SIMULTANEOUS INTRA- AND EXTRAPULMONARY SEQUESTRATION*

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Pulmonary sequestration has attracted considerable attention in recent years. Many papers reporting typical cases have appeared in the literature. Repetition of material already extensively reported will not be attempted (Bariéty and Coury, 1947; Pryce, Sellors and Blair, 1947; Bruwer, Clagett and McDonald, 1950, 1954; Kergin, 1952; Santy, Bérard, Gaby and Huu (1952); Boyd, 1953; Hertzog, Toty, Personne and Gilbert, 1953; Razemon, Houcke and Razemon, 1953; Monod, 1954; Boyden, 1955, 1958; Bruwer, 1955; Pryce, 1946; Smith, 1956; Procházka and Steinhart, 1957).

The case described here is a very rare form of pulmonary sequestration, which justifies its publication.

Case Report

The patient was a 15-month-old boy (No. 1323/58) who at the age of 3 weeks developed pneumonia, seen in the radiograph as a pericardial infiltration above the diaphragm (Fig. 1). Despite the usual antibiotic therapy, it subsided surprisingly slowly (Fig. 2). A follow-up radiograph at 11 months of age showed a sharply defined elliptical shadow, the size of an egg, lying paravertebrally below the diaphragm and superimposed on the gastric bubble (Figs. 3 and 4). Following radiological examination, including upper gastro-intestinal barium studies, retroperitoneal air insufflation, bronchography and pyelography, the provisional diagnosis of retroperitoneal tumour, probably of neurogenic origin, was made. Surgery was advised after further regression of the pneumonitis.

The operation (Kafka, Brodský, Kudrnová, anaesth. Drapka) was performed on October 18, 1958. An oblique incision was made in the left epigastrium. The tumour was found to be on the posterior surface of the stomach (Fig. 5) on the fundus and on the cardia. It was a thin-walled cyst fixed flat against the stomach and was separated by sharp dissection. Its superior end continued as a narrower cylindrical cyst, lying on the oesophagus, right up through the hiatus. The lumen of this narrow cyst was separated from its gastric portion only by an internal membrane.

The oesophagus was pulled downwards and the cyst further dissected. On opening the cyst wall and inserting a probe, it was found that the cyst continued into the thorax in a dorso-latero-cranial direction. By prolonging the incision through the seventh intercostal space an additional thoracophrenotomy was performed. The cyst, or rather its stem, 10×12 mm. in diameter, diverged from the oesophagus to enter the dorso-basal segment of the left lower lobe. A large coiled artery ran into it, traversing the diaphragm and arising from the abdominal aorta. This was ligated and a resection of the dorsobasal segment, together with the entire cystic structure, was carried out (Fig. 6).

Specimen (Fig. 7). The resected specimen consisted of two parts. The superior portion was in the shape of a triangle, at the apex of which were bronchi, artery and vein running into the resected segment of the left lower lobe. On probing, the bronchus did not communicate with the cyst, which formed the posterior wall of the resected segment. The upper portion of the cyst was thick-walled, containing almost cartilaginous elements; its distal part narrowed into a thick stalk 6-7 cm. long, without interruption of the cyst lumen. An ovoid, thin-walled segment the size of an egg was situated on the inferior end.

Histology. (Žaková, Second Institute of Pathological Anatomy, Charles' University, Prague; Case No. 3164/58.) The wall was formed by two layers of smooth muscle fibres. Exterior to this was a layer of connective tissue showing elastic degeneration, and scattered seromucous gland. Ciliated cylindrical epithelium on a basal membrane formed the cyst lining (Figs. 8 and 9).

The post-operative course was uneventful; one year after operation the patient's condition now appears excellent.

Discussion

In the present communication we wish to outline certain characteristics of this syndrome: the simultaneous occurrence of intra- and extralobar sequestration, the subdiaphragmatic position of a part of the dumb-bell-shaped cyst, the blood supply and, finally, problems of diagnosis before and during operation.

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FIG. 1.—Admission radiograph showing consolidation of left lower pulmonary field.

FIG. 2.—Radiograph taken two months after that of Fig. 1, showing slow improvement of pulmonary condition following chemotherapy.

FIG. 3.—Bronchogram showing shadow below diaphragm superimposed on gastric bubble.

FIG. 4.—Bronchogram: lateral view showing shadow below diaphragm just in front of vertebral column.

FIG. 5.—At operation the tumour was found lying at posterior surface of stomach.

FIG. 7.—Excised specimen.

FIG. 9.—Section of cyst wall (high magnification).

FIG. 8.—Section of cyst wall.

FIG. 6.—Schematic drawing showing findings at operation: 1. Spherical cyst; 2. Tubular prolongation; 3. Left lower pulmonary lobe; 4. Bronchial artery.

Neither aetiology nor classification of this disease entity have been agreed upon (Gebauer and Mason, 1959), and the term sequestration itself does not seem to cover the case. Since this term has been widely applied as a result of publications by Pryce *et al.* (1947) and others, we shall continue to use their terminology. The term “foregut cyst” (Hössli, 1950; Roemer and Mollowitz, 1957) or ‘dysembryopathy’ (Bert and Fischer, 1910; Petříková, Polák and Stolz, 1952; Breton and Dubois, 1957) seems more accurate.

It is known that pulmonary sequestration is one of a group of developmental defects occurring at the site of differentiation of the gastro-intestinal and respiratory tracts from the foregut. It naturally follows that various structures in the lower thoracic area may be involved (Cole, Alley and Jones, 1951), particularly the lower lobe arteries (Cole *et al.*, 1951; Findlay and Maier, 1951; Fry, Arnold and Miller, 1953), mediastinum (Maier, 1948), pericardium (Ladd, 1936; Rusby and Sellors, 1945; Warner, Britt and Riley, 1958) and diaphragm (Cockayne and Gladstone, 1917; Raymond *et al.*, 1956).

The majority of cases present a relatively simple lobar form of pulmonary sequestration (Boyd, 1953; Bruwer *et al.*, 1954; Bruwer, 1955), often supplied by an atypical systemic artery, also referred to as a

bronchogenic cyst (Fry *et al.*, 1953; Gross, 1953), Flimmerepithelcysts (Zadek and Riegel, 1958), congenital lung cyst (Kergin, 1952), etc.

Extralobar sequestration has a more complex character. We find it termed ‘lower accessory lung’ (DeBakey, Arey and Brunazzi, 1950), pulmonary aberrations, supranumerary lung (Gans and Potts, 1950; Raymond, Hardy and Robbins, 1956; Boyden, 1958) located in the pleural cavity (Rusby and Sellors, 1945) as well as in the mediastinum (Maier, 1948) or elsewhere. Some authors do not make a genetic differentiation and maintain that these are quantitative variations of the same developmental defect (Bert and Fischer, 1910; Borovanský and Hněvkovský, 1929; Breton and Dubois, 1957).

The site of the lesion and its embryonic stage determine the degree of tissue differentiation and the location and nature of the malformation. Maier (1948) maintains, for example, that there is no formal differentiation between bronchogenic and enterogenic cysts.

Since the aetiology and exact origin of such developmental errors are still not clear, we do not feel qualified to comment upon them.

Only rarely do both extra- and intralobar sequestration occur simultaneously. And even more rarely is this combined with ectopic location of the extralobar part in the abdominal cavity or below the diaphragm. Cases of isolated ectopy are mentioned by Valle and White (1947) and Raymond *et al.* (1956). Very complex malformations may occur. Such a condition was found in our patient, where a cyst of the stomach and oesophagus was connected with a pulmonary cyst supplied by an anomalous artery arising from the abdominal aorta.

Mention must be made here of what Gross (1953) calls ‘duplications of the alimentary tract’. Two cases were presented with simultaneous occurrence of enterogenic cysts in thoracic and abdominal cavities. These should be considered in the differential diagnosis of extrapulmonary sequestration.

From the surgical point of view it is interesting that pulmonary sequestration with arterial anomalies was a problem some 10 years ago, as shown by reports of operative deaths or severe complications in relation (or without relation) to surgery. Fatal haemorrhage was reported by Harris and Lewis (1940), Butler (1947) and Douglass (1948)—see Fry *et al.* (1953). The extensive work of Pryce *et al.* (1947) and others acquainted thoracic surgeons with the problem involved, and today the literature contains more than 110 cases; over 100 have been reported by Bruwer (1955) and 11 cases from the Czecho-Slovakian literature may be added. This

does not include reports of simple bronchogenic cysts (without anomalous artery) with an intrapulmonary or mediastinal localization.

In most uncomplicated cases pre-operative diagnosis is not difficult. Repeated lung infection and a shadow on the radiograph in the region of the dorsobasal pulmonary segment pointing caudally to the midline indicate the lines along which further investigation should be carried out (Wyman and Eyler, quoted by Bruwer (1955)).

There are, of course, circumstances which complicate the diagnosis. These are in the first place inflammatory processes, including lung abscess and tuberculosis, or non-inflammatory processes, such as atelectasis, bronchiectasis, etc. In the second place, difficulties may be encountered in differentiation from pulmonary or extrapulmonary neoplasms

(Barthel, 1958). Cases of pulmonary sequestration treated as tuberculosis, even including thoracoplasty, or as pneumothorax from a burst lung abscess have been reported (Table 1).

Even in the course of operation to make the correct diagnosis is not always simple or even possible. Fry *et al.* (1953) admit having performed an operation, supposedly for an enterogenic cyst, without being directed to the right conclusion even by the discovery of an extension through the diaphragm. Only histological examination revealed that it was a case of a pulmonary sequestration.

Similarly, in our case pulmonary sequestration was not suspected, in spite of the finding of a gastric cyst, until probing of the oesophageal cyst revealed its true extent.

TABLE 1
PULMONARY SEQUESTRATION TREATED SURGICALLY IN CHILDREN UNDER 20 YEARS

Author	Cases (no.)	Sex	Age (yr.)	Site	Clinical Diagnosis	Anomalous Artery Derived from:	Operation
Rusby and Sellors (1945)	1	F	19	Left extralobar	Pulmonary cyst or tumour of mediastinum	Pulmonary or bronchial artery	Extirpation
Ravitch and Hardy (1949)	2	M M	12½ 6	L.L.L. R.L.L.	Infected cyst " "	Thoracic aorta (3) Intercostal artery	Lobectomy Lobectomy
Gans and Potts (1950)	1	F	5/12	Left extralobar communication to oesophagus	Oesophageal diverticulum	Pulmonary artery	Extirpation
Hössli (1950)	1	M	5	L.L.L. communication to stomach	Pulmonary abscess		Lobectomy
DeBakey <i>et al.</i> (1950)	1	F	7	L.L.L.	Pulmonary cyst (dermoid)	Thoracic aorta	
Cole <i>et al.</i> (1951)	1	M	3	L.L.L.	Cyst	Thoracic aorta	Lobectomy
Kergin (1952)	3	M F F	17 19 16	L.L.L. L.L.L. R.L.L. Intra- and extralobar	Cystic disease Infected cyst Intralobar sequestration	Thoracic aorta Subdiaphragm Intercostal artery	Lobectomy Lobectomy Lobectomy
Boyd (1953)	6	M M F F F M	7 5 4 13 9½ 6	R.L.L. R.L.L. L.L.L. L.L.L. L.L.L. L.L.L.	Tuberculosis Pulmonary affection " " " " " " " "	Intercostal artery Subdiaphragm Thoracic aorta " " Subdiaphragm "	
Fry <i>et al.</i> (1953)	1	M	19	R.L.L.	Pulmonary cyst (abscess)	Subdiaphragm	Lobectomy
Čermák, Váňa and Rozhold (1955)	2	F F	6 14	R.L.L. R.L.L.	Bronchiectasis ?	Subdiaphragm Thoracic aorta (3)	Lobectomy Lobectomy
Smith (1956)	1	M	18	L.L.L.	Bronchiectasis	Thoracic aorta	Lobectomy
Procházka and Steinhart (1957)	4	M M M F	6 5 6 7	L.L.L. L.L.L. R.L.L. L.L.L.	Tumour or pulmonary cyst Vulvular pneumothorax Sequestration Cyst (Tb empyema)	Thoracic aorta Subdiaphragm Thoracic aorta	Segmental resection Lobectomy Lobectomy Thoracoplasty else- Lobectomy [where
Warner <i>et al.</i> (1958)	2	F M	3 7½	Intrapulmonary Extrapulmonary (pericardial defect)	Cyst Mediastinal cyst	Subdiaphragm (4) Pulmonary and bronchial artery	Lobectomy Extirpation
Kafka and Beco	1	M	1½	L.L.L. and extralobar (sub-diaphragm)	Neurogenic tumour	Subdiaphragm	Segmental resection and extirpation (thoracophrenolaparotomy)

Conclusions

The history of our case clearly shows signs typical of pulmonary sequestration, which together with the type of infiltration shown by radiography in the left lower lobe should have directed us towards a correct diagnosis.

The misleading element was the finding of an abdominal tumour in a paravertebral position. This and its shape appeared to indicate a ganglioneuroma.

Tomography and retroperitoneal insufflation were not helpful, since the tumour was in fact partly situated in the retroperitoneal space.

Barium meal showed no pathological changes. On bronchography three branches of the dorsobasal bronchus did not fill right up to their periphery, which was considered incidental to the suspected postpneumonic condition.

For these reasons operation was started with a diagnosis of retroperitoneal tumour, a neurinoma, and no connexion was at first seen between the two conditions, pulmonary and abdominal.

In the course of operation we were surprised to find that (1) the mass was not a ganglioneuroma, (2) it was not wholly retroperitoneal, arching into the abdominal cavity and connecting with the stomach, and (3) it had penetrated through the hiatus along the oesophagus into the thoracic cavity.

Only probing the cyst lumen revealed that the cyst continued high up into the thorax, and the approach was widened into a thoraco-phrenolaparotomy. It then became clear that this was a case of pulmonary sequestration. The anomalous artery could be severed and the entire cystic mass was removed.

Summary

A rare case of dysembryoplasia (foregut cyst) in the region of the respiratory and upper alimentary tracts is described, falling under the heading of pulmonary sequestration as known in the literature.

Thoracophrenolaparotomy in a boy of 15 months revealed connecting masses of cysts, localized in the left inferior lobe (intralobar sequestration with an aberrant artery), passing to the oesophagus (extralobar pulmonary sequestration) and through the hiatus to the stomach (forming the subdiaphragmatic part of the tumour).

The entire cystic complex was removed.

Diagnostic errors and special anatomical conditions in the described case are pointed out. The discussion is supported by a Table covering exclusively surgical cases in children.

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