CYSTIC PNEUMATOSIS OF THE LARGE INTESTINE

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Cystic pneumatosis of the intestine is a rare condition, but perhaps not so rare as the small number of reported cases suggests. Although recognized since 1876, when Bang first noted gas cysts in the intestine of a woman who had died of volvulus, cystic pneumatosis of the intestine has remained a baffling diagnostic problem.

The condition may be defined as the presence of gas in cyst-like formations in the body. These may be localized in any part of the gastro-intestinal tract; they have a marked predilection for the ileocaecal region and the duodenum, but are also found in the bladder, vagina, mesentery, parietal peritoneum, and pleura.

Incidence and Distribution.—Cystic pneumatosis is more common in men than in women. It occurs at all ages, but mainly between 25 and 50. About 200 cases have been reported in the literature (Dressler, 1939; Sauser-Hall, 1940). Ferrandu in 1935 collected 180 cases, perhaps not all proven. Jackson in 1940 published a survey and added a case of his own, bringing the total of cases in the available literature to 172. Of these, twelve occurred in children and 160 in adults.

Aetiology.—The aetiology of cystic pneumatosis is obscure and none of the theories advanced is entirely convincing. Theories of a neoplastic or a chemical origin of cystic pneumatosis can be disregarded. The clinical picture of the reported cases is also unlike that of a gas-forming infection, the tissue reactions are not suggestive of an inflammatory condition, and bacterial cultures are usually negative. Animal experiments (Sauser-Hall, 1940) have produced no evidence in support of a bacterial origin.

The mechanical hypothesis postulates a break in the intestinal mucosa through which gas is forced into the intestinal wall. The defect can be caused by a localized infection or over-distension of the intestine by gas. There is some evidence in support of this view. In a large proportion of cases cystic pneumatosis is associated with ulcerative diseases of the gastro-intestinal tract, with obstruction and hyperperistalsis—for example, duodenal ulcer and pyloric stenosis, intestinal tuberculosis, chronic ileus caused by adhesions, etc. This theory, however, fails to explain the generalized forms, and the fact that not all people with pyloric stenosis develop cystic pneumatosis. Dressler (1939) suggests that there may be a constitutional weakness in the walls of the lymphatics which enables a comparatively slight rise in the abdominal pressure to cause a dilatation of the lymphatics.

Pathology.—The gross pathology depends on the extent of the gaseous infiltration. As a rule the bowel is covered with grape-like cysts varying in size, shape, and number. They can be localized in all strata of the intestinal wall, usually in the submucosa and in the subserous tissue. Moore (1929) says that in children gas is formed mainly in the mucosa and submucosa, but in adults the gas accumulates in the subserosa. Microscopically the cysts are separated by loose connective tissue lined by endothelial cells resembling the endothelial cells of the lymphatics; or they may show no lining at all. Giant cells of the foreign-body type are a fairly common feature. The cysts may be empty or may contain some serous material. Haemorrhage, oedema, and eosinophil cells have been described in the interstitial tissue.
FIG. 1.—Under the intact mucosa the submucosa is transformed into a system of numerous cyst-like cavities resembling emphysema of the lungs. × 20.

FIG. 2.—The cavities are lined by epithelial and numerous giant cells of foreign-body type, which can also be seen lying freely in the loose connective tissue separating the cavities. × 80.

FIG. 3.—Shows the cytological details more clearly. × 235.
Clinical Picture.—Primary cystic pneumatosi
unassociated with an organic change in the
abdomen is rare (Urban, 1937). According to
various authors, in 45 to 75 per cent of all cases
reported cystic pneumatosi was associated with
stomach or duodenal ulcer. The clinical diagnosis
of cystic pneumatosi is very difficult (Reverdin,
1924), most cases so far reported having been
recognized only at operation or at necropsy.
Mengis (1938) considers sudden severe abdominal
pain the leading clinical symptom of cystic
pneumatosi. Recently, however, several cases
have been diagnosed by radiography (Lerner and
Gazin, 1946; Berglund, 1939).

Cystic pneumatosi of the intestine is in itself
a benign process, and the cysts may be very quickly
absorbed. Surgical treatment, if necessary, should
therefore be directed against the underlying disease,
but there is no need for drastic resection of the
cystic intestine.

Pelnář (1900) described a case of cystic pneumatosi
associated with tuberculosis of the intesti-
tine. Bartáč (1941) reported on the histology of
a case of cystic pneumatosi of the caecum which
was surgically removed. More recently Vahala
(1946) described in some detail another case of
cystic pneumatosi of the caecum in which the
patient, a 44-year-old physician, was admitted to
hospital with a diagnosis of acute appendicitis. He
had been suffering for years with occasional
bouts of pain in the lower abdomen which came
on after exertion but subsided in a few days with-
out treatment. At operation the appendix appeared
normal, but the caecum was studded with grape-
like cysts in the serosa and deeper in the caecal
wall. In this patient the underlying condition was
a chronic duodenal ulcer, and the cystic pneumato-
sis may have been provoked by a mild attack of
enteritis.

In the present case, too, the clinical picture was
misleading and suggestive of malignant growth of
the large intestine.

Case Report

An undernourished woman of 65 was admitted to a
provincial hospital with a long history of indigestion
and constipation. She had recently been losing much
weight. A movable lump, the size of a man's fist,
was found on palpation in the ileocecal region, and
malignant growth of the large intestine was suspected.
At laparotomy the caecum and ascending colon were
found to be transformed into a grape-like mass that
showed many partly pedunculated vesicles which
collapsed when they were cut open. There was no
malignant growth, and the affected parts were resected.

The patient developed bronchopneumonia and died
soon after the operation. Necropsy could not be
performed.

Histology.—Three small pieces were excised from
the ascending colon and fixed in formalin. No
bacteriological or chemical examination could be
brought. The macroscopical appearance of the
specimens was characteristic: the cut surface showed
small round or oval cavities 1 to 3 mm. in diameter
situated in the intestinal wall. The consistency of the
specimens was sponge-like, not unlike emphysematous
lung tissue, and the cavities appeared empty.

Sections stained with haematoxylin and eosin
showed the glands to be unchanged and no signs of
inflammation in the mucosa (Fig. 1). Closely
arranged cystic cavities, varying in size and shape,
occupied the whole intestinal wall. They were separ-
ated by slightly oedematous connective tissue, and
some were lined by fairly large endothelial cells.
Others showed flat cells only, or no lining at all.
Numerous giant cells of foreign-body type lined some
of the cysts; more often they lay freely in the loose
connective tissue of the submucosa. Round, small
cysts were seen close to the glandular surface, larger
ones in the deeper layers. Some of these seemed to
have been formed by fusion of two or more smaller
cysts under pressure (see the "torn" wall in the
larger cyst, Fig. 2). There were a few round cells,
but no tuberculous or ulcerative changes and no signs
of malignancy.

Most of the cavities were empty. Some contained
a faintly blue-staining homogeneous material. In sec-
tions stained for fat some red-staining material was
seen in a few cysts.

Discussion

This case demonstrates how numerous are the
disguises under which cystic pneumatosi of the
intestine may occur. As in other cases, there was
a long history of dyspepsia and constipation. The
exact origin of these signs could not be established.
The tumour-like mass was entirely formed by the
cystic caecum and ascending colon. The cysts
were located in all the layers of the intestinal wall,
and they collapsed when incised. They contained
some serous and fatty material. This might
suggest that the cysts were formed from distended
lymphatics. As Lamont (1929) points out, the
anatomical arrangement of the lymphatics, which
form a complicated series of plexuses, would be
quite compatible with the tier-like arrangement of
the cysts. Dressler (1939) assumes that the cysts
are formed within the lymphatics.

The case was typical in that it simulated another
disease—malignant growth—and was only recog-
nized at operation. Its occurrence in an elderly
woman was unusual.
Summary

A case of cystic pneumatosis of the large intestine in a woman aged 65, simulating symptoms and signs of malignant growth, is described.

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