Pulmonary Intralobar Sequestration

A REVIEW OF THE LITERATURE WITH TWO CASE REPORTS

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SUMMARY

Two cases of pulmonary intralobar sequestration are presented, with aortography and selective arteriography. These examinations are prerequisites for the diagnosis and management of a patient with suspected sequestration. There should be a high degree of suspicion in a case of a persistent left lower-lobe tumour.

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Pulmonary sequestration is a well-recognised developmental anomaly, in which a portion of the lung in its primitive form has become separated from the rest of the lung.[1] It receives its blood supply from the aorta via an abnormal aberrant vessel.

There are two types of sequestration—an extra- and an intralobar variety. In the extralobar type, ectopic lung tissue is situated below or above the diaphragm and is enveloped in its own pleura, with venous drainage into the systemic circulation, that is, the inferior or superior vena cava, the azygos, or portal vein.[2,3] The intralobar type differs in that the venous drainage is invariably into the pulmonary veins[4] and is without its own pleural covering.

The first report of pulmonary sequestration by Rokitansky and Rektorzik appeared in 1861,[5,6] but it was not until after the Harris-Lewis publication in 1940[7] and the classical description by Price *et al.*[8,9] that this entity became better known.

More than 250 cases of intralobar sequestration have been published.[3] However, failure to recognise this condition may result in the inadvertent incision of the anomalous arterial supply, with subsequent exsanguination. Fortunately, surgeons keep sequestration in mind, but according to Bruwer *et al.*,[10] 3% of the patients undergoing operation have died because of a failure to recognise the entity.

Findlay and Maier[11] mention that surgeons should palpate the pulmonary ligament as a routine before any pulmonary resection, as anomalous vessels frequently arise in this structure, even without sequestration. In view of the surgical implication, a diagnosis of intralobar sequestration should be considered in any persistent inflammatory process in a lower lobe (usually the left), especially in a young patient.

CASE REPORTS

Case 1

A 12-year-old girl was admitted to hospital with recurrent and persistent infiltration in the right lower lobe. Two months before this admission, the patient had been hospitalised in another institution for recurrent right lower-lobe pneumonitis.

The chest X-ray examination, including tomograms, showed a density in the posterior basal segment of the right lower lobe (Fig. 1), and a subsequent bronchogram revealed no filling of this region with contrast material. Bronchoscopy failed to show the posterior basal segment bronchus, and in view of the above findings, sequestration was considered. She was treated for pneumonia and discharged in excellent condition.

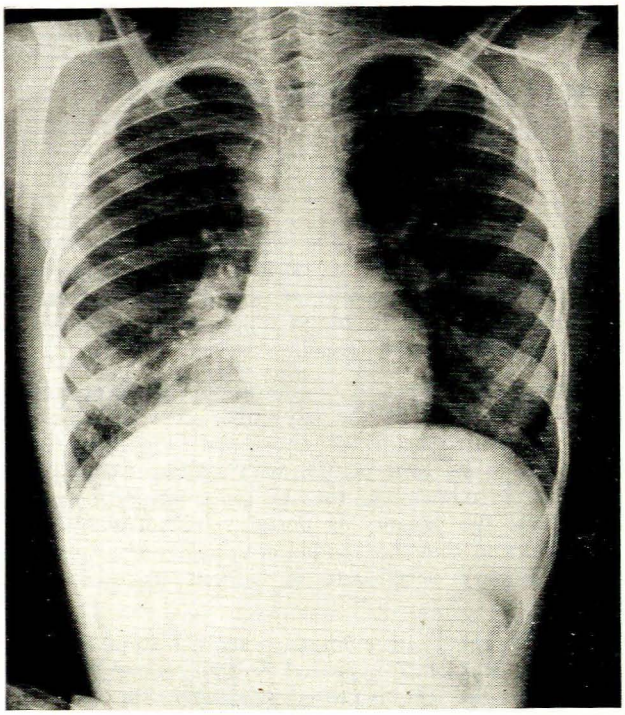

Fig. 1. Case 1. Infiltration in the posterior basal segment of the right lower lobe.

On a second admission, aortography and a selective arteriogram established the diagnosis by demonstrating an aberrant vessel from the aorta (Fig. 2), supplying the sequestered portion of the lung with venous drainage via the pulmonary vein. Resection of the diseased segment was undertaken, and the aberrant artery and pulmonary vein were divided. A specimen showed no bronchi, but an ovoid, cystic tumour in the centre of the specimen, containing homogeneous, gelatinous, pale yellow material, was demonstrated. The patient was discharged without complications.

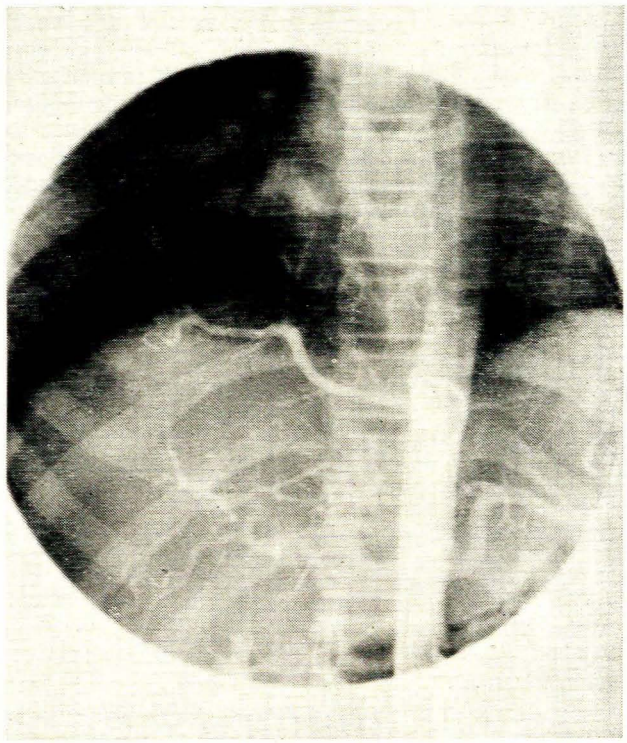

Fig. 2. Case 1. Aortography and selective sequestered arteriogram demonstrates an aberrant vessel from the aorta supplying the sequestered portion of the lung.

Case 2

A 15-year-old girl was admitted to hospital with a history of left lower chest pain, coughing, high fever, and chills, of 2 days' duration. The history indicated 4 previous similar episodes over several years. The chest radiograph showed an infiltrate in the left lower lobe (Fig. 3), which was treated with antibiotics, but 2 weeks later no significant changes in the X-ray film were demonstrated. However, an air-fluid level in the same area was noted. On tomography, air-fluid levels with a lobulated tumour were distinctly shown. Pulmonary sequestration was therefore considered.

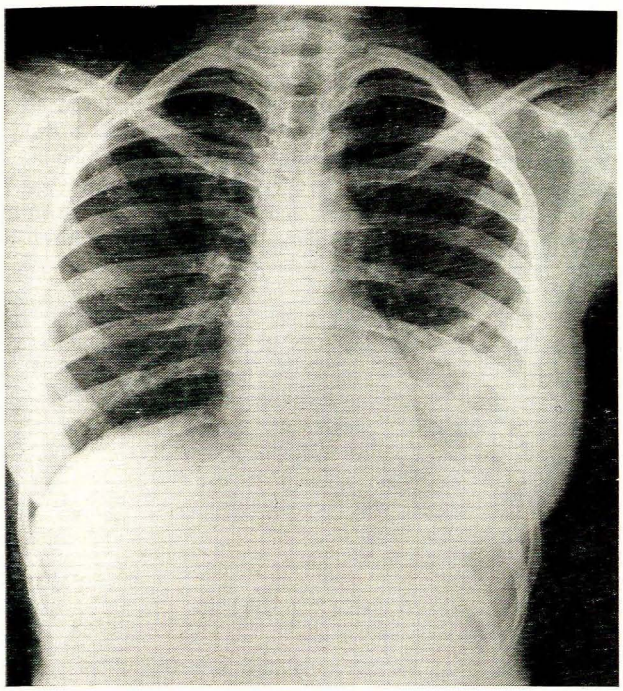

Fig. 3. Case 2. Postero-anterior chest film demonstrating an infiltration in the left lower lobe.

Subsequent aortography and a selective arteriogram showed a large aberrant vessel arising at the level of T10 and T11 (Fig. 4), from the abdominal aorta which supplied the lobulated left lower lobe tumour. Venous drainage was by a pulmonary vein draining into the left atrium. Another smaller vein drained downward and medially into the hemiazygous system (Fig. 5).

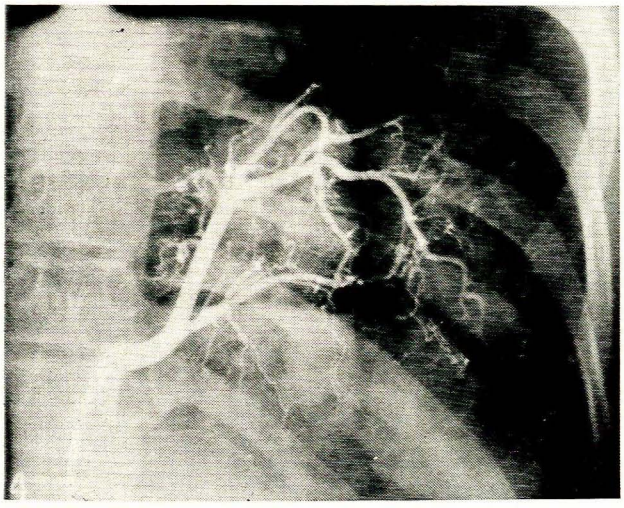

Fig 4. Case 2. Aortogram and selective sequestered arteriogram showing a large aberrant vessel arising at the level of T10 and T11 from the abdominal aorta and supplying the left lower lobe tumour.

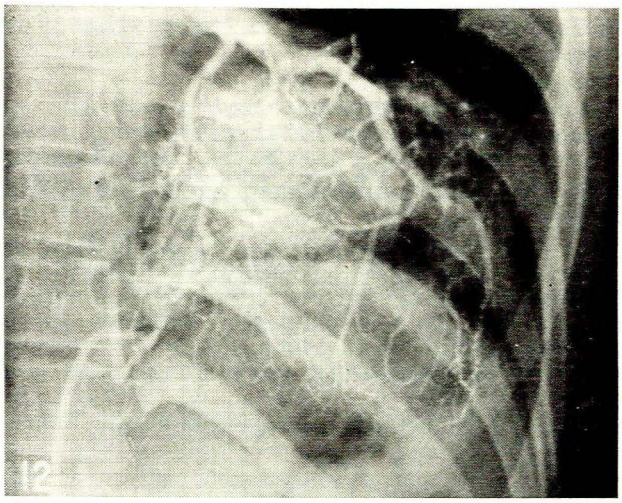

Fig. 5. Case 2. Late film of arteriogram demonstrating the small vein draining into the hemiazygous system.

Bronchography demonstrated marked diminution in the number of bronchioles, and very few tertiary and smaller bronchioles which showed no connection with the cystic structure. Normal bronchi were stretched around this area.

At operation, a large aberrant artery arising from the aorta, and a smaller vessel from the pulmonary artery, were encountered, and noted to be supplying the sequestered region. The arteries were clamped and ligated. Segmental branches arising from the left lower lobe, and veins, were identified, ligated, and the sequestered segment resected. A specimen showed a well-circumscribed multilocular structure filled with pale yellow, gelatinous material. Bronchi of the normal lung were stretched around the cystic structure.

DISCUSSION

The symptomatology of pulmonary intralobar sequestration is non-specific. It includes cough, fever, and chest pain. Infrequently patients are asymptomatic. Clinically it may be confused with pneumonia, bronchiectasis, empyema, or bronchitis. Treatment is usually by antibiotics, but only temporary relief is achieved, with recurrence of symptoms being quite characteristic. Definitive therapy is resection of the segment. The sexes are equally involved. More than 50% of patients are below the age of 20 years,[12] with the posterior segment of the left lower lobe being involved in two-thirds of all reported cases, and the posterior segment of the right lower lobe being next in frequency.[12] The upper lobes are rarely involved.

The chest X-ray film usually shows a tumour suggesting bronchiectasis, and when infection supervenes, infiltration with air-fluid levels may be demonstrated (case 2). Tomography of this area may show the aberrant arterial supply.[13] The bronchogram fails to show filling of the bronchi in the region of the tumour, but an arcuate displacement of the bronchi around this area.[10] The latter differs from the

findings in bronchiectasis, where contrast-filled dilated bronchi are readily seen. Zelefsky *et al.*[14] reported on 6 cases in the literature (2 of which were their own), where, on bronchography, the contrast medium entered the cystic area, suggesting cystic bronchiectasis, and in view of this the importance of arteriography was stressed in making a diagnosis. Arteriography confirms the diagnosis by showing an aberrant vessel which usually arises from the lower thoracic or abdominal aorta and is usually single. Turk and Lindskog,[12] in their review of the literature consisting of 114 surgically-proved cases, reported the aberrant vessel to be arising from the thoracic aorta in 74 cases; the abdominal aorta in 25; the intercostal arteries in 5; and to be of multiple origin in 17 cases.

Selective studies of the aberrant vessel have several advantages,[3] in that the venous drainage may be more readily demonstrated, and therefore helpful in the diagnosis of intra- or extralobar sequestration; and an adequate extent of the region supplied by the abnormal vessel may be shown, so helping to assess the amount of resection to be undertaken, as this depends greatly on the amount of aberrant vessel supply.

In their report, Turk and Lindskog[12] noted 6 cases as having venous drainage to the right side of the heart via the inferior vena cava, instead of the much more usual drainage to the pulmonary veins. These veins were shown to be smaller than the supplying artery, suggesting that only a portion of the venous blood returns to the right heart and that some must enter the pulmonary circulation. Venous drainage to the portal system is rarely encountered and only one such case has been reported.[15]

The second case reported in this article is rather atypical, as the venous drainage passed into the hemiazygous vein (systemic circulation) and a small vein drained into the pulmonary vein (pulmonary circulation). Only one other such case appears in the literature.[4]

Gross pathology of intralobar sequestration has been classified by Price[9] as follows: (*a*) a large cyst in the sequestered region in 50% of the cases; (*b*) a polycystic tumour in 40% of cases; and (*c*) a sequestered tumour containing bronchi which branch parallel to the artery.

The cystic or bronchial elements are usually completely dissociated from the normal bronchial tree, with the anomalous vessel lying in the centre of the sequestered tumour. The vessel has a thick wall and is elastic, similar to the pulmonary artery,[10] and is frequently arteriosclerotic. The cysts are lined with respiratory epithelium, and usually filled with thick, brown, mucoid material.

Different theories on the origin and embryological significance of the sequestration have been presented by various authors. The anomalous vessel is believed to be a persistent branch from the dorsal or ventral aorta, to the splanchnic plexus, although the exact manner of its persistence has not as yet been satisfactorily established.[14,17] As the lung bud develops ventrally from the primitive foregut, one of its developing branches apparently becomes detached and acquires a systemic arterial blood supply from the persistent branch of the splanchnic plexus.[12] A relatively complete survey of the various theories of the aetiology was presented by Nielsen[2] and Gallagher.[18]

Findlay and Maier[11] found that when the origin of an anomalous pulmonary vessel was from the systemic system above the level of the lung roots, there were associated anomalies of the heart and great vessels in 75% of the cases. If the origin was below this level, 16% of cases showed such abnormalities.

CONCLUSION

It is obvious that aortography and selective arteriography are absolute prerequisites for the diagnosis and proper management of a patient with suspected sequestration. A high degree of suspicion should be present in cases of a left, lower-lobe, persistent tumour. This should be an indication for angiography.

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