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A case of middle lobe pulmonary sequestration

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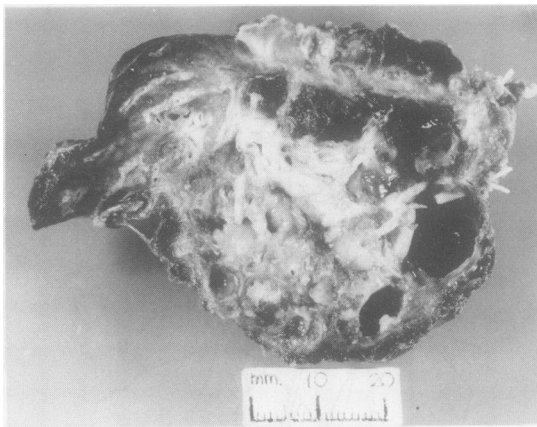
Pulmonary sequestration is a rare anomaly, occurring in nine out of 88 000 patients admitted to a children's hospital.[1] It is defined as a mass of lung tissue that does not communicate with the tracheobronchial tree in the normal fashion and receives its blood supply from systemic arteries.[2] It is subdivided into intralobar and extralobar sequestrations. Intralobar sequestrations occur within a pulmonary lobe, while extralobar sequestrations lie outside the visceral pleura.[2] There are numerous anatomical variations[3] and it has been argued that intralobar and extralobar sequestration are variants of the same abnormality.[3 4] We report a case of intralobar sequestration in which the unusual location and anatomical features support the concept of a range of anomalies from intralobar to extralobar sequestration.

Case report

A female infant was delivered normally at full term with a birth weight of 2·83 kg. From 3 months of age she had recurrent upper respiratory tract infections, and presented at 8 months with right sided pneumonia and heart failure. A continuous murmur was heard over the right chest, and a radiograph showed extensive shadowing with multiple cysts in the right lung. A sweat test gave a normal result. At cardiac catheterisation at 10 months no cardiac abnormality was found, but a large artery was shown to arise from the right subclavian artery and to supply a mid zone cystic area in the right lung. At left ventricular angiography contrast medium passed into the lesion and thence into the right pulmonary artery, suggesting an associated arteriovenous malformation. A xenon perfusion lung scan indicated considerable disparity in uptake between left (63%) and right (37%) lungs, and bronchography showed displacement of bronchi above and below the cystic area.

Because of recurrent chest infections and poor weight gain the right middle lobe was resected when the child was 30 months of age. At operation the middle lobe was rubbery, airless and densely adherent to the chest wall, pericardium, and diaphragm. It was completely separated from the lower lobe by the oblique fissure and incompletely from the upper lobe by a poorly developed transverse fissure. By inflation of the lung demarcation of the inflated upper lobe and airless middle lobe was made obvious, allowing separation of the two with minimal air leak. Numerous systemic arteries entered it from the intercostals laterally, a large and tortuous internal mammary artery medially, and the diaphragmatic vessels inferiorly. It was also supplied by two pulmonary artery branches, one running behind the middle lobe pulmonary vein and the other passing up from the oblique fissure. Venous drainage was via the middle lobe pulmonary vein. There was no middle lobe bronchus. The postoperative course was uneventful and the child was well at follow up nine months later, though her weight was still under the third percentile.



Cut surface of the right middle lobe sequestration showing mucus filled bronchi and cystic spaces.

The resected lobe measured 8 × 5 × 2 cm. Its external surface was covered by a fibrinous exudate and fibrous adhesions. The cut surface showed mucus filled dilated bronchi and cystic spaces of varying sizes (figure). The adjacent lung was compressed and contained focal areas of fibrosis. Microscopy showed bronchi with well developed cartilage and submucosal glands. The cysts were lined by ciliated respiratory epithelium, and some of the alveoli were lined by cuboidal cells. There was no evidence of immature or dysplastic parenchyma. There was interstitial fibrosis, a mild mononuclear cell inflammatory infiltrate, and occasional lymphoid aggregates. Several pulmonary and thick walled muscular systemic arteries were seen.

Discussion

A pulmonary sequestration situated in the right middle lobe is extremely rare. In a review of 80 children with this malformation,[5] 56 had an intralobar sequestration, all of which were in either the right or the left lower lobe. Savic *et al*

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reviewed 400 intralobar sequestrations, and found only one case in which the right middle lobe was affected. All of the others were in the lower lobes.

We believe this to be the second reported case of sequestration in the right middle lobe, in this instance affecting the whole lobe and representing a form that is intermediate between the intralobar and extrapulmonary variants, with features of both. It is also unusual in having a dual systemic and pulmonary arterial supply. For 373 of the intralobar sequestrations reviewed by Savic *et al*[6] the origin of the supplying artery was known. It arose from the thoracic or abdominal aorta in 346 cases (92·6%), and in the remainder from other systemic arteries in the mediastinum or abdomen. None of the sequestrations was supplied by the pulmonary artery. In contrast, five out of 91 extralobar sequestrations had a pulmonary arterial supply. Only 55 (14·7%) of the intralobar sequestrations had a multiple systemic arterial supply. Our patient also had no communication between the sequestration and the bronchial tree. Bronchial communication is rare in extralobar sequestration but occurs occasionally in intralobar sequestration.[3 6] These features support the concept of a "sequestration spectrum"[3] that includes wide variations in vascular supply, anatomic location, and associated anomalies.

The venous return was via the pulmonary vein, as in most cases of intralobar sequestration. At left ventricular angiography contrast passed from the right subclavian artery via the internal mammary artery into the sequestration and thence into the right pulmonary artery, indicating a dual arterial supply as well as a degree of left to right shunt. The shunt was small, and there was no significant step up in oxygen saturation in the main pulmonary artery. Congestive heart failure, however, has been reported as a rare presenting symptom in infants with pulmonary sequestration.[7] This results from volume overload in the left atrium and ventricle due to the extra pulmonary venous return from the systemic arteries via the sequestrated lobe.

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