Intralobar Pulmonary Sequestration*

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Intralobar pulmonary sequestration is a developmental anomaly in which a portion of developing lung tissue receives an independent blood supply from the aorta, usually immediately above or below the diaphragm. This sequestrated portion of lung is non-functioning and develops cystic changes. Wall found three cases in 385 thoracotomies pre-operatively on clinical and roentgenographic findings.

Case Report

A. T., a 16 year-old Puerto Rican girl was admitted to the hospital with complaints of productive cough for three weeks, recent hemoptysis and weakness. She stated that she was very prone to upper respiratory infections and every winter she had had numerous "colds." Three weeks prior to admission she developed a cough which was non-productive initially. In the week prior to admission she had expectoration of bright red and dark blood, associated with right chest pain. Physical examination revealed slight dullness at the right axilla without other abnormal findings. Her temperature was 98°, pulse 70, blood pressure 120/70, and vital capacity 3,000 cc. The sputum was negative for acid fast bacilli or other pathogenic organisms on smear and culture. The erythrocyte count was 4,300,000; white blood cells 6,800; hemoglobin 14.0 gm.; Mantoux tuberculin test 1:1000 negative. Chest x-ray film showed a well-defined rounded mass about 6 cm. in diameter in the medial portion of the right lower lobe having a multi-cystic appearance. Bronchography showed good filling of all the bronchi of the right lower lobe with no filling of the cystic areas and no evidence of bronchiectasis. The branches of the posterior basilar bronchi of the lower lobe revealed accuation of their normal course due to pressure by the extra-bronchial mass. Laminograms demonstrated that the mass was sharply delineated, most clearly seen at 5 to 8 centimeters from the table-top with the patient supine. A gastrointestinal series was negative with no evidence of hernia. Angiocardiography failed to opacify the mass; an aberrant blood vessel could not be demonstrated. Bronchoscopy showed hyperemia of the right lower lobe bronchius with a moderate amount of white mucopurulent secretion in the basilar branches. The diagnosis of pulmonary sequestration was suggested on the basis of the history of repeated upper respiratory infections and recent hemoptysis in a young person with no evidence of tuberculosis and with radiographic evidence of a cystic mass in the right lower lobe which on bronchography did not communicate with the bronchial tree but displaced some of the basilar branches.

At operation, the diagnosis of sequestration of a portion of the right lower lobe was confirmed and the right lower lobe was resected. The sequestrated mass was in the posterior medial aspect of the right lower lobe with a large aberrant artery arising from the thoracic aorta just above the diaphragm and entering the sequestrated segment through the leaves of the inferior pulmonary ligament. Examination of the resected lower lobe revealed an irregular rectangular area, in its posterior inferior portion, which was firm and rubbery to palpation and appeared almost completely non-aerated. The regional bronchi were injected with methylene blue and none of the dye entered the mass. On section, this mass showed no recognizable normal lung parenchyma but contained cystic spaces of various sizes. The mass was not encapsulated but was sharply demarcated from surrounding well-aerated normal appearing lung parenchyma. The aberrant artery leading to this segment was injected with fuchsin and its small branches could be seen lying within the fibrous strands between the cystic areas.

DISCUSSION

Intralobar pulmonary sequestration represents a complete developmental separation of a portion of a lobe of a lung from its normal bronchial tree. In early fetal life the lungs extend so rapidly into the enlarging pleural cavities that in embryos of 30 mms. the definitive lobes and fissures are already established. Intralobar developmental anomalies occur before this stage is reached. There is considerable speculation as to whether the mode of origin of the anomalous artery is responsible for, or is secondary to, the pulmonary sequestration. Bemki observed that ectopic tissue may obtain vascular supply from the vicinity if malformation occurs early enough in the embryonic

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FIGURE 1: Poster-anterior chest film showing a localized zone of increased density at the right base, medially.

FIGURE 2: Poster-anterior tomogram, 7 cm. above the table-top, showing the posterior location of the mass which contains multiple luencies.

FIGURE 3: Right lateral bronchogram, showing normal filling of the bronchial tree and accretion of the posterior basal branches by the mass.
development. Wail et al., on the basis of this observation, favor the concept that sequestration results from "fractioning" of part of the development lung buds; subsequently the separated fraction seeks its own blood supply. Pryce considers the pulmonary anomaly secondary to the vascular anomaly which in turn results from the persistence of one or more of the connections between the dorsal aorta and the postbronchial pulmonary plexus. These persisting arterial connections cause separation of some of the developing lung buds by traction. Pryce is of this opinion mostly because cases have been encountered in which an anomalous artery from the aorta supplies one or both lower lobes, without any evidence of other abnormalities. Occasionally intralobar sequestration occurs in which the anomalous non-functioning portion of pulmonary tissue is found between the diaphragm and the lower lobe, enclosed in a pleural sheath of its own.

The sequestrated lung tissue is usually well demarcated from adjacent lung parenchyma. It may be composed chiefly of normally differentiated lung, a solid mass of branching bronchi, or a multicystic mass. The fact that such cysts sometimes contain air suggests the probability of communication with the bronchial tree. However, injection of dyes into the bronchi of resected specimens has not demonstrated any communication with the abnormal tissue.

Microscopically the picture is variable, with pseudostratified, cuboidal columnar epithelium, mesenchymal elements, irregular dilated cysts, and scattered cartilaginous and fibrous tissue. In about 85 per cent of all cases found in the literature the anomalous artery arises from the lower part of the thoracic aorta. In the remaining 15 per cent the anomalous vessel arises from the upper part of the abdominal aorta or from one of the branches of the celiac axis. This aberrant artery differs histologically from the pulmonary artery in that it has a thicker wall with more muscle fibers and larger elastic fibers. The venous drainage is to the normal pulmonary venous system.

The diagnosis of intralobar pulmonary sequestration should be considered whenever, on X-ray film examination, there is a sharply demarcated multi-cystic density in either lower lobe. A tapering projection may be directed toward the aorta. The distribution of the lesion is not segmental. Although the great majority of cases have been in the posterior medial portion of one of the lower lobes, cases have been reported in a somewhat different anatomic location, such as in the superior segment of the right lower lobe. The walls of the cysts are usually thin and are not always well visualized. Bronchography reveals displacement of some of the bronchial branches and failure of the opaque material to enter the region of the lesion even if cysts with air-fluid levels are present. In addition bronchography helps to rule out bronchiectasis and demonstrates the mass as extra-bronchial. The lesion can simulate a neoplasm. However, this anomaly usually is discovered before the age of 30 years. In the differential diagnosis, herniation through the Bochdalek foramen should also be considered and barium studies of the gastro-intestinal tract may be required to exclude this possibility.

REFERENCES