Extralobar pulmonary sequestration

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Extralobar sequestration of pulmonary tissues is relatively rare and seldom produces symptoms. An incidental finding is described of extralobar sequestration of the lung in a Nigerian child, presenting as an anterior mediastinal polycystic mass. The mass was attached to the right pulmonary artery by a fibrovascular pedicle which contained a small elastic artery, veins, and nerve bundles. Sequestrated lobes are usually situated in the posterior basal lobes of the lungs and may be associated with other congenital abnormalities, but the case reported here differs in these respects. Death was due to enterocolitis unrelated to the congenital abnormality.

Tissues of bronchopulmonary origin, which have no communication with the bronchial tree and are supplied by anomalous vessels, are relatively rare and have been referred to by a variety of terms. These include accessory lung, sequestrated lobe of lung, ectopic lobe of lung, dissociated or dislocated lobe, aberrant lobe, or supernumerary lobe. The term ‘sequestrated lobe’ may not be entirely appropriate but is consecrated by use. Such a lobe may be classified as intralobar, extralobar, or complete sequestration, depending on its anatomical relationship to the main lung masses. The earlier examples of the abnormality were described by Rokitansky (1861) and Rektorzik (1861), and since then the entity has been recognized more frequently (Harris and Lewis, 1940; Gruenfeld and Gray, 1941; Arce, 1943; Davies and Gunz, 1944; Pryce, Sellors, and Blair, 1947; Lemmon, Kirklin, and Dockerty, 1954). In fact, this condition is not peculiar to man; at least 21 cases have been reported in cattle and one case in a horse (Davidson, 1956). The intralobar variety appears to be the commonest and more than 250 cases have been reported in the literature (Ranniger and Valvassori, 1964). There are, however, fewer reports of the extralobar and complete varieties.

The disparity in occurrence between the intralobar variety and the extralobar and complete varieties may be due to the fact that the latter two are not as susceptible to infective complications as the intralobar variety and therefore are less frequently detected. Furthermore, the intralobar variety from its anatomical location is often detected in the course of investigations for other pulmonary lesions which it may simulate on clinical or radiological grounds. The case presented here is an incidental necropsy finding in a Nigerian child who died from bacterial enterocolitis but had a right-sided extralobar sequestrated lobe attached anteriorly to the right pulmonary artery by a fibrovascular pedicle.

CASE REPORT

A 3-year-old Nigerian boy was admitted to University College Hospital, Ibadan, complaining of a discharging right eye, generalized abdominal pain, and diarrhoea and vomiting of seven days' duration. On admission he was moribund, had periorbital cellulitis, and was dehydrated. On examination, he had a right chronic conjunctivitis and a discharging sinus over the right maxilla, but no other significant physical signs were elicited. The clinical diagnoses were gastroenteritis and conjunctivitis.

Bacteriological examination of the conjunctival exudate yielded Staphylococcus pyogenes, and stool cultures revealed Shigella dysenteriae 2 in one and Salmonella group 'G' in the other. The Widal test on a specimen of serum was positive for Salmonella paratyphi B at a titre of up to 1/160 and 1/40 for the H antigen. Haemoglobin 8·6 g./100 ml. (59%), P.C.V. 27%. M.C.H.C. 32%. W.B.C. 10,000/c.mm., with 38% neutrophils, 58% lymphocytes, and 4% monocytes. Electrolytes: sodium, 110 mEq/l.; Cl⁻, 77 mEq/l.; K⁺, 5-2 mEq/l. Serum urea, 17 mg./100 ml. Haemoglobin genotype, AS. The child was given appropriate antibiotics orally as well as locally to the right eye, and was hydrated adequately by intravenous fluid therapy. He gradually deteriorated and died.

PATHOLOGY Necropsy was performed 24 hours after death. The body was that of a 10-kg. boy, 86 cm. tall with bilateral periorbital ulceration of the skin and a yellowish exudate of the right conjunctiva.
FIG. 1. Sequestrated lobe lifted to show that it is not connected to any underlying structures. Note the right pulmonary artery running posteriorly to the superior vena cava (with a black rod); the pedicle is medial to it.

FIG. 2. Microscopic appearance of the wall of a cyst. The lumen contains mucin, the epithelium is thrown into small folds, and mucous glands and cartilage are present. P.A.S.-Tartrazine. × 200.
There was pus in the right middle ear. There were several minute ulcers in the terminal ileum and sigmoid colon and the rest of the intestines appeared congested.

In the anterior mediastinum there was a pedunculated, multicystic mass, measuring \(4 \times 2.5 \times 1\) cm., attached to the right pulmonary artery by a 1-cm. long whitish fibrous pedicle (Fig. 1). The mass was partially overlapped by the anterior margin of the right lung and the mediastinal surface of the lung was its lateral relation. The ascending aorta lay inferior and medial to it. The mass was fluctuant and poorly translucent. Although its pedicle was firmly attached to the right pulmonary artery there was no demonstrable communication with it. The cut surface of the mass confirmed the presence of several cystic spaces containing semisolid, greenish-yellow, mucoid material. The cut surface of the pedicle had a cribriform appearance suggestive of a fibrovascular structure. There were no abnormalities in either lung or other organs examined.

Histologically, the cysts were lined by ciliated, columnar epithelium thrown into folds in places. The wall of a cystic space from without was composed of a thin layer of pleura, fibrous tissue, a few plates of cartilage, and mucous glands (Fig. 2). A few thin-walled capillaries were also present in the wall. The cysts contained an amorphous, eosinophilic secretion shown to be mucin on P.A.S. staining. In all sections examined no lung parenchyma was seen.

The pedicle was composed of fibrovascular tissue with one thick-walled artery, several veins, and nerve bundles. The veins showed no changes, but there was considerable reduplication of elastic tissue extending throughout the media of the artery. The intima of the artery showed no significant changes, and tissues of bronchopulmonary origin were not seen in the pedicle.

No histological changes were seen in several sections taken from the lungs except moderate congestion of alveolar capillaries. There was a necrotizing enterocolitis consistent with Salmonella or Shigella infection.

**DISCUSSION**

The aetiology and pathogenesis of pulmonary sequestration remain poorly understood but have been described and adequately discussed by Spencer (1963). The present case, which appeared as an anterior mediastinal polycystic mass, is an example of extralobar sequestration situated in an unusual site, and its direct attachment to the right pulmonary artery is considered to be very rare. The case (No. 1) described by Baar and d'Abreu (1949) was different from ours although their sequestrated lobe derived its blood supply from the left pulmonary artery and was attached to the upper lobe of the left lung by a thin band of connective tissue within which lay a branch of the left pulmonary artery and vein: this branch was shown to communicate with the left pulmonary artery. Another case was described by Gans and Potts (1951), in which the sequestrated lobe arose from an oesophageal bronchial bud, lay posterior but unconnected to the left main bronchus, and received its blood supply from a branch of the left pulmonary artery.

Smith (1956) suggested that the primary lesion in intralobar sequestration was failure of the pulmonary artery in foetal life to supply the segment of lung involved. It is conceivable that failure of complete canalization of a branch of the right pulmonary artery in foetal life was responsible for sequestration in our case. The frequent association of cystic changes with an aberrant systemic arterial supply in the intralobar variety led to the suggestion that cyst formation resulted from high pressure arterial blood flow (Smith, 1956). There was no demonstrable systemic arterial supply in the present case, but one could postulate that it was present in foetal life (presumably from the right pulmonary artery) and subsequently became occluded. Although the pathogenesis of the occlusion is not clear, the elastic structure of the artery observed in the pedicle is similar to systemic aberrant arteries in intralobar varieties (Pryce, 1946; Baar and d'Abreu, 1949; Nielsen, 1964). The stimulus for the formation of such an artery presumably occurred in foetal life, thus supporting the suggestion that high-pressure arterial blood flow from the pulmonary artery might be responsible for the cystic change observed in the present case of extralobar variety. It would therefore appear that cystic change could occur irrespective of the type of sequestration.

There were no symptoms or signs referable to the cardiorespiratory system in our patient, a feature consistent with the extralobar variety, when respiratory function is not usually affected, though most cases are detected in early life. It is compatible with life, and there are a few cases, reported in elderly patients (Sjølde and Christiansen, 1938; Rektorzik, 1861). The variety encountered here should be amenable to surgical treatment without the risk of an anomalous vessel frequently encountered in the intralobar type.

The majority of sequestrated lobes are situated in the left side of the thorax, posteriorly and basally (Freedlander and Gebauer, 1939; Pryce, 1946; Valle and White, 1947; Findlay and Maier, 1951; Turk and Lindskog, 1961) and derive their blood supply from the thoracic or abdominal aorta, intercostal arteries, and aortic arch, in that
order of frequency. The venous drainage of the intralobar variety is usually to the pulmonary veins or occasionally to the vena cava, and of the extralobar variety to the hemiazygos and azygous vein, occasionally to the portal vein or to the suprarenal vein (Vogel, 1963). There are, however, a few recorded cases of the intralobar variety draining into the azygos venous system (Douglas and Shaw, 1964; Kergin, 1952), and the venous drainage in our case was into the azygos vein. Pulmonary sequestration may be associated with other congenital abnormalities, such as diaphragmatic defects, herniae, and foregut and tracheobronchial connexions, but none was observed here, and the cause of death was bacterial enterocolitis due to simultaneous Shigella and Salmonella infection, quite unrelated to the congenital abnormality.

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REFERENCES
Extralobar pulmonary sequestration

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