particular interest as an apparent intermediate condition between intralobar sequestration and accessory lung and because of the unusual situation of the anomaly.

RESUMEN

Se descubrió el secuestro intrapulmonar en la lingula en el cadáver de una mujer blanca de 62 años. Como el tejido afectado estaba adherido al pericardio, recibía irrigación arterial extrapulmonar tanto de la arteria pericárdica como de un vaso emanado de la aorta. Éste caso es de interés especial como una condición aparentemente intermedia entre la sequestración intraloblar y el pulmón accesorio a causa de la situación inusitada de la anomalía.

RéSUMÉ

L'auteur a découvert une séquestation intralobaire de la lingula pulmonaire à l'autopsie d'un homme de race blanche de 62 ans. Le par enchyme pathologique était adhérent au péri cardie et recevait ainsi sa vascularisation extrapulmonaire, à la fois de l'artère pé ricardiophrénique et d'un vaisseau systémique issu de l'aorte. Ce cas présentait un intérêt particulier car d'une part il représentait un état intermédiaire entre la séquestration intralobaire et le pou mon accessoire et d'autre part à cause du siège inhabituel de l'anomalie.

ZUSAMMENFASSUNG

REGIONAL PULMONARY BLOOD FLOW

Carbon dioxide labeled with oxygen-15 has been used by Dollery and co-workers to study the regional blood flow in the lungs of patients with pulmonary stenosis. The upper and lower zone flows were normal in patients with isolated pulmonary stenosis, but both were low in patients with the tetralogy of Fallot. A comparison of the two upper zones showed a significantly higher blood flow through the left upper zone in patients with both isolated pulmonary stenosis and the tetralogy of Fallot. A significantly higher flow was also found through the left upper zone than the right normal subjects. By contrast, patients with atrial septal defect had a higher flow through the right upper zone than the left. Neither difference was as large as in the patients with right ventricular outflow tract obstruction.

Patients with ventricular septal defect and those with patent ductus arteriosus had almost identical flow through the two upper zones.


TOTAL CORRECTION OF TETRALOGY

An analysis of the postoperative course in 28 patients who had complete correction of tetralogy of Fallot is presented. Pertinent preoperative findings are summarized and the operative procedure is briefly described. During or following surgery, seven patients had a syndrome of low cardiac output which was lethal in four. The operative mortality rate was 14 per cent. One patient died during re-operation for persistence of the ventricular septal defect; the overall mortality rate was 18 per cent. Congestive heart failure developed in 42 per cent of surviving patients. This was transient in some and had a more protracted course in others. All patients recovered and late improvement was impressive. The ultimate result from surgery was considered excellent in the majority of cases.

It is concluded that total correction is feasible in most patients with tetralogy of Fallot, with an acceptable surgical mortality.