PNEUMATOSIS CYSTOIDES INTESINALIS
ASSOCIATED WITH ABDOMINAL CARCINOMATOSIS

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PNEUMATOSIS CYSTOIDES intestinalis is a rare condition in which gas-containing cysts arise in the intestinal wall and elsewhere within the abdominal cavity. The cysts vary in size from a millimetre to several centimetres in diameter and may occur in large numbers. In most cases the cysts are subserous but occasionally they arise in the submucous layer of the intestine. The pneumatosis is most often found in the small intestine and this may be the only area affected, but caecum, colon, parietal peritoneum, mesentery, omentum and lymph nodes are sometimes involved.

In over 50% of cases there is a peptic ulcer present, usually with some degree of stenosis, but the condition has also been described in association with tuberculous enteritis, carcinoma of stomach, appendicitis, intestinal obstruction, enteritis, colitis, peritonitis, intestinal parasites, and even following Caesarian section. In some cases the disease appears to be idiopathic.

In this patient the pneumatosis was associated with microscopic, and later macroscopic, infiltration of the intestine with neoplastic tissue.

Case Report

A widow of 60 was admitted to hospital with a three week history of pain, worse on deep breathing, in the region of the right scapula. She had no cough, breathlessness or oedema and she was a non-smoker. For two years she had experienced occasional attacks of abdominal discomfort and vomiting, and soon after their onset, certain investigations were done elsewhere. These, including X-rays of chest and abdomen, were normal. Appendicectomy had been performed at 28 and hysterectomy at 53.

On examination there were signs of a small, right-sided pleural effusion and the abdomen was distended and resonant. Chest X-ray showed the effusion and a pneumoperitoneum with gas beneath both diaphragms.

During the next eight weeks the patient deteriorated steadily. She lost weight, had bouts of fever and at times became very dyspnoeic and clammy. The pleural effusion increased in size, the pneumoperitoneum persisted, and the abdominal distension varied greatly. Sometimes the abdomen was inflated like a balloon and at other times the abdominal wall was flat and relaxed.

As investigations had not revealed the cause of the persistent pneumoperitoneum and pleural effusion, a laparotomy was performed by Mr. A. Desmond and pneumatosis cystoides intestinalis was discovered. In the peritoneal cavity there was a little free fluid which was sterile on culture. The parietal peritoneum was studded with small gas cysts varying in size from 1 mm. to 1 cm. in diameter. Two cysts were present on the upper surface of the left lobe of the liver, and the jejunum was covered over its whole surface with masses of cysts, giving the appearance of froth. Some of the cystic tissue was excised and on microscopy this showed cystic serosal spaces and small foci of mucin secreting carcinoma.

The patient recovered from the operation but continued to deteriorate and she died a month later.

A specimen of gas was aspirated from the abdominal cavity three hours after death and on analysis this contained nitrogen 54.1%, carbon dioxide 38.8%, hydrogen 7.0%, methane 0.1% and oxygen nil.

Necropsy. The right lung was bound to the chest wall by malignant tissue and a considerable amount of fibrinous fluid was present in the right pleural cavity. The abdomen contained a small quantity of yellow fluid but the striking feature was a large number of gas cysts on the intestines and parietal peritoneum. In addition there were multiple white plaques and bands of malignant tissue in the wall of the small intestine, and to a lesser extent, in the large intestine (Fig. 1). The stomach and oesophagus were normal. Several lymph nodes infiltrated with carcinoma were present in the mediastinum and lower para-aortic regions. No metastases were present in the liver, spleen, adrenals, kidneys or brain.
Histological sections of the right lung showed fibrosis and squamous carcinoma. Some areas resembled pavement epithelium, others contained vacuolated cytoplasm and areas of necrosis. Secondary deposits of similar pattern were seen in the various organs. The serosal surface of the small intestine showed patchy neoplastic infiltration with fibrinous exudate. Empty spaces, macroscopically interpreted as gas cysts, were often lined with malignant cells but this appeared to be fortuitous rather than causal.

Discussion. Duvernoy (1730) was the first to draw attention to abdominal gas cysts which he discovered during cadaver dissection, but these may have been due to post mortem changes in the tissues.

Several authors described a similar condition in pigs and it has also been found in chickens and sheep, but Bang (1876) produced the first satisfactory and well-documented description of the human disease finding it at autopsy in a woman who had died of volvulus.

Hahn (1899) described pneumatosis in a living person.

In his authoritative article on this subject, Koss (1952) reported a case of his own and analysed 212 others from the world literature. Since this time, reports of individual, and short series, of cases have appeared in various countries, especially America.

The association of pneumatosis cystoides intestinalis with malignant disease is uncommon. The earliest example of this is provided by the first case in the American literature (Finney, 1908). At operation a male patient of 60 was found to have a carcinomatous mass involving the pyloric third of the stomach with nodules of secondary carcinoma in the liver and retroperitoneal nodes. This pneumatosis was limited to the ileum.

In his large series, Koss (1952) reported seven cases of carcinoma of stomach. In one of these (Dale and Pearse, 1950, Case 2), the pneumatosis followed laparotomy. At operation a carcinoma of stomach was found with spread to lymph nodes and liver but no gas cysts were seen. However, autopsy five weeks later revealed pneumatosis involving mesentery, omentum, retroperitoneal nodes, jejunum, ileum and colon.

Mujahed and Evans (1958) report a series of seven cases of pneumatisis, and one of these, a patient with chronic lymphatic leukaemia, also had a carcinoma of lung, but no mention is made of intra-abdominal metastases.

Various theories have been advanced concerning the aetiology of pneumatosis cystoides intestinalis. In the earlier articles the neoplastic theory was suggested and Bang (1876) thought that the pneumatosis was "a distinct variety of tumour, the cells of which have the faculty of secreting gas."

From time to time bacteria and yeasts have been isolated from the cysts but none with any consistency, and infective and neoplastic theories have now been discarded.

Another view is that the gas is formed by chemical action in the chyle and this seems to be supported by the fact that pneumatosis is common in pigs fed on dairy refuse.

In the mechanical theory it is postulated that gas is formed from the intestinal lumen under pressure during peristalsis; a kind of surgical emphysema. Support for this view is provided by the fact that where the pneumatosis is associated with some other disease of the gastrointestinal tract, ulceration or inflammation of the mucosa and obstruction of the lumen are commonly found. Furthermore, treatment of the associated condition e.g. pyloric stenosis, is often followed by disappearance of the cysts. In one case the cysts were observed to increase in size when vomiting occurred during laparotomy.

It seems likely that several mechanisms exist whereby the cysts may be formed, but the most probable explanation of the human disease is provided by the mechanical theory.

The present case shows many of the typical features of pneumatisis cystoides intestinalis, but the most interesting finding is the association of the gas cysts with neoplastic infiltration of the intestine. In view of the discovery in the biopsy specimen taken at laparotomy of carcinoma cells in intimate relationship with the cysts, and the subsequent widespread infiltration of the intestinal wall with malignant tissue, there seems little doubt that this infiltration was an important factor in the production of the pneumatosis.

Summary

A case of pneumatisis cystoides intestinalis associated with abdominal carcinoma is described. The historical aspect, the association with malignant disease, and the aetiology, of this strange condition are briefly discussed.

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