Intralobar Bronchopulmonary Sequestration with Bronchial Communication*

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Surgical resection in unsuspected intralobar sequestration can result in fatal hemorrhagic complications. The roentgen criteria for establishing this diagnosis are: 1) chronic and recurrent pulmonary infiltrate; 2) most often located in basilar segments of the lower lobes; 3) multiple cystic areas; 4) failure of contrast media to enter the cystic area during bronchography. We studied two cases of intralobar sequestration where on bronchography the contrast medium entered the cystic areas. To our knowledge only four other similar cases are reported in the literature. In our cases the preoperative diagnosis was confirmed by aortography. Since the roentgenographic and bronchographic findings alone would have suggested the possibility of cystic bronchiectasis, the importance of aortography is stressed.

The importance of establishing a preoperative diagnosis of intralobar sequestration has been emphasized by sporadic case reports of fatal hemorrhage during lobectomy for unsuspected cases.1,2 The occurrence of such complications is easily understood when one appreciates that the feeding vessel of intralobar sequestration is a medium-sized elastic artery arising in most instances directly from the aorta. The difficulty in controlling bleeding from this vessel is compounded by the fact that the vessel frequently originates from the abdominal aorta3 and in most cases shows marked arteriosclerotic changes.

Aortography and selective angiography will readily confirm this diagnosis.3-5 However, these procedures are not routinely performed on patients with lung disease unless one entertains the possibility of pulmonary sequestration. The suspicion is usually raised by the roentgen findings of: 1) chronic and recurrent pulmonary infiltrate; 2) most commonly located in the medial basilar segment of the lower lobe, especially on the left; 3) multiple cystic changes.

As was pointed out by Bruwer and co-workers6 "bronchography practically invariably reveals failure of the opaque material to enter the region" even when there are airfluid levels in the cystic areas.

This paper reports two cases of intralobar pulmonary sequestration where the cystic spaces filled with contrast media during bronchography and reports the incidence of this finding from review of the literature.

FIGURE 1. Case 1. PA chest.
INTRALOBAR BRONCHOPULMONARY SEQUESTRATION

CASE REPORTS

CASE 1

The patient is a white woman first admitted to Bronx Municipal Hospital at the age of 18 with infiltrates in the left upper and lower lobes. The infiltrates in the left lower lobe were associated with multiple cystic changes suggesting bronchiectasis. Sputum cultures for tubercle bacillus were positive on three consecutive specimens. Because the patient was five months' pregnant, further radiologic studies were deferred. She was treated with antituberculous agents and showed a good clinical response.

Three years later, she was readmitted because of productive cough and persistent infiltrate in the basilar segment of the left lower lobe (Fig 1). Bronchography revealed opacification of the cystic spaces with contrast material and she was discharged with the diagnosis of cystic bronchiectasis. Four years later the patient returned complaining of several episodes of hemoptysis and was readmitted. Bronchography once again showed filling of the cystic areas with contrast (Fig 2). Aortography demonstrated a medium-sized artery arising from the abdominal aorta supplying the involved portion of the left lower lobe. Selective injection of the abnormal vessel demonstrated drainage into the pulmonary veins and also resulted in opacification of peripheral branches of the pulmonary artery (Fig 3). A diagnosis of intralobar sequestration was made and confirmed at surgical resection.

Examination of the specimen (Fig 4) revealed the lobar bronchus to be slightly tortuous and minimally dilated. All segmental bronchi could be traced except for the one to the posterior basal segment. The latter segment was entirely occupied by an irregular lobulated cystic structure measuring 3.5 cm in diameter. The lumen was devoid of contents. The inner aspect was shiny gray and wrinkled and there were several openings measuring up to 0.4 cm in diameter through which the lumen of the cyst communicated with distal bronchi in the adjacent segments of lung. The wall of the cyst was white gray, fibrous, measuring from 0.2 to 0.4 cm in thickness. Numerous bronchi in all other segments showed moderate cylindrical dilatation of the lumens, but the parenchyma was not consolidated nor disrupted. A ligated stump of a vessel was present in the midportion of the diaphragmatic aspect of the specimen. The vessel penetrated into the lung and could be traced for a distance of 1.5 cm where it was located within the fibrous tissue separating the cystic structure of the posterior segment from the remainder of the pulmonary parenchyma. The vessel measured 0.5 cm in diameter and it had a patent lumen although yellow, raised sclerotic plaques were present on the intimal surface. There were extensive fibrous adhesions covering the diaphragmatic pleura. Microscopically the cyst proved to be lined by a single layer of ciliated epithelium. The wall was fibrous connective tissue with occasional muscle bundles but no cartilage. Adjacent to the cyst was the aberrant vessel which showed intimal sclerosis and numerous elastic lamellae arranged in a concentric fashion, typical of a systemic artery. There were occasional inflammatory cells in the wall of the cyst and in the bronchi of other portions of the lung.

CASE 2

The patient is an 18-year-old white youth who was admitted to the Hospital of AECOM for investigation of the cause of recurrent episodes of pneumonia in the left lower lobe since the age of 12. The chest x-ray film showed basilar infiltration in the left lower lobe (Fig 5). Bronchography revealed an irregular cystic area filled with contrast in the posterior segment of the left lower lobe (Fig 6).

An aortogram demonstrated a large vessel originating from the abdominal aorta supplying the involved segment of the left lower lobe. Late films in the series showed drainage into the pulmonary veins (Fig 7).

A left lower lobectomy was performed and the abnormal

Figure 2. Case 1. A (left) Post-tussive PA film from bronchogram. B (right) Lateral film from same study. Note extensive filling of cystic spaces with contrast.
The specimen, left lower lobe of lung, was essentially similar to that of case 1. The cystic spaces in the posterior basal segment showed no obvious communication with the bronchial tree and no bronchiectasis was present elsewhere. The fibrous wall of the cyst was lined by a single layer of ciliated epithelium. The aberrant artery, which penetrated the lung through the diaphragmatic surface, showed abundant elastic fibers.

Intralobar sequestration as described by Pryce and associates is a condition in which a bronchopulmonary mass is supplied by a systemic artery; or in other words the diseased area is sequestered from its normal pulmonary arterial supply.

Although he believed the etiology to be related to traction by the abnormal vessel during embryologic development, the questions raised by Abbey-Smith and his alternative hypothesis are very intriguing. He proposed that the primary defect is a failure of development of the peripheral branches of the primitive pulmonary artery and as a result there is persistent supply from the primitive dorsal aorta, as exists in early embryonic life. The cystic changes result after birth secondary to the establishment of systemic pressures in the aberrant circulation and to the proliferation of the mesodermal elements induced by the systemic artery. The reader is referred to the original reports for detailed discussion.


The frequent association of inflammatory changes with intralobar sequestration and the known association of systemic to pulmonary collateral circulation in cases of chronic inflammation of the lung have prompted some authors to question the existence of the entity of intralobar sequestration. However, there are reports of cases of intralobar sequestration without infection. Furthermore, the systemic vessels in chronic inflammation most frequently originate from bronchial or intercostal arteries as opposed to the aorta in sequestration. Histologically, the aberrant artery in intralobar sequestration has a prominent elastic component and atheromatous changes are very common. Our first case also demonstrated communication from the aberrant systemic artery to the pulmonary artery, probably as a result of chronic inflammation. Similar findings were reported by Cole and colleagues (cases 1 and 6) and others.

On review of the literature we found only four other cases where bronchography demonstrated opacification of the cystic areas. The possibility that the bronchial communication as demonstrated on bronchography is a result of the associated inflammation is quite likely. In both of our cases, and in case five of Cole, case 1 of Pryce and co-workers and case 1 of Abbey-Smith where this was demonstrated, inflammatory changes were noted on pathologic examination.
The significance of this finding is more than academic. As pointed out before, had aortography not been performed, chronic bronchiectasis would have been thought to be the most likely clinical diagnosis. Although we have found only four other cases of bronchial communication demonstrated on bronchography in over 200 reported cases of intralobar sequestration, the surgical implications of intralobar sequestration make it mandatory that aortography be performed in any persistent inflammatory process in the lower lobes, especially in a young patient.

CONCLUSIONS

We studied two patients with intralobar sequestration where the cystic spaces in the involved area filled with contrast medium during bronchography. Four similar cases were found described in the literature. Although this finding is uncommon, the importance of establishing a preoperative diagnosis of intralobar sequestration is emphasized. Aortography should be performed in all suspected cases.

REFERENCES

3 Raniger K, Valvassori G: Angiographic diagnosis of intralobar pulmonary sequestration. Amer J Roentgen
Figure 7. Case 2. A (upper) Selective injection aberrant vessel. B (lower) Venous phase demonstrating drainage into left atrium.


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