Extralobar Pulmonary Sequestration associated with Congenital Diaphragmatic Hernia

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INTRODUCTION

Sequestration of the lung has been traditionally defined as a mass of lung tissue that receives a systemic arterial supply and has no communication with tracheobronchial tree. This sequestrated, non-functioning lung tissue can occur either within the definitive lung without a separate pleural covering or as a mass separated from the lung. The former is called an intralobar sequestration and the latter, an extralobar sequestration (Gerle et al., 1968; Kissane, 1971; Iwai et al., 1973; Spencer, 1978).

The extralobar type of pulmonary sequestration has often been called an "accessory lung" (Müller, 1928; Gruenfeld and Gray, 1941; Spencer, 1978), because it has a distinct pleural investment, which maintains complete anatomic and physiologic separation from the adjacent normal lung. The left lower lobe is involved in about two thirds of the cases (Turk et al., 1961) and is often associated with diaphragmatic hernia (St. Raymond, 1956; Carter, 1969).

Recently authors experienced a case of extralobar pulmonary sequestration which was incidentally found during a reparative surgery for left Bochdalek hernia in a newborn infant.

CASE REPORT

This 2 day old male infant (S.N.U.H. 1359698) was admitted to the hospital because of dyspnea. He was delivered at full term to a 26 year old para 2-0-0-2 woman after uneventful prenatal course, weighing 2800g at birth. Soon after the birth respiratory difficulty was noted. It became progressively worse and was accompanied with cyanosis.

Examination on admission revealed an irritable infant with scaphoid chest, moderate dyspnea and mild cyanosis. The temperature was 37.3°C, the pulse rate was 140/min, and the respiration rate was 96/min. Simple chest X-ray (Fig. 1 and 2) revealed mild hyperexpansion of the right lung and total obliteration of the left chest with intestinal gas shadow. Mediastinum was shifted to the right side and the heart was located in the left chest. Under an impression of diaphragmatic hernia emergency operation was performed.

At operation, the left chest cage was filled with transverse colon, a considerable portion of small intestine, fundus of stomach and a part of left kidney, which were herniated through a large Bochdalek foramen, with measurements of 3.5×3.0cm. The left lung was almost entirely collapsed and was pushed medially by the displaced intestinal loops. A mass was found at the dome of the diaphragmatic hernial sac and had no communication to bronchial or pulmonary arterial system of the collapsed left lung. It was covered by a thin translucent membrane and located between parietal pleura and peritoneum. The mass was round, dark red and meaty and measured 2×2×1cm. This mass was first
thought to be displaced accessory spleen. Frozen section was done from the mass, which turned out to be abnormal lung tissue.

After the surgery he was placed in Bourns respirator for 16 days because of respiratory failure and several episodes of cardiac arrest. The small, collapsed left lung was aerated and fully expanded by the 4th postoperative day. Twenty-four days after the surgery symptoms of respiratory difficulty was improved and he was discharged.

Pathology: The specimen (S-80-4193) consisted of pale reddish and unusually resilient soft tissue mass that was covered by a thin translucent membrane, measuring $4 \times 2 \times 1$ cm. No crepitancy was noted. After the fixation in 10% formalin for 24 hours, the main mass was flat-round and dark red with measurements of $2 \times 2 \times 1$ cm. Cut sections showed honeycomb appearance, with the largest cyst being 3 mm in diameter. No bronchial tree was seen on the cut surface. However, a single large artery, 1 mm in external diameter, entered into this lesion through a cord-like fibrous tissue attached to the main mass.

Microscopically, almost entire mass consists of irregularly dilated bronchiolar structures that are closely packed together only with a small amount of fibrous stromal tissue. Mature alveolar structures are not recognizable throughout the specimen. The bronchiole-like spaces are lined by pseudostratified columnar epithelium with prominent long cilia. The lumens are mostly empty, but amorphous watery material is present in some of these spaces. Focal papillary ingrowths as well as cystic dilation are also seen. The stroma is composed of loose fibrous tissue with minimal mononuclear cell infiltration. Striated muscle cells are scattered in clumps or individually lying free in the stroma. There are only two bronchi that contain hyaline cartilage ring. One is near the periphery of the mass and the other is seen, embedded in the fibrous tissue pedicle. These two bronchial elements were cut cross and are in close proximity. Hilus-like structure is present, where scattered lymph nodes, a few blood vessels, and peripheral nerve trunks are included. The artery is of elastic type.

**DISCUSSION**

Among many theories that have been raised for the explanation of pulmonary sequestration, the strongly favored one was postulated by Eppinger and Schauentein in 1902 and subsequently supported by others. According to this theory, an additional tracheobronchial bud develops from the embryonic foregut distal to the normal lung bud. This pluripotential tissue, with its own blood supply, migrates caudal with the normally developing lung, giving rise to the sequestration. Whether the malformation is intralobar or extralobar is probably determined by the time at which the accessory foregut outpouching occurs. Sequestration arising later from a more caudal portion of the now elongated foregut remains extrapulmonary and acquires a covering of parietal pleura, i.e., "extralobar".

Waddell (1949) proposed that the mesenchymal tissue investing the primitive bronchial tree was responsible for the development of the distal respiratory tissue (respiratory bronchioles and alveoli) of the normal lung. Summation of the accessory lung bud theory and Waddell's view might explain the wide spectrum of histologic differentiation of pulmonary sequestration on the basis of the extent of bronchoalveolar development. This view is strongly favored by Spencer (1978). Examples of sequestrated lung with well developed conducting passages with relative failure of the mesenchymal component
are grouped in the category of grade 1 of the series of Iwai et al. (1973). Here, the main component is cystic large bronchi with well developed cartilage, bronchial glands, smooth muscles, and tall ciliated epithelium. In other extreme of the spectrum, grade 4 of Iwai et al. series, bronchioloalveolar structures are well differentiated. Our case would belong to the category between above mentioned two extremes, because the great majority of the mass is terminal bronchioloes and yet true alveolar differentiation is not recognizable. These features are somewhat resembling so-called adenomatoid malformation of the lung (Chi and Ahn, 1979). In fact there might be causal relationship between these two entities.

Striated muscle fibers seen in this case is of special interest in the sense of its histogenesis. Striated muscle has been seen in other case of extralobar pulmonary sequestration (Aterman and Patel, 1970). We experienced an other case of massive striated muscle inclusion in the hypoplastic lung in a 20 week old fetus. The presence of skeletal muscle might be related to degree of maturation of the lung.

The absence of a patent foregut communication can be explained by involution of the communication during the development, possibly because it outgrows the blood supply (Flye and Izant, 1972; Spencer, 1978). Remnants of the connection can sometimes be found in a fibrous pedicle which accompanies the arterial blood supply to the anomalous tissue.

Approximately 90 per cent of the cases of extralobar sequestration are found in the left side of the chest (Smith, 1962; Askin, 1975). Their occurrence on the left side is probably related to the later closure of left diaphragm and the absence of a large underlying left lobe of the liver (Spencer, 1978). Carter (1969) reported 50 per cent incidence of anomalies associated with extralobar pulmonary sequestration, with the most common one being diaphragmatic hernia. Valle and White (1947) noted congenital diaphragmatic hernias on the left side in approximate 30 per cent of cases of extralobar sequestration. The frequent association of hernia with this condition has been interpreted as being due to mechanical interference by the accessory mass of lung with normal closure of the pleuropertitoneal canal (Berman et al., 1952). Pulmonary agenesis, evagination of the diaphragm, esophageal diverticulum, basilar artery aneurysm, horseshoe kidney, ectopic pancreatic tissue, congenital megacolon, and duplication of the colon are other reported anomalies associated with extralobar pulmonary sequestration (Berman et al., 1952; Carter, 1969; Flye and Izant, 1972). This frequent association of anomalies is contrast to the rarity of combined defects in intralobar type of pulmonary sequestration.

The arterial supply to extralobar sequestration is usually one or more arteries arising from the descending thoracic aorta, subclavian, intercostal or inferior diaphragmatic arteries, though it may derived from the pulmonary artery (Baar and d'Abreu, 1949). They are usually small and inconspicuous, and frequently enter the sequestated lobe through the ligamentum pulmonis (Spencer, 1978). In most of extralobar types of pulmonary sequestration the venous drainage is usually via azygos system, but pulmonary venous drainage is also recorded (Gerle et al., 1968). Since angiography has not been performed in our case, it is difficult to determine any specific arterial supply to the lesion. But this case appears to have a systemic arterial supply because the vessels found on the surface of the mass are not connected with pulmonary artery.

Differences between intralobar and extralobar types of pulmonary sequestration are well discussed by Smith (1962) and could be summa-
Table 1. Comparison of the features of intralobar and extralobar pulmonary sequestrations*

<table>
<thead>
<tr>
<th>Feature</th>
<th>Intralobar type</th>
<th>Extralobar type</th>
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<tbody>
<tr>
<td>Bronchopulmonary tissue</td>
<td>Confined to posterior basilar segments; no pleural separation</td>
<td>Found above or below diaphragm; separate pleural covering</td>
</tr>
<tr>
<td>Arterial supply</td>
<td>From aorta, above or below diaphragm; well developed, large artery</td>
<td>From pulmonary or systemic artery; usually small inconspicuous vessel</td>
</tr>
<tr>
<td>Venous drainage</td>
<td>Pulmonary veins</td>
<td>Azygos, hemiazygos or portal venous system</td>
</tr>
<tr>
<td>Side involved</td>
<td>60% on the left</td>
<td>&gt; 90% on the left</td>
</tr>
<tr>
<td>Foregut communication</td>
<td>Very rare</td>
<td>More common</td>
</tr>
<tr>
<td>Other anomalies</td>
<td>Rare</td>
<td>Frequent and severe</td>
</tr>
<tr>
<td>Found on neonates</td>
<td>Never</td>
<td>Often</td>
</tr>
</tbody>
</table>

*Proposed by Smith (1962)

rized as Table 1. However, despite these differential features, the increasing number of cases that have now been reported (Park, 1962; Gerle et al., 1968; Iwai et al., 1973) which have shown features of both intralobar and extralobar varieties makes it somewhat difficult to distinguish clearly these two types by means of Smith’s criteria alone. Although intralobar sequestration is almost invariably associated with recurrent infection, extralobar variety is much less likely to become infected, except when there is a gastrointestinal communication. Thus, this lesion is usually managed by total excision of the mass, in contrast to lobectomy frequently applied to intralobar sequestration. The surgeon should realize the fact that the anomalous origin of the arterial supply to the sequestrated lung can cause a lethal hemorrhage due to transection of this artery during lung resection (Harris and Lewis, 1940).

SUMMARY

Authors reported a case of extralobar pulmonary sequestration at the site of congenital diaphragmatic hernia in a 2 day old infant. The accessory lung was found incidentally during a reparative surgery for a left Bochdalek hernia. This mass, 2cm in size, was far apart from the poorly aerated left lung, and was embedded within the membranous tissue of the diaphragmatic hernial sac.

Neither bronchial nor pulmonary arterial connection was seen. The hernia was successfully repaired and the accessory lung mass was removed.

Microscopically the removed mass consisted entirely of terminal bronchiolar mass with focal cystic dilatation and abundant stroma which contained scattered striated muscle fibers. No evidence of alveolar differentiation was recognized.

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同時に Extralobar pulmonary sequestration 1例を発見し、後に
2例を発見し、我々の観察した例を報告し、文献を参照する。

患児は出生直後に呼吸器に不調を認め、X-rayを施行したが、特異
な所見は認められなかった。左側胸腔内に肺組織の存在が示唆され
たため、剖検を施行した。剖検所見では、左側胸膜に沿って硬固な
肺組織があり、胸部の形態に影響を及ぼしていた。剖検所見の根
拠により、Extralobar pulmonary sequestration の存在が示唆されました。

REFERENCES

St. Raymond, A.H., Jr., Haréy, J.D., and Robbins, S.G.: Lower accessory lung communicating with esophagus and associated with congenital diaphrag-
- Chi, J.G. et al.: Extralobar pulmonary sequestration —

[Diagram: Illustration of a human silhouette with labeled parts: Heart, LL, RL, S, S, T, C, PS.]

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[Images: Close-up views of medical samples labeled A, B, and a section labeled B.]

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- Hernial sac & opening
- Kidney.

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**LEGENDS FOR FIGURES**

Fig. 1. Rentgenogram of the body. Air-filled small and large bowel loops are filling the left chest cavity. Mediastinal structures as well as the left lung is markedly shifted to the contralateral side.

Fig. 2. Schematic drawing of diaphragmatic hernia, showing the relationship of the normal and abnormal lung tissues. PS: sequestrated lung tissue, LL: left lung, RL: right lung, SI: small intestine, TC: transverse colon, S: stomach.

Fig. 3. Gross specimen of the sequestrated lung, showing external (A) and cut (B) sections. Note honey-comb appearance with focal cystic change on cut section.

Fig. 4. Low power microscopy of the specimen, revealing the relationship of parenchymal portion and pedicle of the accessory lung. L: lymph node, V: vessel, P: parenchymal portion, B: Bronchus. H&E ×3

Fig. 5. Medium power photomicrograph of the parenchymal portion (P in Fig. 4). Bronchiole-like structures are closely packed. A small amount of stromal tissue is seen between these structures. H&E ×40

Fig. 6. Bronchiole-like structure is lined by pseudostratified columnar epithelium with prominent cilia (A). Also note the striated muscle fibers in the stroma (B). H&E ×400