

INTRADIAPHRAGMATIC MESOTHELIAL CYSTS

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Primary tumours and cysts of the diaphragm are rare. In a review of the literature Clagett and Johnson (1949) were able to record 30 cases, to which they added four of their own. Of these cases, 18 were malignant and 16 were benign.

Primary cysts constituted only a very small proportion of the cases, and in recording a case of mesothelial cyst of the diaphragm Aufses and Oseasohn (1949) stated that they were only able to find records of five other primary cysts in the literature.

We have recently encountered a case of mesothelial diaphragmatic cyst; a second almost identical case which was under the care of Mr. Geoffrey Flavell, of the London Hospital, is also reported by his courtesy.

CASE REPORTS

Case 1.—W. B., a man aged 41, was radiographed when a mass radiography unit visited the factory in which he worked. He had no complaints. Radiographs (Figs. 1 and 2) revealed a small rounded opacity associated with the antero-medial part of the right diaphragm. A diagnostic pneumoperitoneum was carried out as it was thought that the most likely diagnosis was a small liver hernia, and this demonstrated clearly (Fig. 3) that the lesion was in or above the diaphragm, the former appearing more likely, as the outline of the diaphragm appeared to enfold the tumour. As no precise diagnosis could be established, thoracotomy was advised and accepted.

On October 31, 1950, the right chest was opened through the bed of the seventh rib. The pleura was completely free of adhesions. In the anterior part of the diaphragm about two inches from the anterior chest wall and from the pericardium lay a tense, bluish cyst, the size of a hen's egg, covered by diaphragmatic pleura and having muscle fibres of diaphragm splayed out over it. The overlying pleura was incised and the cyst shelled out without difficulty, leaving a defect some 2 in. in diameter in the diaphragmatic musculature, in the floor of which intact peritoneum was visible. This defect was closed with interrupted silk sutures and the chest closed without drainage. The post-operative course was uneventful, the patient being discharged home on the tenth day.

Dr. G. R. Osborn reported that the cyst from the diaphragm measured about 2 cm. in diameter. Microscopically it was of the simple chylous type; the lumen was lined with a single layer of endothelial cells which occasionally had rudimentary papillary projections. Outside this there was a little areolar connective tissue and then skeletal muscle fibres from the diaphragm. It was not a true tumour.

Case 2 (Mr. G. Flavell's case).—A woman, aged 49, was admitted to the Ear, Nose, and Throat Department of the London Hospital. Ten months before admission she had had a severe cough lasting two weeks, accompanied by laryngitis; during this attack she lost her voice, which returned gradually to normal. Laryngoscopy revealed a papillomatous area in the interarytenoid region from which a biopsy was taken. The

FIGS. 1 and 2.—Radiographs showing small rounded opacity associated with the antero-medial part of the right diaphragm in Case 1.

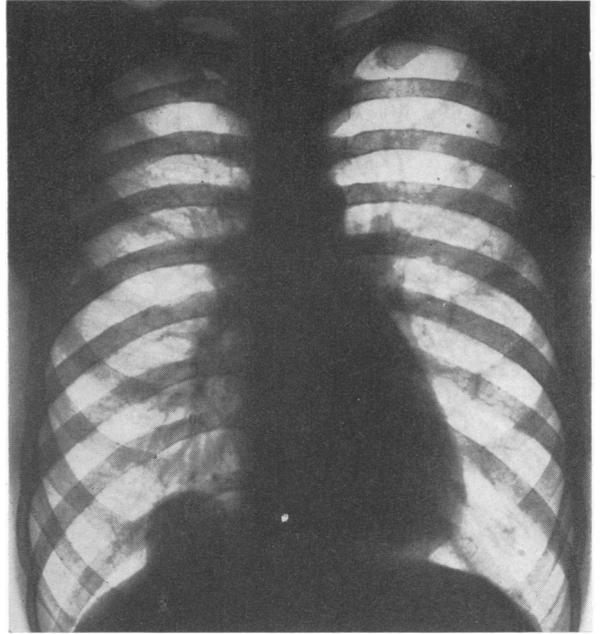


FIG. 1.

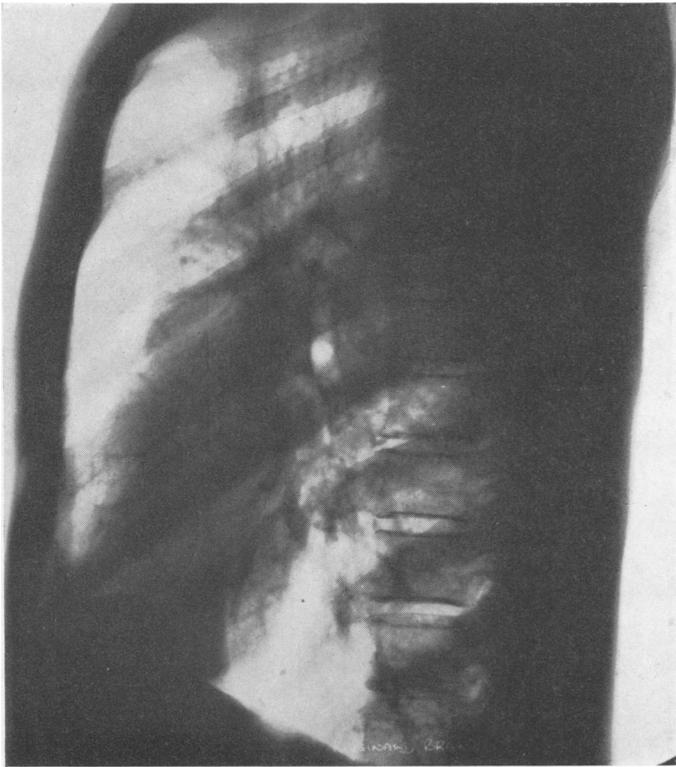


FIG. 2.

FIG. 3.—Radiograph after the induction of diagnostic pneumoperitoneum.

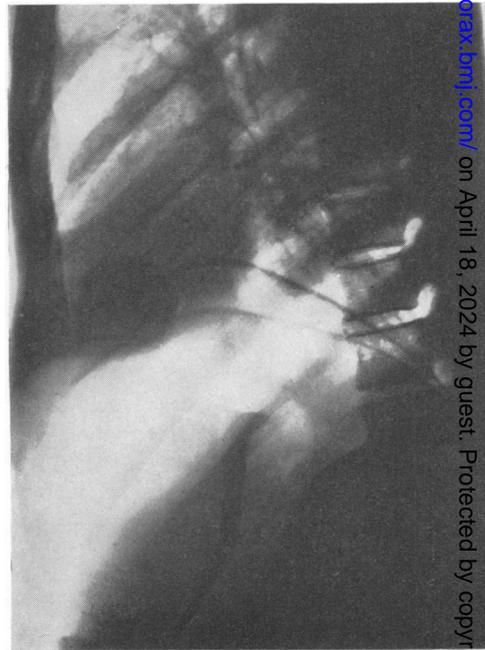


FIG. 3.

report concerning this tissue stated that there was hyperplasia and chronic inflammation, but no neoplasm.

She had slight dyspnoea on exertion, but no other chest symptoms.

A radiograph revealed a rounded opacity in the region of the left anterior cardiophrenic angle. Thoracotomy was advised and accepted (Figs. 4 and 5).

On July 3, 1950, the chest was opened through the bed of the second rib and a multilocular cyst about 3 in. in diameter was found completely submerged in the antero-medial part of the left dome of the diaphragm presenting from above an appearance like a Byzantine dome. The upper surface was covered by muscle fibres of the diaphragm itself stretched over the cystic area. To free the cyst it was necessary to incise the diaphragm so that when the cyst was removed a substantial hole was left communicating with the peritoneal cavity. The cyst itself presented about equally into the pleural and peritoneal cavities, so that the upper surface was covered by pleura and the lower surface by peritoneum.

The histological report described "a cyst from the left diaphragm lined by a single row of flattened cells and containing voluntary muscle in its wall."

The patient made an uninterrupted recovery.

DISCUSSION

The rarity of primary diaphragmatic cysts is evidenced by the few reports in the literature. There seems little doubt that with the increasing use of mass radiography more cases will come to light.

In a careful search through the literature we have been unable to discover any case corresponding to the two now reported. Aufses and Oseasohn (1949) described an example which occurred in a woman aged 42 who had for three months a dull, aching pain in the chest. At thoracotomy a cyst was found in the right cardiophrenic angle with fibres of the phrenic nerve crossing over its surface. Histologically the cyst had a thin fibrous wall with flattened epithelial lining and some lymphatic tissue in the wall. They described it as a "mesothelial cyst of the diaphragm."

Nylander and Viikari (1948) described three cases; all occurred in men and all were upon the right side. Their first patient had complained of indefinite chest pain, and they describe the cyst as "attached to the diaphragm by a fairly broad pedicle and slightly to the pericardium too." Their second patient had a large cyst "slightly adherent to the pericardium and widely to the diaphragm in its medial part. The cyst obviously originated in the anterior part of the diaphragm close to the pericardium." In their third case "a plum-sized translucent tumour projected from the upper surface of the diaphragm, located medially at about 2 cm. from the ventral insertion of the right half of the diaphragm: it was fairly easily detached from the musculature of the diaphragm."

Histologically all three cysts showed a fibrous wall and endothelial lining. The authors concluded that the cysts arose from a remnant of the infracardiac bursa.

Pickhardt (1934) described a "pleuro-diaphragmatic cyst" occurring in the left anterior cardiophrenic angle. It was a bilocular cyst with a thin wall of fibrous tissue and an endothelial cell lining. It does not appear to have had any close relation to the diaphragm, and the description is that of what we would now call a pericardial coelomic cyst.

Clagett and Johnson (1949) reviewed the literature on diaphragmatic tumours and added a case of their own, a right-sided diaphragmatic cyst with calcification

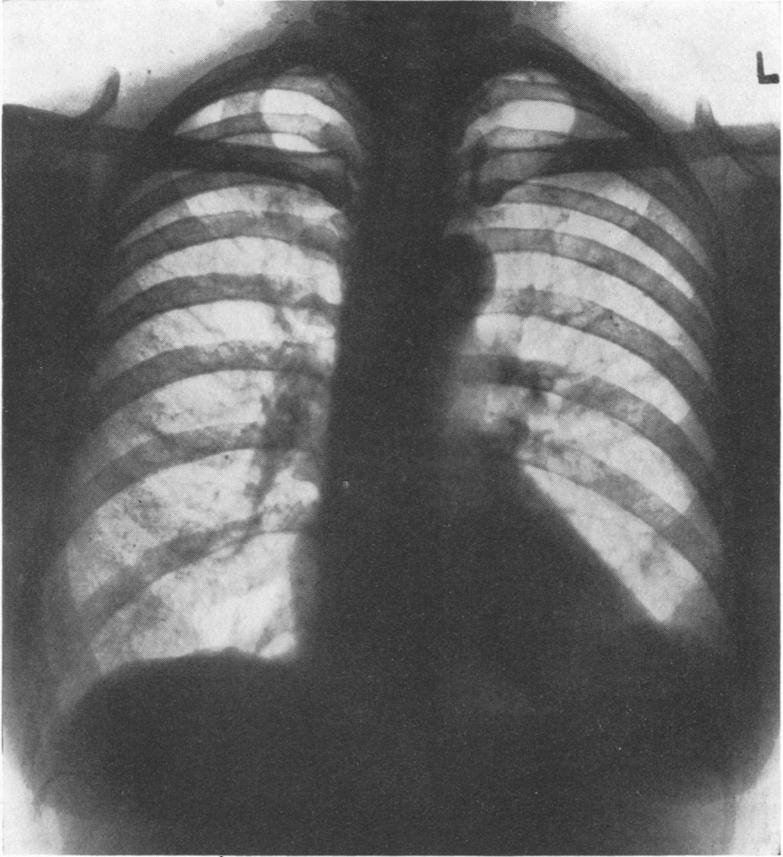


FIG. 4.

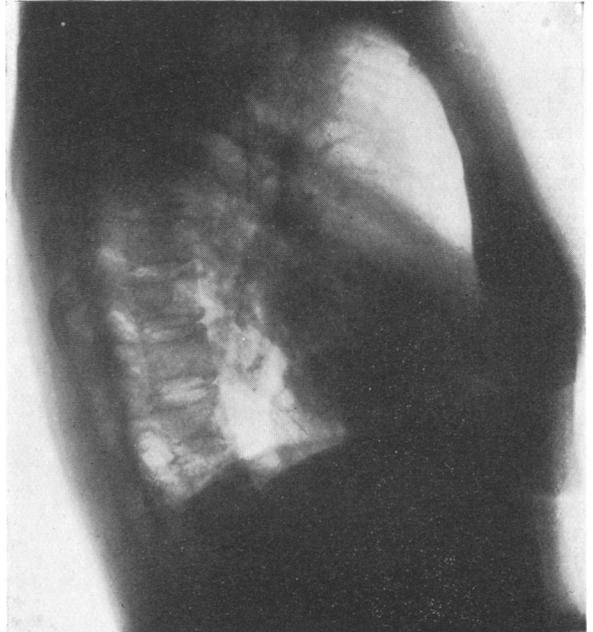


FIG. 5.

FIGS. 4 and 5.—Radiographs showing a rounded opacity in the region of the left anterior cardiophrenic angle in Case 2.

in its wall, which, when removed, left a defect in the diaphragm. Histologically it was a "fibrous walled cyst containing putty-like material."

Scott and Morton (1946) described a cyst in the dome of the left diaphragm containing chocolate-coloured fluid with cholesterol crystals and having a fibrous wall, calcified in places. It was lined with a single to double layer of flat to cuboidal epithelial cells.

In the case of Robson and Collis (1944) a fibrous-walled cyst lined by granulation tissue was thought to be tuberculous.

DIAGNOSIS AND TREATMENT

Both the cases now described were first discovered by a routine radiograph.

Some of the cases described in the literature had symptoms of which pain and dyspnoea were the most frequent.

The finding of an opacity in the region of the diaphragm will naturally lead to a full investigation. Consideration should be given to the possibility of diaphragmatic hernia and a barium examination of the alimentary tract should be carried out, including a "follow-through" examination: if this investigation is omitted an anterior costosternal (foramen of Morgagni) hernia may be missed. The upward and anterior displacement of the transverse colon is characteristic.

Pneumoperitoneum is a valuable diagnostic measure, and will exclude the possibility of a liver hernia. Its use in relation to diaphragmatic neoplasms was described by Binney (1931) and Söderlund (1937); in our first case it demonstrated the intradiaphragmatic position of the cyst quite clearly.

Artificial pneumothorax has been employed by some authors (Pickhardt, 1934, and Nylander and Viikari, 1948). Pickhardt also employed thoracoscopy as a diagnostic measure.

In the majority of instances the diagnosis can only certainly be made by exploratory thoracotomy, and this should be carried out in every case. The morbidity of such a procedure is minimal in skilled hands. In a proven diaphragmatic "tumour" the fact that more than half the recorded cases have been malignant (Clagett and Johnson, 1949) provides an extra argument (if such is required) in favour of operation.

EMBRYOLOGICAL HYPOTHESES

It will be observed that our two cases differ from any others in the literature (except possibly the third case of Nylander and Viikari) in that both were situated in the substance of the diaphragm and had muscle fibres splayed out over them. Their situation was identical, although one was right-sided and the other left. From their structure, there can be no doubt that both were of congenital origin.

Intradiaphragmatic Cysts.—The situation of these cysts within the substance of the diaphragm and lying above intact peritoneum suggests the following embryological origin (Fig. 6). In their early development the pericardium and peritoneum are connected by the pleuro-peritoneal canal, this canal being defined by partial rims at the pericardial end (duct of Cuvier) and at the peritoneal end (pleuro-peritoneal fold). The lung bud appears on the medial wall of this canal between these rims and, growing laterally and away from the medial edges of these rims,

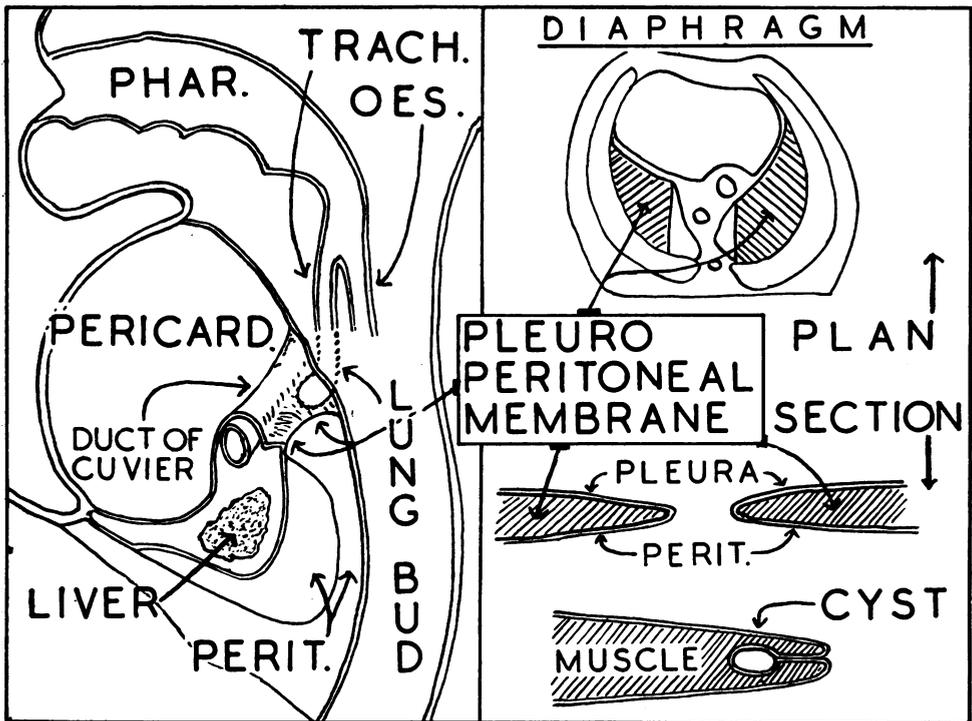


FIG. 6.—Diagram to illustrate the significance of the pleuro-peritoneal membrane in relation to intradiaphragmatic cysts. (Modified from Hamilton, Boyd, and Mossman, 1945; *Gray's Anatomy*, 1944.)

develops two membranes, the pleuro-pericardial and pleuro-peritoneal. The latter membrane, with which we are now concerned, stretches across the lower end of the canal and eventually assists in sealing it off and so separating the pleural and peritoneal cavities; it persists as part of the diaphragm which is thus produced by a “folded” membrane with mesothelium on its upper and lower surface. Muscle appears within this fold at an early stage. The cysts, being in the substance of the diaphragm and having peritoneum below, presumably developed within this double membrane. There are two possibilities: first, that fluid collected between the double layer which was otherwise normal, but the difficulty here would be to explain how secretion of fluid occurred as the mesothelium would, of course, be on the outside of such a cyst. The second—and more likely—explanation is that an invagination occurred in the free crescentic margin of the membrane at the time when it was fusing with the neighbouring structures, and that from this invagination the cyst developed. Incidentally, invagination of this type could not occur far from the sharp crescentic margin and such a cyst must therefore lie somewhere along the line of the margin.

Mesothelial Cysts.—If we accept that the cyst arose by such a means, the interest lies in the fact that it forms but one of a series of cysts which may arise from various parts of the mesothelial membrane lining the three coelomic cavities—cysts which

are generally called mesothelial or coelomic cysts, the commonest variety being the pericardial or costophrenic angle cyst. It becomes useful, therefore, to review the several theories which have been advanced to account for these cysts (Fig. 7).

TEXT	DESCRIPTION	PROCESS	RESULT	EXAMPLES
1	PERSISTENCE OF LACUNAE	LACUNAE MESENCHYME	CYST PERICARD.	IRREGULARLY SITUATED SOME PERICARDIAL
2	CLOSURE OF DIVERTICULUM	ATRIUM VENTRAL DIVERTIC.	CYST	PERICARDIAL
3	PNEUMATO-ENTERIC RECESS	P.E.R. BURSA DIAPH. OMENTUM		SOME RIGHT-SIDED PARA-OESOPHAGEAL
4	FOLDING DURING CLEAVAGE	COSTO-PHRENIC ANGLE	CYST	COSTO-PHRENIC UNRELATED TO PERICARDIUM
5	JUXTAPOSITION OF SURFACES	DORSAL LATERAL MESENCHYME	CYST	ATTACHED TO (OR WITHIN) THE DIAPHRAGM (POSTERIORLY)
6	INVAGINATION OF SURFACE	FREE CRESCENTIC EDGE PLEURO-PERIT. MEMB.	CYST	SOME INTRA-DIAPHRAGMATIC

FIG. 7.—Diagram showing ways in which mesothelial coelomic cysts may arise (modified from various authors).

1. One of the primitive mesenchymal clefts or lacunae, which normally fuse to produce pericardial coelome, does not do so and so persists as a separate cavity or cyst (Lambert, 1940).

2. An embryological ventral diverticulum (of the pericardium) becomes sealed off and so results in a cyst (Lillie, McDonald, and Clagett, 1950).

3. The closure of the diaphragm *normally* isolates the supra-diaphragmatic end of the pneumato-enteric recess (a right-sided para-oesophageal transdiaphragmatic diverticulum of the greater omentum) to produce the infracardiac bursa, a closed mesothelial-lined space which may develop into a cyst (Nylander and Viikari, 1948).

4. During the expansion of the margins of the pleural cavity by cleavage of the mesenchyme, irregularities or infoldings occur in the advancing "edge" of the pleural space, these giving rise to cysts (Kindred, quoted by Drash and Hyer, 1950).

5. In the final closure of the pleuro-peritoneal and pleuro-pericardial openings (which may, of course, persist and produce congenital defects in the diaphragm or

pericardium respectively) mesothelial surfaces are approximated, and in this process a loculus may be produced and so initiate a cyst. (It should be noted that two *different* mesothelial surfaces are here involved.)

6. In this same process of closure and fusion invagination of the free edge of a membrane may occur, as in the present cyst, where only one single mesothelial surface was involved.

Thus it becomes possible to systematize the aetiological factors involved in the production of these cysts if we accept that all arise from vagaries in the development of the mesothelial surfaces lining the primitive coelomic cavity. Which of the six mechanisms is invoked as explanation will depend mainly on the anatomical situation of the cyst, and on the relationship of the cyst to the mesothelial surface of origin; the relative frequency with which these different causes operate will be proportional to the probability of the particular embryological abnormality arising. As regards the relationship of the cyst to the mesothelial surface, it will be appreciated that, while in the histological sense cysts always lie *outside* the cavity from which they arise, anatomically they may appear to lie *within* the cavity of origin: for example, a micro-cyst arising outside the parietal pleural cavity during the process of mesenchymal cleavage may subsequently, with growth, raise the parietal pleura from the chest wall and hence lie inside the pleural space.

Nomenclature.—The value of a clear picture of the aetiology is that we are then in a better position to consider the nomenclature which is most appropriate for these cysts; in this connexion the following facts are important. (1) The cysts are mesothelial. (2) The cysts are coelomic. (3) The cysts are related to the pericardium, peritoneum, or pleura, and may be further classified as visceral or parietal. (4) The cysts have a definite anatomical location. (5) The cysts arise by one of several embryological abnormalities. Categories (1) and (2) define the class; (3) and (4) define the site; (5) defines the aetiology. Clearly we cannot include all these considerations in a brief descriptive name for each cyst. In any given case our knowledge may be partial or hazy on some of these five points. There will, however, usually be two pieces of factual information: (1) histological evidence that the cyst is mesothelial, and (2) the anatomical location of the cyst. It is suggested that this combination should serve as a “conventional minimal title” for cysts which are considered to belong to this series; for example, “mesothelial cyst of the diaphragm”; “mesothelial pericardial cyst”; “mesothelial costophrenic angle cyst.” Supplementary information covering the residual three points can be given in the accompanying description. A further advantage of this “conventional minimal title” is that it is purely descriptive and therefore free of inaccurate or uncertain implications regarding aetiology. Titling in terms of the cyst content is generally unsatisfactory; for example, the term “springwater cyst” has been applied both to those cysts lined with columnar ciliated epithelium and with mesothelium, that is to cysts arising by entirely different embryological processes from entirely different embryological structures (bronchiogenic; coelomic); and the term “serous cyst” may well include mesothelial cysts, columnar ciliated bronchial cysts, and lymphangiomatous cysts.

SUMMARY

Two cases of intradiaphragmatic mesothelial cysts are presented and the literature is reviewed.

The various embryological theories advanced to explain the origin of these cysts are collected and systematized.

The most appropriate nomenclature for cysts of the mesothelial-coelomic series is briefly discussed.

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