CASE REPORTS

PULMONARY SEQUESTRATION

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Pulmonary sequestration is a disturbance in embryonic development that results in a cystic mass of nonfunctioning lung tissue. The authors present a case of this disorder and review the pertinent literature.

Pulmonary sequestration is a disturbance in embryonic development resulting in a cystic mass of nonfunctioning lung tissue. The mass is supplied by an anomalous systemic artery and has its own bronchial tree, which usually does not communicate with the normal bronchial tree.[1-3] Its venous drainage is via the azygos system, pulmonary veins, and inferior vena cava.[2]

The term *sequestration,* derived from the Latin word sequestare, "to separate," was first used by Pryce in 1946.[1,3] In 1771 Huber gave the first description of an aberrant systemic artery supplying the lesion.[1] He noted an anomalous artery arising from the thoracic aorta supplying the right lower lobe of normal lung. In 1861, Rokitansky described a case of pulmonary sequestration. He defined the condition of extralobar sequestration, and believed the anomaly to be an accessory pulmonary lobe. Rokitansky's lobe is synonymous with extralobar sequestration. The full description of intralobar sequestration by Pryce in 1946 established pulmonary sequestration as a distinct clinical entity.

There are two types of pulmonary sequestration, intralobar and extralobar. Intralobar sequestration is confined within the lung, usually lying in the substance of the posterior basal segment of the lower lobe, although it may often extend into adjacent segments. It is intimately associated with adjacent lung and no pleural envelope exists.[2] This lesion is generally nonfunctioning. It is supplied by an anomalous systemic artery or arteries from the aorta; the venous drainage is usually via the pulmonary veins. The bronchial system often communicates with the tracheobronchial tree. When such an intercommunication exists, repeated infections of the involved area are not uncommon.[1]

Extralobar sequestration involves accessory lung tissue that is completely enclosed in its own pleural sheath. There is complete separation of the mass and the surrounding normal lung tissue. It can be found between the lower lobe and the diaphragm or in the upper portion of the abdomen.[2,3] This mass is also supplied by an anomalous systemic artery. Its venous drainage is directed into the azygos or hemi-azygos system. In this form, bronchial intercommunications are rare. In the extralobar type, as in the intralobar type, the left lower lobe is the site most commonly involved.[1,4,5] The approximate incidence of intralobar sequestration of the upper lobe is 14 percent.[6]

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ETIOLOGY

Many theories have been postulated with regard to the formation of pulmonary sequestration. It is

generally accepted that the aberrant lung tissue is the result of a congenital abnormality.[1] During normal embryonic development there is a separation of embryonic pulmonary tissue from the tracheobronchial tree, and this tissue degenerates into a cystic area alongside the normally developing lung.[1] Pryce postulated that traction from an anomalous branch of the aorta to a segment of the developing lung resulted in damage to the segment and separation from normal lung tissue.[3] Boyden advanced the theory of coincidental occurrence of lung cyst and anomalous systemic pulmonary arteries.[1,3] Other theories also exist but the theory first proposed by Eppinger and Schauenstein and expanded upon by Gerle and associates tends to explain most aspects of the etiology of both intralobar and extralobar sequestration. This theory states that an additional lung bud forms early in normal embryogenesis, developing distal to the site of normal lung bud formation. This accessory lung bud migrates with the normal embryonic lung. The vascular supply is derived from the splanchnic plexus and has many connections to the primitive ventral and dorsal aorta. These connections remain to form the anomalous systemic artery that will supply the aberrant lung tissue.[3]

The type of sequestration that occurs, intralobar or extralobar, depends at what embryologic point the accessory lung bud develops. If it develops early it becomes enveloped by normal lung tissue and intralobar sequestration occurs. However, if development occurs late in embryogenesis, there will be a separation of the sequestration from the normal lung tissue and an extralobar sequestration results.[3]

Pulmonary sequestration is a relatively uncommon developmental anomaly, but this disorder cannot be called rare. Sequestrations account for 1.1 to 1.8 percent of all pulmonary resections. Intralobar sequestration occurs six times more frequently than the extralobar variety. A possible explanation is that extralobar sequestration is often asymptomatic and therefore may go undetected.[1,3] Pulmonary sequestration occurs slightly more often in males than in females, the ratio for intralobar being 1.5:1 and extralobar being 3:1.[1] Although pulmonary sequestration has been reported in all ages, the majority of patients manifest symptoms early in life. Extralobar sequestration generally is diagnosed in children 10 years of age and under.[1]

Pulmonary sequestration is associated with a number of congenital malformations. Intralobar sequestration is rarely associated with any congenital anomalies. Fifteen to 40 percent of extralobar sequestrations are found to have associated congenital abnormalities.[1-4] The most common abnormality is diaphragmatic hernia. A proposed explanation is that the aberrant lung tissue interferes with the proper closure of the pleuroperitoneal canal.[3] Among the other abnormalities are diaphragmatic eventration or paralysis, congenital heart defects, pericardial cyst and defects, pulmonary agenesis, foregut duplications or diverticulum, ectopic pancreatic tissue, congenital megacolon, and complete duplication of a colon.[1,3]

CASE HISTORY

A 20-year-old, asymptomatic white woman presented to the Queens Hospital Center outpatient clinic for a routine examination. Results of the physical examination and the past medical history were unremarkable. A routine chest radiograph revealed a large ill-defined infiltrate of nonhomogeneous density in the left upper lobe with associated atelectasis (Figure 1). The patient was placed on antibiotic therapy; however, she failed to return for a follow-up examination. One year later the patient returned for another routine examination and the previous lesion was noted again, unchanged from prior examination. Multiple tomograms taken through the lesion showed a fairly well circumscribed mass-like density with a necrotic center (Figure 2). At this juncture, the presumptive diagnosis was changed to pulmonary sequestration. A left upper lobectomy was performed, and the specimen subjected to histologic examination (Figure 3).

Microscopic examination revealed cystically dilated bronchi and bronchioles filled with mucopurulent exudate and lined by respiratory epithelium that was focally ulcerated. The surrounding parenchyma contained large plates of cartilage within fibrotic chronically inflamed stroma. Focal foreign body giant cells, foamy histiocytes, and pigment-laden macrophages were also noted (Figure 4). The adjacent pulmonary parenchyma showed organizing pneumonitis and vascular sclerosis. Sections of an aberrant artery revealed a prominent internal elastica but no significant inti-

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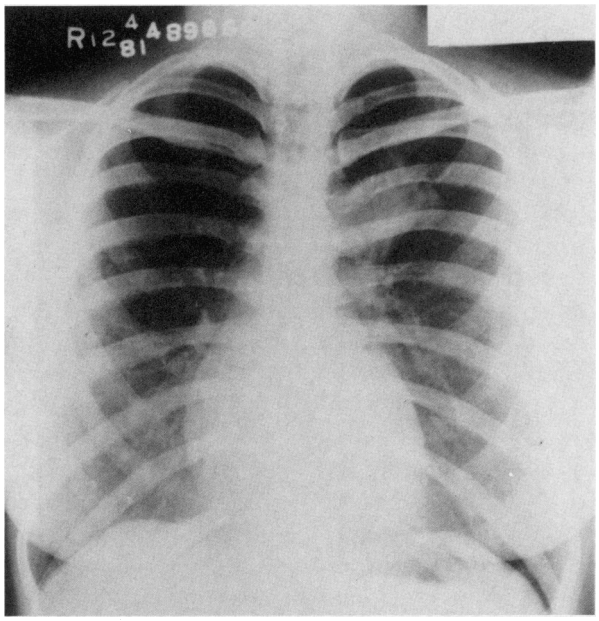

Figure 1. Plain film of the chest. Lesion seen in the left upper lobe

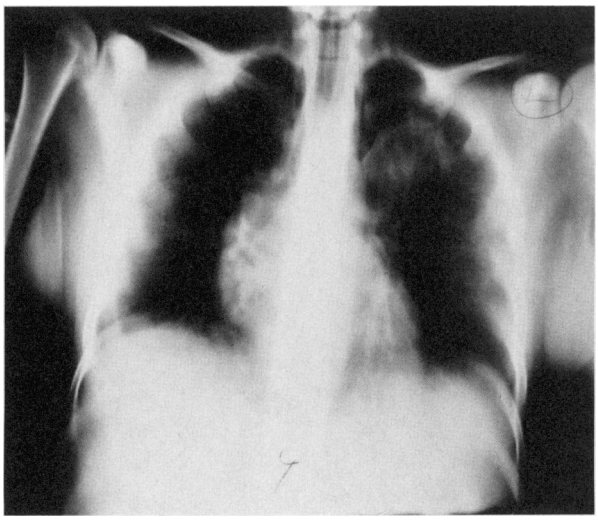

Figure 2. Tomogram of the chest. Lesion is noted in the left upper lobe

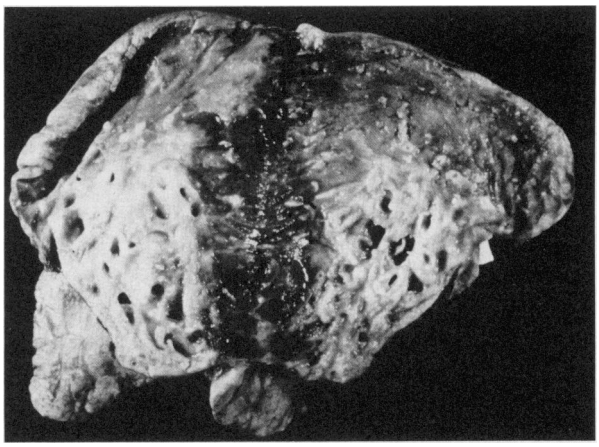

Figure 3. Intralobar bronchopulmonary sequestration in the left upper lobe of a 20-year-old woman. The large, wedge-shaped mass present within the lung is composed of collapsed and fibrotic pulmonary parenchyma containing multiple cystic spaces

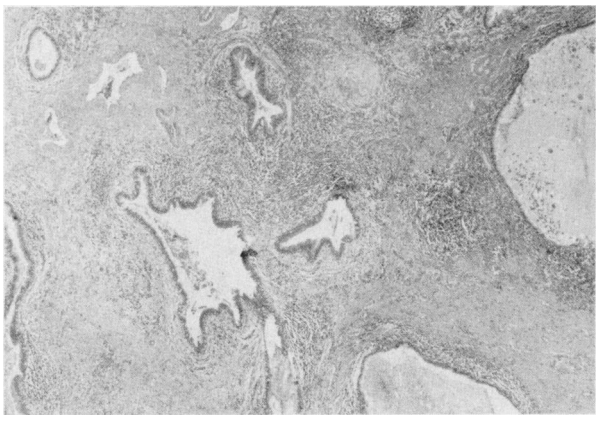

Figure 4. Light photomicrograph of mass illustrated in Figure 3 demonstrating multiple epithelial-lined cystic spaces filled with mucus and polymorphonuclear leukocytes. The surrounding pulmonary parenchyma is replaced by dense, chronically inflamed fibrous tissue

mal thickening. Thus the final pathologic diagnosis was intralobar pulmonary sequestration of the left upper lobe.

The patient's postsurgical course was uneventful. Follow-up was unremarkable.

DISCUSSION

The clinical presentation of pulmonary sequestration is characterized by many signs and symptoms. In many cases it is discovered by routine roentgenographic examination of the chest. However, often these patients may present with symptoms.[3]

Pulmonary sequestrations may be manifested clinically by a recurring localized infection in a fluid-filled cyst. Intralobar sequestration is diagnosed more frequently because of the increased incidence of lung infection.[1] In the older age group, symptoms are usually due to the presence of airway communication between the sequestration and the normal bronchial tree. Recurrent fever, chills, and purulent sputum production are the most common signs. Secretion of mucus within an intralobar sequestration causes a cystic swelling with subsequent compression and atelectasis of the normal surrounding lung tissue.[7] Extralobar sequestration can very often be asymptomatic. Usually it is diagnosed from an incidental finding on a routine chest roentgenographic examination or noted during repair of congenital diaphragmatic hernia. Extralobar sequestration can present with recurrent pulmonary infection, but this is rare.[3] Extralobar sequestrations often have a fistulous type of communication to the esophagus or stomach. These can present with gastrointestinal symptoms, dysphagia, regurgitation, vomiting, and hematemesis. These findings are most likely secondary to compression and discharge of blood and infected material into the gastrointestinal tract.[3]

Additional symptomatology can be related to cardiovascular malformations. Congestive heart failure may develop as a result of an increase in cardiac output. The increased output results from a left to right shunt involving the aberrant systemic artery through the sequestration with drainage into the pulmonary veins. These hemodynamic difficulties are usually manifested during the first few weeks or months of life. Occasionally, patients may present with pulmonary hypertension or a pansystolic murmur, which is the result of the large arterial blood supply through the sequestration.[3,7]

The differential diagnosis commonly includes pneumonia, empyema, bronchiectasis, lung abscess, bronchogenic or enterogenous cyst, Bochdalek hernia, and mediastinal or pulmonary neoplasm.[1]

Diagnosis can be based on plain roentgenograms of the chest, which almost always show some abnormality. If the index of suspicion is high, correlation of the radiographic findings and the clinical presentation can lead to the diagnosis. Intralobar sequestration will appear on a chest film as a dense, diffuse mass or infiltrate, usually seen in the posterior basal segment of the left lower lobe. The density may appear cystic; occasionally, an air fluid level is seen. Extralobar sequestration tends to appear as a homogeneous triangular density.[1,3-5]

Many authors advocate the use of bronchography, which may show displacement of the bronchial tree due to the space-occupying lesion. Bronchography, tomography, and bronchoscopy are usually performed, but tend to offer little specific diagnostic information.[3] The definitive study for diagnosis is aortography with selective arteriography.[3,4,6] The demonstration of the aberrant vessel is considered diagnostic.[1] In cases of suspected extralobar sequestration, a fistulous communication to the gastrointestinal tract should be excluded by means of a contrast study of the upper gastrointestinal tract, which will usually demonstrate the tract, if present.[3]

The definitive treatment of pulmonary sequestration is surgical resection. Preoperatively, infection should be controlled by antibiotics.[7] Extralobar sequestration, although usually asymptomatic, should be resected to prevent the hemodynamic manifestations.[3]

In the present case the location of the sequestration is atypical.[6] However, it clearly shows the need for the clinician and radiologist to work together to arrive at the diagnosis of pulmonary sequestration.

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