

Case report

Cystic pneumatosis in coeliac disease

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SUMMARY This report presents a patient with proximal small bowel cystic pneumatosis associated with poorly controlled coeliac disease and pseudo-obstruction. Cystic pneumatosis is rare in the proximal small bowel and we can find no report of its occurrence in association with coeliac disease.

The finding of intramural gas in a hollow viscus is well recognised and has been reported in a large number of conditions, although there is sometimes uncertainty as to aetiology (Table). We wish to present a case of cystic pneumatosis in a patient with coeliac disease, an association not previously recorded.

Case report

An 83 year old woman was admitted as an emergency with a one week history of abdominal pain, distension, and vomiting. She had suffered with coeliac disease for 40 years, the diagnosis having been confirmed by several jejunal biopsies showing subtotal villus atrophy. She did not adhere to a gluten free diet and these indiscretions led to repeated admissions for reinforcement of dietary advice. She had osteoarthritis and was taking ibuprofen. For several weeks before admission she had steatorrhoea but had no bowel action for the two days preceding presentation, although she had passed flatus. On examination she was dehydrated but not clinically shocked. The abdomen was distended with signs of generalised peritonitis and rectal examination revealed soft, grey, frothy stool. Examination of blood revealed hypokalaemia, hypoalbuminaemia, and a macrocytic anaemia. Arterial blood gases were normal. An abdominal radiograph (Figure) showed free intraperitoneal gas, small bowel distension, and small bowel intramural gas. At laparotomy she was

seen to have pneumoperitoneum, but no free fluid. In the small bowel mesentery 5 cm distal to the ligament of Treitz, there was a collection of gas containing cysts ranging from 2 to 5 mm in diameter, and extending for approximately 100 cm along the small bowel. The mesenteric vessels were all pulsatile, and the bowel appeared healthy. There was no sign of acute or chronic peptic ulceration. No further intervention was considered necessary and the abdomen was closed. Predictably, she developed a chest infection which was treated with prolonged oxygen therapy and antibiotics. The remainder of the post-operative course was uneventful and on the seventh day no gas was visible on a plain abdominal radiograph. She was discharged 11 days after surgery on a gluten free diet and remains well 18 months later.

Table *Reported conditions associated with cystic pneumatosis*

	Reference
Ulcerative colitis	1
Asthma	2, 3
Mesenteric infarction	4, 5
Ischaemic colitis	6
Scleroderma	7
Colonoscopy (postbiopsy)	8
Crohn's disease	9, 10
Hirschsprung's disease	10
Imperforate anus	11
Meconium plugs	11
Leukaemia	12
Whipple's disease	12
Steroid therapy	12
Jejunol-ileal bypass	12
Chilaiditi's syndrome	7, 13-20

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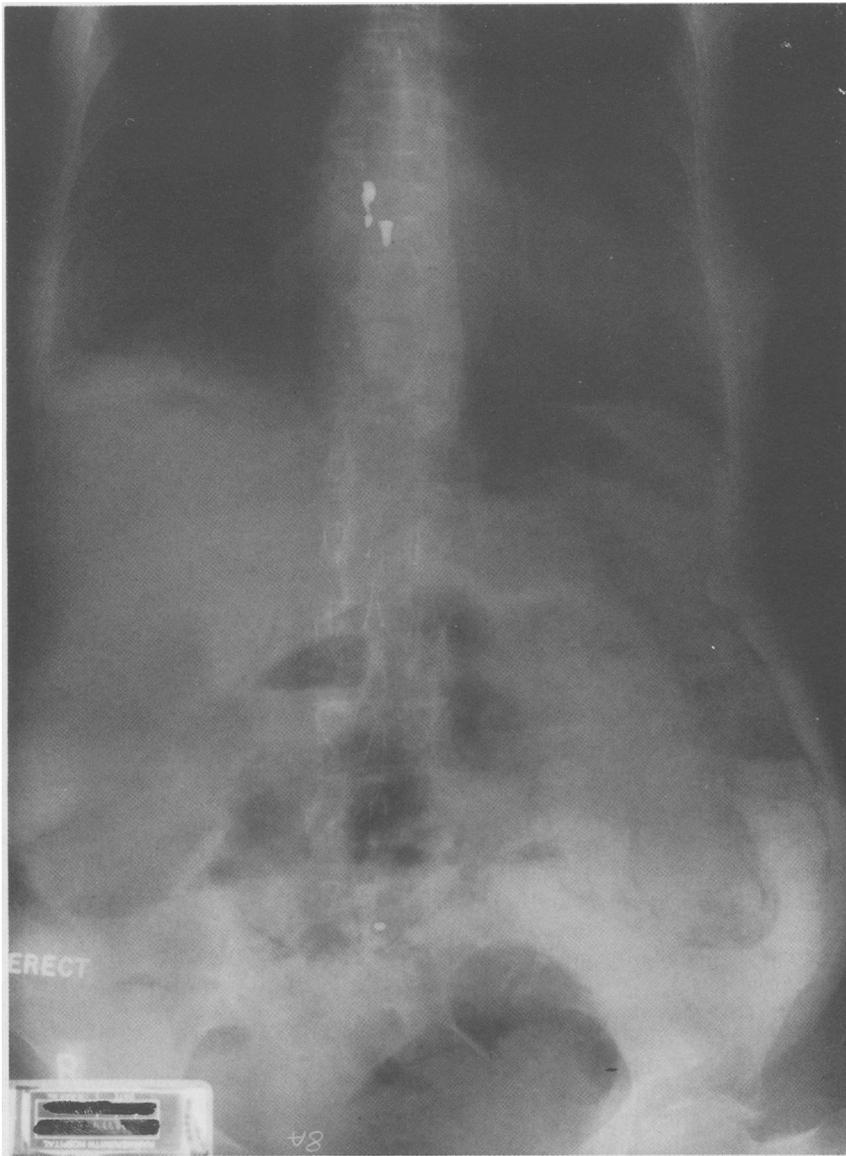


Figure Plain abdominal radiograph showing free gas, small bowel distension and intramural gas.

Discussion

Cystic pneumatosis was first described in 1730 by Du Vernoy and in 1786 Hunter described the condition in the intestine of a pig, that had been sent to him by Jenner. Thus the condition is not unique to man, but in animals is usually considered to be of no significance.²⁰ The cysts range in size from 0.5 mm to 10 cm, and are most common in the terminal ileum

and colon, but are rarely found in the stomach and proximal small bowel.^{21–26} The gaseous contents vary, but nitrogen predominates. The cysts probably represent distended tissue spaces, although cystic lymphatic vessels have also been suggested.²⁷ Giant cells similar to foreign body giant cells are often seen on histological examination. The aetiology of cyst formation has variously been thought to be caused by gas forming tumours,²⁸ gas forming bacteria,²⁹ dietary

causes,³⁰ or disturbances of acid based metabolism.³¹

In 1965, Colquhoun³² classified the various types of presentation of intramural gas in hollow viscera as: (1) gas-forming infections; (2) interstitial emphysema; (3) cystic pneumatosis (pneumatosis cystoides intestinalis). The role of gas forming organisms has been extensively documented and will not be discussed further. Interstitial emphysema is defined by the typical appearance of linear gas streaks on plain radiographs. There are no cysts identified and no clinical evidence of gas forming infection. Therefore cystic pneumatosis is really a diagnosis of exclusion.

In 1952 Koss hypothesised that one required raised intraluminal pressure and a break in mucosal integrity to develop these cysts, but in 15% of his reported cases, neither of these parameters were obviously fulfilled.¹³ Other authors³ have found the condition in patients with asthma and proposed a pulmonary origin of the gas. Keyting³ showed that rupture of an emphysematous bulla could lead to gas tracking along blood vessels to the lung root, and then through the aortic sheath to the mesenteric vessels. These authors noted that the gas bubbles tended to be grouped around the distribution of the major branches of the mesenteric vasculature.

Cystic pneumatosis has been noted in association with mesenteric infarction⁴⁻⁶ where, together with gas in the portal vein, it is a grave prognostic feature, and a similar association exists with ischaemic colitis.

In 1933 Jung and Ngai³³ documented the formation of subserosal gastric cysts in a patient undergoing surgery for a perforated gastric ulcer. The finding of cystic pneumatosis in perforated ulcers and jejunal diverticuli is rare,³⁴ however, which may be due to the lack of raised intraluminal pressure caused by the massive intraperitoneal leak.

Cystic pneumatosis has been seen in a number of varying conditions and treatment should always be directed at the primary disease. Should a complication occur as a result of the cysts, then surgery may be required. The use of hyperbaric oxygen has recently been advocated,³⁵ but such therapy must be prolonged for 48 hours after radiographic dissolution of the cysts.³⁶

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