Annals of Clinical and Medical Case Reports

Case Report

ISSN 2639-8109 | Volume 10

Pneumatosis Cystoides Intestinalis in Ulcerative Colitis; Histological Characteristic of the Cystic Structures

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Received: 15 May 2023
Accepted: 21 June 2023
Published: 29 June 2023
J Short Name: ACMCR

Keywords:

Penumatosis cystoides intestinals; Hypertrophic cystic wall; Ulcerative colitis

Citation:

Mori H, Pneumatosis Cystoides Intestinalis in Ulcerative Colitis; Histological Characteristic of the Cystic Structures . Ann Clin Med Case Rep. 2023; V10(24): 1-4

1. Abstract

Pneumatosis cystoides intestinalis (PCI) is a rare disease characterized by multiple gas-filled cysts in the intestinal wall, and may be easily missed or misdiagnosed. We report here a case of 40-year-old man who was suffering from ulcerative colitis (rectal type) for 14 years. He was referred to the gastroenterology department of our hospital for the complaint of consistent abdominal pain. Colonoscopy revealed multiple nodular protuberances covered with normal-looking mucosa in the cecum and ascending. Diagnosis of PCI was made by cyst collapse after puncturing. Biopsy from the PCI revealed that gas-filled cysts were mostly located in the submucosal space with rather hypertrophic cyst wall, and a layer of multinucleated histiocytes was located at the surface of the inside of the cysts. Surrounding the cysts, lymphocyte infiltration was apparent. These evidences suggested that gas-filled cysts survived for a long term. It is considered that destiny of PCI without surgical treatment will be variable with the type of onset, process and histological structures of the PCI.

2. Introduction

Pneumatosis cystoides intestinalis (PCI), which was first described by Du Vernoi in autopsy specimens in 1730 [1], is a rare disease without obvious symptoms that most frequently occurs in the large and small intestine. Preoperative diagnosis of PCI is difficult, and it may be easily misdiagnosed. PCI can be primary or secondary to other diseases, and the secondary type accounts for 85% of the cases [2-6]. A vast number of factors for the secondary type such as inflammatory bowel disease (IBD) involving Crohn's disease and ulcerative colitis, drug use, and chronic obstructive pulmonary disease [7]. Thus, diagnosis is challenging because of the asymptomatic or atypical symptoms.

Up to date, mechanism of the onset and advance of PCI are not clear, although different hypothesis has been proposed. In 1998, Pear [8] introduced a classification of the major pathogenic mechanisms causally involved intramural gas formation, which included (i) bowel necrosis, (ii) mucosal disruption, (iii) increased mucosal permeability, and (iv) pulmonary disease. Industrial chemicals and drugs are also known to concern with the occurrence of PCI. Trichloroethylene which has been is widely used as an organic solvent is a potent risk factor for PCI [9, 10]. Alpha-glucosidase inhibitor (α-GI) to control blood glucose has been also reported to associate with pathogenesis of PCI [11, 12].

Presently, we report a case of PCI of the cecum and ascending colon of the patient with ulcerative colitis (rectal type). To our knowledge, detailed histological property of PCI has not been clear, present investigation may give rise a unique clue to understand the growing process of PCI.

3. Case Presentation

A 40-year-old man was referred to the gastroenterology department of our hospital for a thorough examination. He was suffering from consistent abdominal pain and intermittent bloody stool starting age of 26. The patient was given salazopyrin by a practitioner under the diagnosis of ulcerative colitis. This patient worked at a chemical company where he used organic solvents (details unknown) for about 5 years starting age of 31 or 32.

On the consultation, the patient had no obvious abdominal distention, vomiting and diarrhea. Abdominal CT revealed no evidences of urinary calculus, hydronephrosis and accumulation of ascites. No gross abnormality of the gastrointestinal tract was found. Furthermore, no sign of neoplastic findings of retroperitoneal organs including pancreas and kidney, was present. However, colonoscopy revealed multiple nodular protuberances with size of 1-2cm covered with normal-looking mucosa in the cecum and ascending colon suggesting the presence of PCI (Figure 1). To further clarify the diagnosis of PCI, we used a mucosal injection needle to puncture the cyst. The cysts collapsed, and no return of blood was present after puncturing the cysts. Biopsy was done from the cysts and other unaffected colon and rectum. Biopsy from the PCI revealed that gas-filled cysts were mostly located in the submucosal space. The wall of the cyst was thickened with fibrous tissues. Inner side of the cyst was covered by a layer of multinucleated histiocytes (Figure 2). In some of the histiocytes, englobed small bubbles of the gas was recognized (Figure 3). Surrounding the gas-filled cyst, prominent lymphocyte infiltration was apparent suggesting that these cysts survived a fairly long term. On this thorough examination, the patient was given the diagnosis of ulcerative colitis (rectal type) in remission stage. Considering that patient had no peritonitis or other complication, conservative approaches including oxygen inhalation and oral probiotics were used. No recurrence of digestive tract symptoms or other discomfort occurred during the 1 year of follow-up.

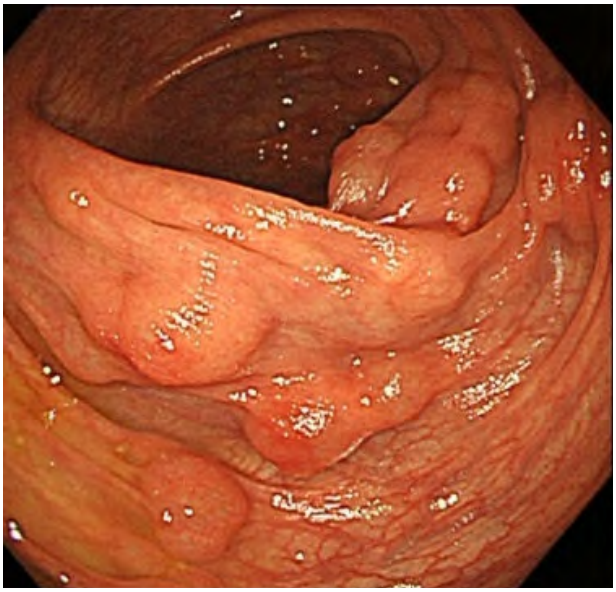

Figure 1: Colonoscopy reveals multiple nodular protuberances with size of 1-2cm covered with normal-looking mucosa. Some protuberances are conglutinated.

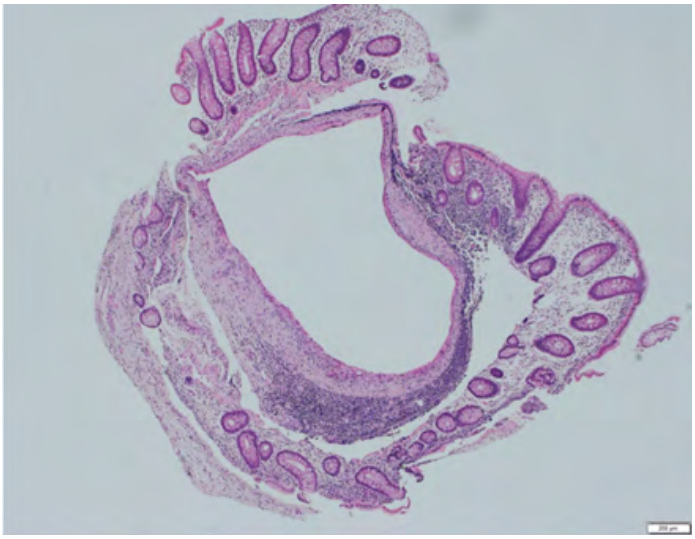

Figure 2: A low magnification of PCI. Gas-filled cysts are located in the submucosal space. The wall of the cyst is thickened with fibrous tissues. Inner side of the cyst is covered by a layer of multinucleated histiocytes. Surrounding the cyst, prominent lymphocyte infiltration is apparent.

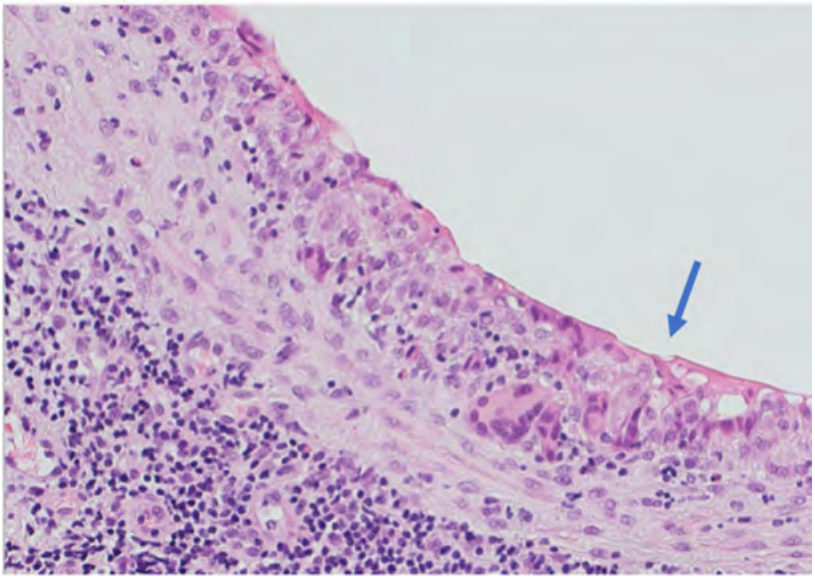

Figure 3: High magnification of a part of the cyst. In some histiocytes, englobed small bubbles of the gas are recognized (arrow).

4. Discussion

PCI is a rare disease by multiple gas-filled cysts in the intestinal wall. The symptoms of PCI vary and may be asymptomatic or atypical. PCI can be primary or secondary to other type accounts for 85% of the cases. The clinical relevance of PCI also varies widely and ranges from benign to life-threating conditions depending on the underlying cause of PCI [13]. Present case probably belongs to the benign group. Up to date, true mechanisms through which gas enters the intestinal wall are not well understood [5, 13-15]. Multiple hypotheses have been proposed to explain this mechanism. The three most-well accepted hypotheses are the pulmonary, mechanical and bacterial theories. Pulmonary theory suggests that chronic lung diseases may rupture alveoli, causing mediastinal emphysema and trapping gas into the intestinal wall along the aorta and mesenteric vessels [5]. The mechanical theory refers to the increased intraluminal pressure caused intestinal obstruction or other conditions that can lead to mucosal damage and promote cyst formation [13]. The bacterial theory refers to the intestinal bacteria that produce gas and trap the gas into the submucosa or aerogenic bacteria that directly penetrate the intestinal mucosa in the submucosa and produce gas [14, 15]. Bacterial theory is supported by the concept of counterperfusion supersaturation, whereby the intraluminal bacterially produced hydrogen tension exceeds the nitrogen tension in the blood, causing a hydrogen diffusion gradient towards the submucosal vessels [13].

Ulcerative colitis is regarded as one of representative precursor diseases for PCI. Interestingly, for these cases, PCI occurred preferably in unaffected sites of ulcerative colitis [16]. According to Nakano et al [17] with 9 cases of ulcerative colitis accompanying PCI, 4 cases exerted consistency in sites of both lesions, remaining 5 cases had inconsistency for both lesions. Furthermore, in almost all cases, PCI occurred in right site of the colon in remission stage of ulcerative colitis [16-18]. This is almost in agreement with the present case.

To our experience regarding ulcerative colitis, mucous membrane of unaffected part is tended to be atrophic. Recently, in a study of gastrointestinal manifestation of COVID-19 infection, histopathologic examination of the patients underwent colectomies (mostly right colectomies) revealed a spectrum of disease from superficial mucosal ischemic colitis to frank transmural ischemic colitis and associated changes being consistent with PCI [19]. This imply that ischemic gastrointestinal changes are importantly concerned with the etiology of PCI. Accordingly, superficial mucosal ischemic colitis or chronic atrophic colitis following additional iatrogenic trauma or manipulation increasing intraluminal pressure is a plausible pathway leading to the PCI.

Meanwhile, the present study showed that structure of gas-filled cysts of PCI may be fairly strong. It is confusing that both of ulcerative colitis and PCI have similar symptoms. Moreover, this patient has a long history of ulcerative colitis. Thus, onset of PCI of this patient may need to retroact to over 10 years. As one of his medical histories, is present the use of organic solvents. This might be related to the occurrence of PCI in this patient, although details are unknown.

After examining of 123 patients with PCI, Boerner et al [3], reported that remission was achieved in 70% of patients using nonsurgical treatment. However, destiny of PCI without surgical treatment will be variable with the type of onset, process and histological features of the PCI.

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