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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 pulmonar ysequestration 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 4×2×1cm. 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 2×2×1cm. Cut sections showed honeycomb appearance, with the largest cyst being 3mm in diameter. No bronchial tree was seen on the cut surface. However, a single large artery, 1mm 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 bronchioles 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 occurence 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 pleuroperitoneal canal (Berman et al., 1952). Pulmonary agenesis, evernation 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 sequestrated 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*

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

*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 membraneous 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 structures with focal cystic dilatation and abundant stroma which contained scattered striated muscle fibers. No evidence of alveolar differentiation was recognized.

≫國文抄錄≪

先天性 橫隔膜 脫腸에 同伴된 副肺症
(Extralobar Pulmonary Sequestration)

서울大學校 醫科大學 病理學敎室
池堤根·李正彬·金明淑
서울大學校 醫科大學 外科學(小兒外科)敎室
朴 貴 媛·金 宇 基

著者들은 Bochdalek氏型 先天性 左側 橫隔膜 脫腸에

同伴된 Extralobar pulmonary sequestration 1例를 생후 2日 된 男兒에서 觀察하고 이를 記述하였다.

患兒는 出生直後부터 呼吸이 순조롭지 못해 X-ray를 촬영하였던 바 左側 橫隔膜脫腸이란 진단을 받고 復元手術을 시행하였다. 左側 胸膜腔은 완전히 小腸, 大腸 胃底部 및 左側 腎 一部로 차 있었고 左側肺의 通氣現象은 거의 認知할 수 없었으며 縱隔部와 함께 右側으로 밀려 있었다. 手術中 우연히 脫腸膜 中央部에서 2 cm정도 크기의 원형의 암적색 組織을 發見하였다. 이 組織은 側壁 胸膜과 腹膜으로 싸여 있었으며 氣管支나 肺動脈과의 연결은 觀察할 수 없었다.

顯微鏡的으로 이 組織은 部分的으로 囊性으로 팽창된 最終 細氣管支라고 생각되는 構造와 약간의 間質로 形成되어 있었으며, 肺組織邊緣部에서 2개의 氣管支가 觀察될 뿐이고 肺胞의 發育이 전혀 되지 않아 그 組織像은 先天性 腺腫樣 肺畸形과 유사했다. 間質에는 橫紋筋 細胞 및 그 纖維가 덩어리지서 혹은 개개로 散在해 있었다.

REFERENCES

Askin, F.B.: *Congenital anomalies of lungs in Kissane, J.M., Pathology of infancy and childhood. 2nd ed. St. Louis: C.V. Mosby. 1975. pp. 473-476.*

Aterman, K. and Patel, S.: *Striated muscle in the lung. Amer. J. Anat.*, **128**:*341, 1970.*

Baar, H.S. and d'Abreu, A.L.: *Duplications of the foregut. Brit. J. Surg.*, **37**:*220, 1949.*

Berman, J.K., Test, P.S., and McArt, B.A.: *Congenital esophagobronchial fistula in adult. J. Thorac. Surg.*, **24**:*493, 1952.*

Boyden, E.A.: *Development of the lung. in Kelky's practice of pediatrics. Vol. IV, Part two. Chapt. 64, Harper and Row. 1975, p. 1.*

Boyden, E.A.: *Segmental anatomy of the lungs; A study of the patterns of the segmental bronchi and related pulmonary vessels. New York; McGrow-Hill, 1955.*

Carter, R.: *Pulmonary sequestration; collective review. Ann. Thorac. Surg.*, **7**:*68, 1969.*

Chi, J.G. and Ahn, Y.K.: *Congenital adenomatoid malformation of the lung (An autopsy case report). Seoul J. Med.*, **20**:*55, 1979.*

Eppinger, H. and Schauenstein, W.: *Krankheiten der Lungen. A. Angeborene Krankheiten. Ergebn. allg. Path. Anat.*, **8**:*267, 1902.*

Flye, M.W. and Izant, J.R.J.: *Extralobar pulmonary sequestration with esophageal communication and complete duplication of the colon. Surgery*, **71**:*741, 1972.*

Gerle, R.D., Jaretzki, A. III, Ashley, C.A., and Berne, A.S.: *Congenital bronchopulmonary-foregut malformation; Pulmonary sequestration communicating with the gastrointestinal tract. New Engl. J. Med.*, **278**:*1413, 1968.*

Gruenfeld, G.E. and Gray, S.H.: *Malformation of the lung. Arch. Path.*, **31**:*392, 1941.*

Harris, H.A. and Lewis, J.: *Anomalies of the lungs with special reference to the danger of abnormal vessels in lobectomy. J. Thorac. Surg.*, **9**:*666, 1940.*

Iwai, K., Shindo, G., Hajikano, M., Tajima, H., Morimoto, M., Kosuda, T., and Yoneuda, R.: *Intralobar pulmonary sequestration with special reference to developmental pathology. Amer. Rev. Resp. Dis.*, **107**:*911, 1973.*

Klein, Z.: *An accessory lobe of lung in a newborn. Pediatrics*, **45**:*118, 1970.*

Müller, H.C.: *Handbuch der speziellen pathologischen Anatomie und Histologie. Edited by F. Henke and O. Lubarsch. Vol. 3, Part 1, Berlin; Springer, 1928. pp. 531-598. Cited by Gerle et al.*

Park, W.W.: *Intralobar sequestration of the lingular pulmonalis. Dis. Chest*, **41**:*378, 1962.*

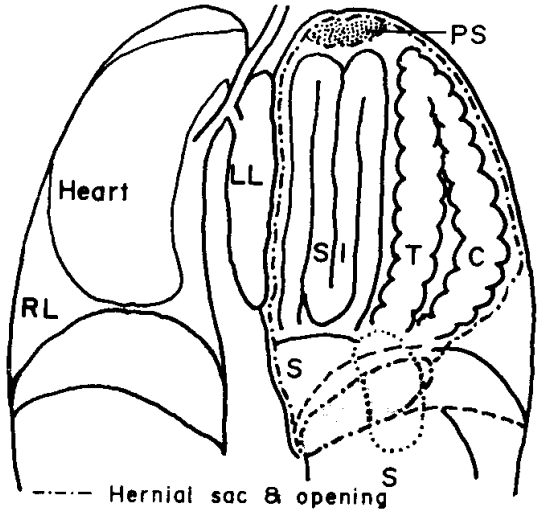
Pryce, D.M.: *Lower accessory pulmonary artery with intralobar sequestration of the lung; a report of seven cases. J. Path. Bact.*, **58**:*475, 1946.*

Pryce, D.M., Sellors, T.H., and Blair, L.G.: *Intralobar sequestration of the lung associated with an abnormal pulmonary artery. Brit. J. Lung*, **35**:*18, 1947.*

Smith, R.A.: *Some controversial aspects of intralobar sequestration of the lung. Surg. Gynecol. Obstet.*, **94**:*57, 1962.*

Spencer, H.: *Pathology of the lung. 3rd Ed. Oxford: Pergamon Press, 1978. pp. 75-87.*

St. Raymond, A.H., Jr., Hardy, J.D., and Robbins, S.G.: *Lower accessory lung communicating with esophagus and associated with congenital diaphra-*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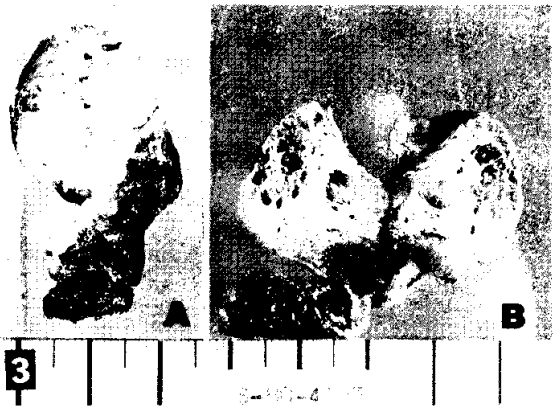
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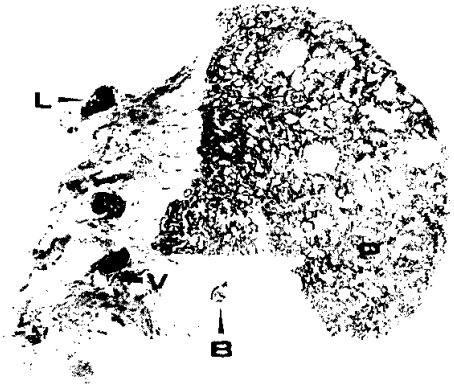
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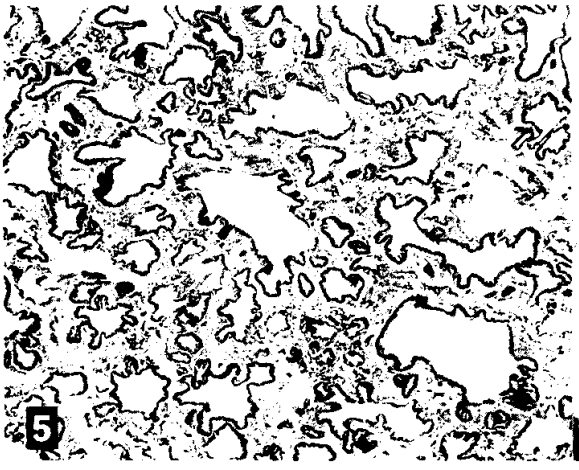

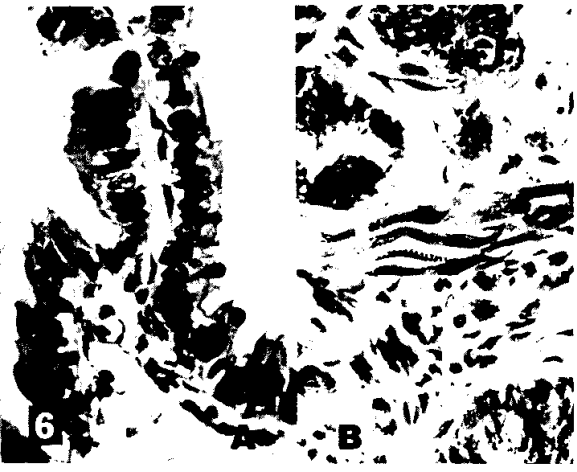
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gmatic hernia. J. Thorac. Surg., 31:*354, 1956.*

Turk, L.N. and Lindskog, G.E.: *The importance of angiographic diagnosis in intralobar pulmonary sequestration. J. Thorac. Cardiovasc. Surg.*, 41:*299, 1961.*

Valle, A.R. and White, M.L., Jr.: *Subdiaphragmatic dberrant pulmonary tissue; case report. Dis. Chest*, 13:*63, 1947.*

Waddell, W.R.: *Arch. Path.*, 47:*227, 1949. cited by Spencer.*

Warkany, J.: *Congenital malformation. Notes and comments. Chicago: Year Book Med. Publ. Inc. 1971. pp. 604-617.*

Yang, K.M., Ahn, G.H., and Kim, Y.I.: *Intralobar pulmonary sequestration; a case report. Korean J. Cardiovasc. Thorac. Surg.*, 4:*55, 1971.*

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