ENTEROGENOUS CYST OF THE MEDIASTINUM

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Enterogenous cyst of the mediastinum is not a common condition. According to Bickford (1949) 40 cases have been reported. Of these, 24 were lined by purely gastric mucosa (this includes the case reported by him), four had purely intestinal epithelium and 12 were composite.

This paper is presented as a further instance of an enterogenous cyst which was removed successfully at thoracotomy.

Case History

The patient was a baby girl born of a healthy mother who had had a normal pregnancy. The baby was admitted when aged 3 months, on March 1, 1950, because of bronchitis, which had been present for one week.

Examination revealed cyanosis and distressed respirations, with diminished expansion of the chest over the right base, and a diminished percussion note. A diagnosis of bronchopneumonia was made. Mantoux tests were negative. Radiographs (Figs. 1 and 2) showed a large oval tumour lying posteriorly in the upper two-thirds of the right side of the chest. From time to time some stridor was noted. Her weight gain was not satisfactory.

Mr. F. Ronald Edwards advised thoracotomy for a possible gastrogenic cyst of the mediastinum or possibly ganglioneuroma.

On May 24, 1950, the chest was opened through the bed of the right sixth rib. In the posterior mediastinum was a tensely cystic swelling 4 cm. x 2 cm. x 1.5 cm. A few millilitres of clear fluid was aspirated. This later was analysed as clear mucoid fluid with no trypsin, pepsin or free hydrochloric acid. The chloride content was 700 mg. %o. The cyst was excised and the chest closed without drainage. There was a small extra posterior upper lobe segment. On June 3, 1950, the baby became dyspnoeic and cyanosed. She had signs of an effusion on the right side of the chest and 160 ml. of milky alkaline fluid was aspirated. This fluid did not recur and she was discharged on June 30, 1950. Since that time she has continued to gain weight and the chest has remained clear.

Section of the cyst showed a thick muscular wall (Figs. 3 and 4) of smooth muscle divisible into two layers running at right angles to each other. There was a thin muscularis mucosae on which there was a thin zone of coiled tubular epithelium. Finally there was a layer of tall columnar epithelium. Oxyntic cells were absent. The mucosa was enterogenous but not typically gastric.

FIG. 1.—Rounded tumour on the right side of the chest.

FIG. 2.—Lateral film showing the rounded tumour in the posterior mediastinum.
The fluid removed from the chest contained large numbers of lymphocytes and many fat globules.

Incidence

These cysts may be found on either side of the chest, but a little more commonly on the right (1·5 to 1) (Olenik and Tandatnick, 1946).

Males are affected a little more often than females (1·5 to 1) (Olenik and Tandatnick). It is predominantly a condition of young children. Eighteen cases occurred under 4 years of age (Steele and Schmitz, 1945). A case has been described in which the patient was a 15-year-old girl (Steele and Schmitz), another a 10-year-old girl (Dickson, Clagett and McDonald, 1946) and the oldest appears to be that of a 54-year-old man (Adams and Thornton, 1943). The cysts are invariably confined to the chest. One extended below the diaphragm (Dickson et al.) and three thoracic cysts described by Gross, Neuhauser and Longino (1951) were found to be intestinal diverticula arising in the abdomen. Truly thoracic cysts may, however, be associated with intra-abdominal cysts (Poncher and Milles, 1933).

Pathology

The cysts vary in size, shape and position in the chest. They may be as small as 2×3×2 cm. (Ward and Krahl, 1942) or as large as 14×20×5 cm. (Black and Benjamin, 1936). They are usually unilocular but may be multilocular (Ward and Krahl). The wall consists of two muscle layers placed at right angles to each other. There is a muscularis mucosae. The lining may be of gastric, intestinal or bronchial epithelium, singly or all found in the one specimen. The lining membrane may be atrophied and thus unidentifiable. When gastric mucosa is present there may be peptic ulceration with haemorrhage into the cyst or perforation of its wall. The blood supply of the cyst is derived from local vessels. Specially stained sections of the wall show the presence of ganglion cells (Böss, 1937; Seydl, 1935). The contents may be viscid, opalescent, milky or haemorrhagic. Davidson and Brown (1947) analysed the fluid from a cyst; the cholesterol content was 10 mg. %, protein 1·6 g. %, pH 7·8, and there was no free hydrochloric acid or pepsin.

Ladd and Scott (1944) analysed one specimen and showed that the cyst contained 360 ml fluid. The reaction was acid, the specific gravity was 1010, chlorides 114 mg. %, bicarbonate 34 volumes %, and cells 560 per ml. (80% polymorphs). In a second case they withdrew 10 ml. of thick, viscid fluid containing 700 cells per ml.

Symptomatology

It must be realized that there may be no symptoms at all, and the condition is found on routine radiology (Steele and Schmitz). The symptoms may be due to the swelling itself, to local pressure effects, to complications within the cyst or to all three. There may be general systemic disturbances.

Symptoms Due to the Swelling. These are not common but Ward and Krahl in a report on a child aged 9 weeks remark that the mother noticed a
progressive enlargement of the right side of the chest.

Symptoms Due to Pressure Effects. The main structure affected in this way is the trachea. Stridor, respiratory wheezing and a brassy cough may be present. Cyanosis and dyspnoea and signs of bronchitis, bronchopneumonia, atelectasis and bronchiectasis may be present as secondary manifestations of the presence of the cyst. Erosion of the vertebral bodies and ribs may occur (Steele and Schmitz, 1945; Ladd and Scott, 1944).

Symptoms Due to Complications in the Cyst. These are due to the occurrence of peptic ulceration and its associated sequelae. Perforation into adjacent structures gives rise to well marked symptoms and signs. Thus haemoptysis, pneumothorax or empyema may develop.

General Symptoms. There may be failure to thrive, feeding difficulties, unexplained anaemia, and some babies have attacks of screaming and drawing up of the legs.

Investigation
Radiological examination of the chest is the most satisfactory way of establishing the diagnosis. There is a rounded swelling in the chest. There may be tracheal or bronchial compression from behind forwards (Lindquist and Wulff, 1947) and radiological signs of lung compression with or without infective changes. Wyllie and Pilcher (1943) have stated that the radiological signs are those of (a) a space-occupying lesion, (b) some degree of collapse of the lung, (c) obstructive emphysema, and (d) a picture suggestive of a pleural effusion, with, however, a clear costophrenic angle or a rounded contour of the angle. They also stress that good lateral films are essential to show the position of the cyst in the posterior mediastinum. The details of the cyst shadow may be obscured by secondary changes in the lungs or by pleural fluid (Ladd and Scott, 1944). Rarely, erosion of the ribs and vertebral bodies may be seen (Mixer and Clifford, 1929; Ladd and Scott).

Cassel, Cunningham and Weisel (1950) report in a man aged 24 a subcarinal widening with elevation of the right main bronchus on bronchoscopy. When the cyst is large bronchoscopy is of little value, but when small the clinical picture may resemble that associated with an inhaled foreign body or tuberculous tracheo-bronchial glands, and here bronchoscopy is essential (Wyllie and Pilcher). They also stress that tuberculosis must be excluded.

Paracentesis of the cyst has been performed. Ward and Kralh withdrew 15 ml. of thick fluid resembling old blood. Wyllie and Pilcher aspirated 10 ml. of blood-stained fluid which had a deposit of serosal cells and some ‘fat’ phagocytes. This mode of examination is deprecated by Lindquist and Wulff. They maintain that the nature of all mediastinal cysts should be proved by thoracotomy. Wulff operated on a child with a so-called cyst and found a large cavernous haemangioma. The cyst may, however, be aspirated if it is causing acute suffocation (Lindquist and Wulff).

Treatment
The general opinion prevailing is that thoracotomy should be performed and the cyst excised (Cassel et al., 1950; Adams and Thornton, 1943; Bickford, 1949).

Ladd and Scott maintain that extirpation may be difficult because of adhesions, especially where there has been peptic erosion in the cyst. They advise marsupialization of the cyst and destruction of the lining membrane. They showed in one case that the cyst and the oesophagus had a common wall. In the operation reported here the dissection was performed under clear vision without any undue difficulty.

Theories of Causation
The aetiology of the condition has aroused much speculation. Although six possibilities have been put forward by Cassel et al., three main theories should be considered.

One is that the cyst is due to a persistent foetal diverticulum. This is based on the work of Lewis and Thyng (1908), who have demonstrated these cysts in the pig, rabbit and human embryos. Ladd and Scott point out that diverticula resembling those described by Lewis and Thyng are rare in early post-natal life and also that they are most common in the region of the duodenum, whereas alimentary duplications are most common in the lower jejunum and liver.

The second theory postulates that the cysts are derived from omphalo-mesenteric remnants. This is based on the writing of Fitz (1884). The theory does not explain the occurrence of alimentary cysts down the gastro-intestinal tract towards the rectum and has difficulty in explaining how these cysts in the abdomen derive their blood supply.

The other theory is based on Keith’s (1948) assertion that alimentary atresia and duplication are analogous. He proposes that this condition is due to an error in vacuolation in which there is a failure of coalescence of the vacuoles in the transverse diameter.

Summary
A case is reported of a 3-month-old baby in whom a cyst, lined by intestinal mucosa, was removed.
from the mediastinum by operation. Convalescence
was complicated by a chylothorax which was cured
by a simple aspiration. The features and the litera-
ture of mediastinal gastro-enterogenous cysts are
reviewed.
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