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Pneumatosis cystoides intestinalis as a complication of celiac disease

The role of diagnostic laparoscopy

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Abstract

Pneumatosis cystoides intestinalis (PCI) is an uncommon but important condition characterized by the presence of gas-filled cysts in the submucosa and subserosa of the gastrointestinal tract. PCI has been associated with several clinical settings. We report a case where PCI developed in a patient with known celiac disease. To our knowledge, this type of coincidence has been described in seven prior cases. Since PCI often results in pneumoperitoneum through rupture of the cysts, it is important to differentiate the benign form of pneumoperitoneum, in which no intervention is indicated, from the life-threatening form with intestinal infarction and/or perforation, in which immediate surgery is mandatory. Differentiating between them can be difficult; this explains the high rate of negative laparotomy, which, in the past, occurred in 27% of cases. By performing diagnostic laparoscopy, we were able to establish the diagnosis of PCI and exclude intestinal perforation or infarction, thus permitting the patient to avoid an unnecessary laparotomy. Although diagnostic laparoscopy is not routinely indicated as a diagnostic tool for the detection of PCI, it proved to be a safe and accurate method to differentiate between the benign form of PCI and the life-threatening form. Whenever this differentiation has to be made, we recommend that diagnostic laparoscopy be performed first rather than primary laparotomy. Herein, the laparoscopic features of PCI are described and the coincidence of PCI with celiac disease is reviewed.

Keywords: Pneumatosis cystoides intestinalis, Celiac disease, Pneumoperitoneum, Laparoscopic features

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Pneumatosis cystoides intestinalis (PCI) is an uncommon but important condition in abdominal pathology. PCI is characterized by the presence of multiple gas-filled cysts and clusters in the submucosa or subserosa of the gastrointestinal tract. PCI has been associated with several clinical settings, including necrotizing enterocolitis in infants, obstructive pulmonary disease in adults, and a wide variety of gastrointestinal conditions, such as pyloric stenosis, ischemic bowel disease, progressive systemic sclerosis. It is also seen in association with drug therapy, particularly when steroids, chemotherapy, and immunosuppression are involved [11].

The pathogenetic mechanisms of PCI are still uncertain, but it appears that there are two main sources for the gas, one mechanical and one bacterial. The mechanical theory proposes that gas enters the bowel wall from either an obstructed intestinal tract or the lung [4, 6]. The bacterial theory proposes that gas-forming bacilli (especially *Clostridia perfringens*) enter the submucosa through mucosal rents and produce gas within the intestinal wall [26]. Since cultures of the cysts are consistently sterile, it is assumed that intraluminal rather than intracystic bacteria produce an

excessive amount of gas, which under pressure enters the intestinal wall [11].

With the exception of a statistical study of 919 cases by Jamart [12], only small series of PCI have been reported. In contrast, celiac disease is a much more common pathological entity. To our knowledge, there have been only seven case reports describing the simultaneous appearance of both pathologies, PCI and celiac disease, in a patient.

PCI may therefore be encountered as a rare complication or concomitant disease of celiac disease. It may mimic intestinal perforation by showing free intraabdominal air on conventional radiographs. In patients with PCI of the small bowel, the subserosal cysts rupture in ~15% of cases, resulting in pneumoperitoneum [12]. In most of these patients, the cyst rupture does not produce bacterial peritonitis. This self-limiting and in most cases asymptomatic pneumoperitoneum has therefore been termed "benign pneumoperitoneum."

However, in a previous study, one of 12 such patients had documented bowel infarction [1]. Therefore, careful stationary observation of these patients seems to be mandatory. At the other extreme, PCI may be associated with a life-threatening intestinal infarction and/or perforation that requires immediate surgical intervention without permitting any diagnostic delay. Since PCI may mimic gastrointestinal perforation and differentiation between the benign pneumoperitoneum and the life-threatening form may be difficult to assess, some patients with the self-limiting form of PCI have erroneously undergone laparotomy.

We report herein on a patient with known celiac disease where the clinical course and radiological follow-up mimicked a gastrointestinal perforation. By performing diagnostic laparoscopy, we were able to diagnose the coincidence of PCI and exclude perforation or infarction of the intestinal tract. We will describe the laparoscopic features of PCI and review the literature on the coincidence of PCI in patients with celiac disease.

Case report

Three months before admission of the 66-year-old male patient to our department, a history of frequent foul-smelling stools, weight loss, and abdominal pain was noted in the patient. The ensuing endoscopic investigation of the upper gastrointestinal tract, including biopsy contributed to the diagnosis of celiac disease. After a gluten-free diet was established, there was some improvement of the intestinal discomfort during the following weeks. The medical history of the patient included a mild chronic obstructive lung disease (COPD), grand mal epilepsy with long-term medical treatment over 50 years, and paranoid schizophrenia, both of the latter conditions without actual activity. The patient's daily medication consisted of 500 mg clozapinum, 10 mg haloperidole, and 400 mg carbamazepine.

At admission, the patient showed all the clinical signs of mechanically induced intestinal subileus. The abdominal examination revealed a distended nontender abdomen with diffuse pain, high-frequency bowel sounds, and shifting dullness. The blood samples revealed a leukocytosis of 12,100 mg/L, a C-reactive protein of 140 mg/L, anisocytosis, and hypochromasia. An abdominal radiograph revealed dilated, air-filled small bowel loops with few fluid levels. After 24 h of conservative treatment, the patient developed the clinical sign of peritonitis in the upper abdomen, and radiographic control of the chest and abdomen revealed free air under both diaphragms, mimicking intestinal perforation (Figs. 1 and 2).

The patient was urgently brought into the operative theater, where a diagnostic laparoscopy was performed. By laparoscopy, multiple gas-filled cysts and clusters in the subserosa of the small bowel wall and its mesentery could be appreciated, and a diagnosis of PCI was established (Fig. 3). The insertion of two additional working trocars allowed the entire intraabdominal intestinal