Extralobar pulmonary sequestration. An autopsy case

Satoshi Furukawa*, Akari Takaya, Tokiko Nakagawa, Ikuo Sakaguchi, Katsuji Nishi

Abstract: We encountered at forensic autopsy a rare anomalous case of extralobar pulmonary sequestration in the thorax. The anomalous tissue had its own visceral pleura and was located under the lower lobe of the lung in the left thoracic cavity of a 34-year-old male victim. It had no vascular or airway connection to the normal lung. However, it had vascular branches from the thoracic aorta and hemiazygos vein. Its long diameter was 8.5 cm; its short diameter, 8.0 cm; and its height, 3.0 cm. It weighed 75.0 g. Histological examination detected blood vessels, nerve cells, lymph nodes, lumens of various shapes, and alveoli in the tissue. This showed that the mass had the same structure as lung tissue. Based on these findings, the anomalous tissue was clearly an extralobar pulmonary sequestration.

Key words: Extralobar pulmonary sequestration, Forensic autopsy, Histological findings

According Gezer et al. [1] and Carrasco et al. [2], aberrant blood supply to the lung was already first reported by Huber in 1777. Rokitansky [3] and Rektorzik [4] in their independent studies from 1861 used the term “accessory pulmonary lobe” to describe extralobar pulmonary sequestration. Pryce [5] in 1946 renamed the phenomenon as “pulmonary sequestration”. Pulmonary sequestration is a rare congenital malformation with an estimated incidence of 0.15% to 6.45% [6]. Pulmonary sequestration is divided into two classes: intralobar sequestration and extralobar sequestration. An intralobar sequestration (which makes up 80% of cases) is contained within the normal visceral pleura and is surrounded by normal lung tissue. An extralobar sequestration (which makes up 20% of cases) is pulmonary tissue that is not connected to the tracheobronchial tree and has a connection to the systemic arterial blood supply [7].

In this report, we describe an autopsy case in which an anomalous soft mass was present in the left thoracic cavity of a 34-year-old Japanese male. Vessels connected the soft mass to the aorta and vein, but the mass was not connected to the bronchus.

Case report

After a storm, a broken sport fishing boat was found on Lake Biwa, which is the largest freshwater lake in Japan. A police search found two middle-aged men on the bottom of the lake. Autopsies were performed the next day.

Autopsy findings

The body of a 34-year-old man was slightly corpulent with no wounds. Both lungs were expanded due to inhaling fresh water. The bronchus contained a large volume of reddish fluid with foam. An anomalous soft tissue mass with a pyramidal form was present in the left thoracic cavity (Fig. 1). The pyramidal mass, having the dimensions of 8.5 cm × 8.0 cm × 3.0 cm, was found in the

*Corresponding author, Department of Legal Medicine, Shiga University of Medical Science, Setatsukinowa, Otsu City, Shiga 520-2192, Japan, Tel & Fax: +81-77-548-2202, E-mail: 31041220@belle.shiga-med.ac.jp
hemoglobin-colored fluid in the left thorax, between the lower lobe of the lung and diaphragm (Fig. 2. and 3). It was separate from the lung and resembled a partially expanded, immature lung with a smooth to wrinkled pleura. The mass consisted of pulmonary parenchyma and was separated from the rest of the lung by its own pleural envelope. It had vascular branches and was linked only by branches from the thoracic aorta and the hemiazygos vein (Fig. 4).

Other organs, including the brain, showed severe congestion. There were no other abnormalities present, except for the pyramidal mass in the thoracic cavity and a fist-sized cyst containing serous fluid on surface of the great omentum in the abdominal cavity. A diatom test performed after autopsy detected several species of diatoms in the lung, spleen, liver, kidneys and brain. The man’s death was the result of drowning.

Histological findings

Light microscopy observation with hematoxylin-eosin (HE) stain showed comparatively large arteries and several veins, nerve cells, and a near-circular lumen surrounded by hyaline cartilage and mucous glands in the hilar region (Fig. 5).

In the peripheral region there were many vessels, lymphocyte colonies (indicating chronic inflammation), and lumena with cuboidal epithelium.

There was a distinct hyaline cartilage surrounding the large lumen, which was lined by a columnar epithelium containing mucus that had been stained red-purple by periodic acid-Schiff (PAS) stain (Fig. 6). Mucus cells (which had been stained by PAS or Azan stain) were present in tracheal glands between the lumen and cartilage (Figs. 7 and 8). These lumena were lined by simple cuboidal epithelium containing goblet cells with mucus (stained purple with PAS stain) and ciliated cells (Fig. 9). Based on these histological findings, we denied hamartoma such as the congenital pulmonary airway malformation. The anomalous tissue clearly had the same structure as the lung and was an extralobar pulmonary sequestration.

Discussions

Savic et al. [6] in their review reported that extralobar sequestration was present in 133 (24.6%) cases of the 540 cases of pulmonary sequestration that were published between 1962 and 1975. In the majority of these cases, the extralobar sequestration was close to the diaphragm (77%) or in the left suprarenal area (90%). There are also reports of atypical localizations of extralobar pulmonary sequestration, such as apical and bilateral, mediastinal, intrapericardial, and abdominal pulmonary sequestrations and pulmonary sequestration of the hilum [6,8].

Extralobar pulmonary sequestrations are often associated with other congenital anomalies, such as diaphragmatic hernias and diaphragmatic defects, cardiopulmonary anomalies, and communication with the foregut. Dyspnea and cyanosis occur in 90% of extralobar pulmonary sequestration cases, especially in children (approximately 50% of whom suffer dyspnea and cyanosis) [6].

Extralobar sequestrations predominantly appear in infants and children, but rarely in adults [9]. Savic et al. [6] report that 10% of extralobar sequestrations are asymptomatic and are discovered incidentally. In their research of 540 published cases, they found that the oldest patient with an extralobar sequestration was an 81-year-old woman. The existence of sequestration in the present case was not discovered until autopsy and there were no associated congenital anomalies such as diaphragmatic defects, anomalous connections with the gastrointestinal system, or cardiovascular
defects. The only defect was a fist-sized cyst, which contained serous fluid, in the abdominal cavity. The etiology of intralobar sequestration remains controversial [5,10]. However, extralobar sequestrations are commonly considered to be of congenital origin. They are included in the class of broncho-pulmonary foregut malformations.

The respiratory system develops during the third week of embryonic stage from an outpouching of the laryngotracheal groove that forms on the ventral face of the foregut and later divides into the right and left bronchiæ, the bronchioles, and alveolar ducts [11]. The extralobar sequestration develops [1] either from a separate outpouching of the foregut or from embryonic lung-forming cells adhering to other organs, which then separates these cells from the remainder of the lung tissue because of the traction of differential growth. In either manner, a separate pleura would develop around the sequestration [12]. Extralobar sequestration almost exclusively presents before the age of 10; it most often presents with dyspnea and cyanosis within the first six months of life [13].

![Fig. 2. The pyramidal mass was found in the hemoglobin-colored fluid in the left thorax between the lower lobe of the lung and diaphragm](image)

![Fig. 3. The pyramidal mass, having the dimensions of 8.5 cm × 8.0 cm × 3.0 cm](image)

![Fig. 4. The vascular branches of the pyramidal mass were linked only by branches from the thoracic aorta and the hemiazygos vein](image)

![Fig. 5. Histological findings. (Hematoxylin-eosin stain)](image)
However, an estimated 10% of extralobar sequestrations are asymptomatic, and discovered by accident [6]. In the present case, the man had no symptoms before his death. To conclude, this case report, allowed us to analyse an extralobar pulmonary sequestration found at the autopsy and in the histological findings.

Acknowledgement. We thank Shiga University of Medical Science for supporting this project and permission to publish the article.

References