

## Report

## A Case of Pneumatosis Cystoides Intestinalis in a Patient with Crohn's Disease

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Pneumatosis cystoides intestinalis (PCI) has been reported to develop in association with conditions that increase intra-abdominal pressure. However, the finding of PCI is relatively rare in Crohn's disease (CD). A case of PCI occurring with ileocecal CD is reported. Difficulties may arise at laparotomy when the surgeon needs to determine the resection margin. A 61-year-old woman was diagnosed with ileocecal CD. She achieved remission with prednisolone and 5-aminosalicylic acid, but was later admitted to our hospital due to abdominal distention. Barium enema revealed an ileocecal stricture due to CD and PCI extending to the ascending colon. Colonoscopy showed multiple, smooth-surfaced, small hemispherical protrusions in the ascending colon. Computed tomography showed wall thickness and intramural air bubbles. Laparoscopic surgery was performed because of the ileocecal stricture from CD 4 months later. The resected specimen showed stricture of the terminal ileum and cobblestoning edematous mucosa. PCI was recognized in the submucosa of the ascending colon. The patient's post-operative course was uneventful. The distal resection margin was able to be determined based on the findings of submucosal edema. Therefore, intraoperative endoscopy is required when the PCI margin is unclear. Although reports of CD associated with PCI are rare, this condition must be kept in mind. The resection margin should be determined based on the distribution of CD lesions and the degree of intestinal edema for a safe anastomosis.

**Key Words:** Crohn's disease, pneumatosis cystoides intestinalis, intestinal pneumatosis, inflammatory bowel disease, surgical margin

## Introduction

Pneumatosis cystoides intestinalis (PCI) is characterized by the presence of gas within the submucosal or subserosal layer of the intestinal wall. The etiological mechanisms are unclear, although PCI has been reported to develop in association with raised intra-abdominal pressure due to ileus surgery<sup>1)</sup>, colonoscopy<sup>2)</sup>, pulmonary diseases such as chronic bronchitis<sup>3)</sup>, trichloroethylene exposure<sup>4)</sup>, connective tissue disorders<sup>5)</sup>, use of immunosuppressants<sup>6)</sup>, and inflammatory bowel disease (IBD) such as ulcerative colitis (UC) and Crohn's disease (CD). However, PCI is relatively rare in CD<sup>7)~13)</sup>.

To illustrate the therapeutic problems related to the co-existence of the two lesions, a case of PCI occurring in a patient with ileocecal CD is reported. Difficulties may arise at laparotomy when the sur-

geon has to determine the bowel resection margin.

## Case Report

## Patient

A 61-year-old woman was diagnosed with ileocecal CD in October 2001. She achieved remission of CD with prednisolone (5 mg/day) and 5-aminosalicylic acid (5-ASA). She had a history of abdominal distension, increased flatus, and constipation in July 2006. She was referred to our hospital for investigation and treatment. The abdominal pain increased in late August 2008.

Past medical history revealed acute appendicitis that required an appendectomy 46 years earlier.

On admission, her height was 144 cm, and her weight was 39 kg. On examination, she was febrile, with a distended and generalized tender abdomen, without signs of peritoneal irritation.

Laboratory investigations revealed anemia and slight elevation of C-reactive protein, with no abnormalities in the white blood cell (WBC) count (Table 1).

Plain abdominal radiography showed small collections of bubbly radiolucent gas along the wall of the colon (Fig. 1). A barium enema examination showed an ileocecal stricture due to CD and PCI extending to the ascending colon (Fig. 2). On colonoscopy, multiple, smooth-surfaced, small, hemispherical protrusions were observed in the right colon (Fig. 3A, 3B). The colonoscope was unable to be passed through the ileocecal stricture to the oral side (Fig. 3C, 3D).

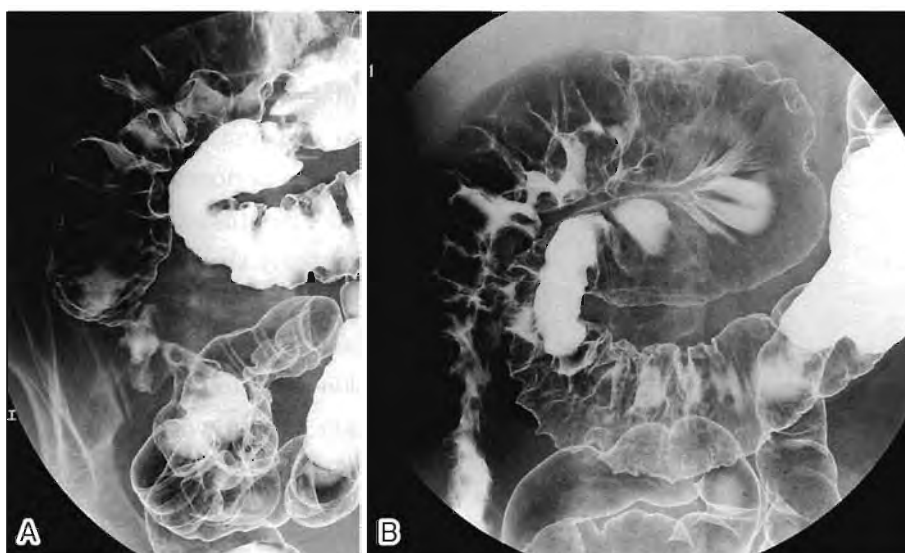
**Table 1** Laboratory findings on admission

Complete blood count		Blood chemistry	
WBC	6,710 / $\mu$ L	Total protein	6.2 g/dL
RBC	$335 \times 10^6$ / $\mu$ L	Albmin	3.9 g/dL
Hb	8.5 g/dL	AST	11 U/L
Ht	28.5 %	ALT	11 U/L
Platelet	$32.3 \times 10^4$ / $\mu$ L	LD	170 U/L
		ALP	145 U/L
		Total cholesterol	210 mg/dL
		Triglyceride	153 mg/dL
		Creatinine	0.77 mg/dL
		BUN	17.7 mg/dL
		C reactive protein	0.83 mg/dL
		HbA1c	5.8 % (JDS)

No ulcerations or active lesions of CD were evident on the anal side of the colon and rectum. Computed tomography (CT) of the abdomen showed a thickened wall and bubbly intramural air in the ascending colon (Fig. 4), with thickening of the bowel wall in the ileocecal area. Intra-abdominal free air and

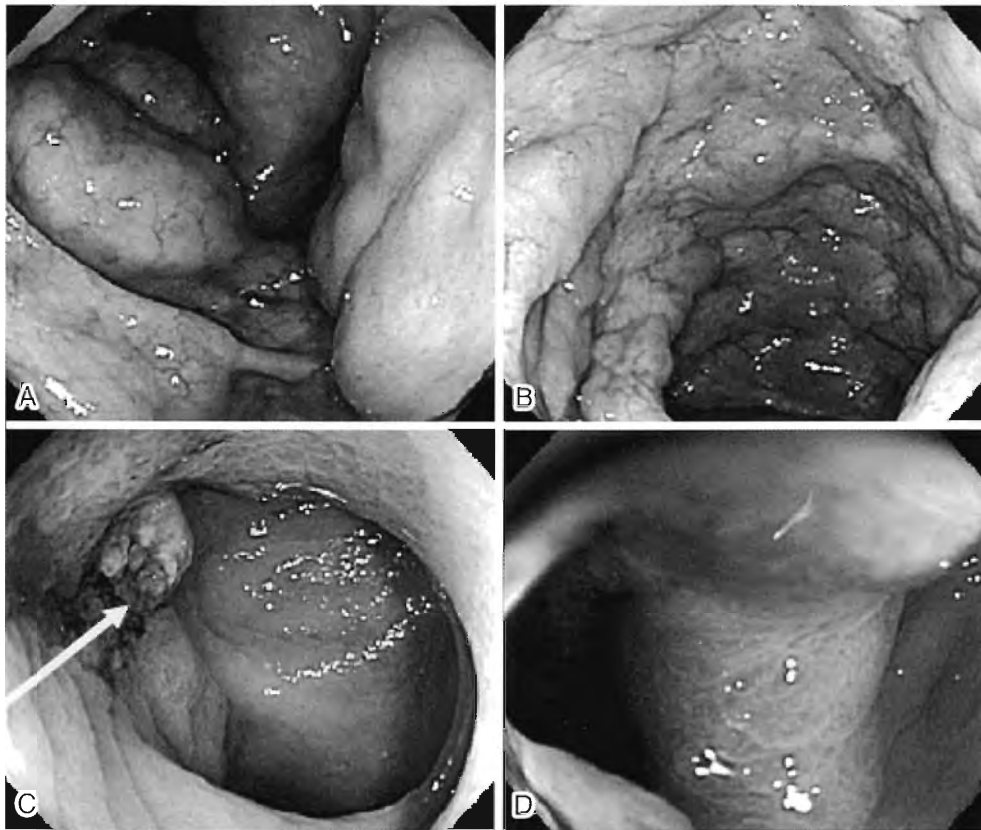


**Fig. 1** Abdominal X-ray findings  
Multiple air bubbles are seen in the ascending colon distal to the ileocecal stricture.  
▲ air bubbles in the ascending colon.  
→ ileocecal stricture.



**Fig. 2** Barium enema findings

A: Ileocecal stricture due to Crohn's disease.  
B: Multiple protruding mucosal lesions in the ascending colon.  
The barium enema shows an ileocecal stricture due to CD and PCI extending to the ascending colon.



**Fig. 3** Endoscopic findings of the ascending colon

A, B: Multiple protruding mucosal lesions in the ascending colon.

C: Anal side of the stricture (arrow).

D: Inside view of the stricture.

Colonoscopy shows multiple, smooth-surfaced, small, hemispherical protrusions in the ascending colon. The colonoscope could not be passed through the ileocecal stricture to the oral side (arrow).



**Fig. 4** Computed tomography findings

Computed tomography (CT) of the abdomen shows wall thickness and intramural air bubbles in the ascending colon.

portal venous gas were not seen. These findings suggested ileocecal stricture due to CD and PCI in

the ascending colon.

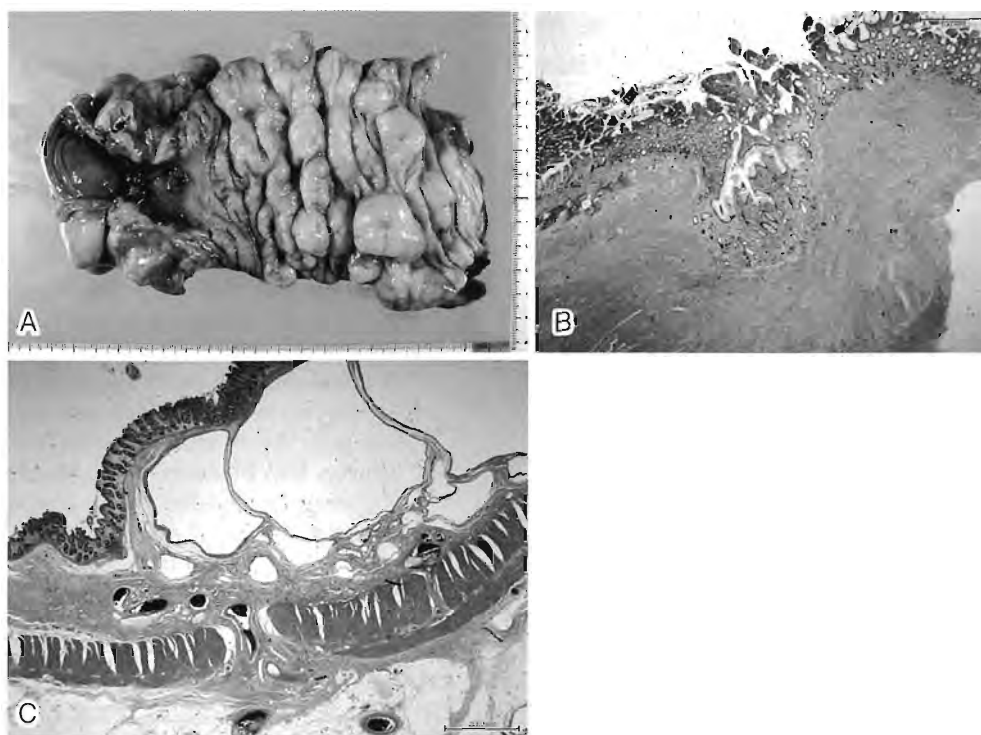
The patient was treated conservatively with an elemental diet because she refused surgical treatment at that time. Therefore, elective laparoscopic surgery was performed due to an ileocecal stricture from CD 4 months later.

Laparotomy showed ileocecal disease with slight dilation of the oral side bowel due to the severe stricture. Submucosal PCI was noted in the ascending colon distal to the ileocecal stricture. The distal resection margin was able to be determined based on the findings of submucosal edema.

The resection included 20 cm of the distal ileum and ascending colon, and a functional end-to-end anastomosis (FEEA) was performed.

**Macroscopic findings of the resected specimen** (Fig. 5A)

The resected specimen showed stricture of the



**Fig. 5** Resected specimen

A: Macroscopic findings. The resected specimen shows the stricture of the terminal ileum and cobblestoning mucosa with edema. No mucosal ulcerations or fistula formations of CD are present.

B: Microscopic findings of Crohn's disease. A noncaseating epithelioid granuloma is seen in the stricture.

C: Microscopic findings of PCI. PCI is seen in the submucosa of the ascending colon.

terminal ileum and cobblestone mucosa with edematous areas. Therefore, the active lesion of CD was limited to the oral side of the colon. No mucosal ulcerations or fistulas of CD were noted.

#### Microscopic findings (Fig. 5B, 5C)

A noncaseating epithelioid granuloma was seen in the stricture. PCI was recognized in the submucosa of the ascending colon.

The patient's postoperative course was uneventful. She now remains well on maintenance infliximab treatment, with no residual pneumoperitoneum.

#### Discussion

PCI is associated with IBD, chronic obstructive pulmonary disease (COPD), acquired immunodeficiency syndrome (AIDS), immunosuppressive therapy, infection and intestinal ischemia. PCI has been infrequently described in patients with IBD. Most reports are limited to case studies and small series. PCI may produce no symptoms and, as such, re-

quires no therapy, as exemplified by the present case.

CT and abdominal radiography are the most frequently used modalities for diagnosing PCI. Low window settings are important in the detection of intraluminal air and obviate the need for intraluminal contrast to outline the circumferential pattern of PCI.

The two most widely accepted theories about the formation of PCI attribute the disorder to either mechanical or bacterial causes. Mucosal ulcerations, increased intraluminal pressure, and hyperperistalsis are considered to be the reasons for development of PCI. Possible explanations for PCI arising in bowel affected with Crohn's disease include increased intraluminal pressure and peristalsis with diarrhea and obstruction, transmural ulcerations in the bowel wall, and increased bacterial flora secondary to obstruction.

PCI may produce no symptoms and requires no

therapy. This is of particular importance for Crohn's patients, in whom small bowel resection should be avoided if possible in anticipation of further surgery and an eventual short-bowel syndrome. Elective laparoscopic surgery was selected for the present patient because of a severe stricture of Crohn's disease in the terminal ileum.

In the present case, the distal resection margin was able to be determined based on the findings of subserosal edema; therefore, intraoperative endoscopy is required when the margin of submucosal edema is unclear. Since PCI is a benign disorder, complete resection is unnecessary. If PCI spreads over long segments, the resection margin should be decided based on the distribution of CD lesions and the degree of intestinal edema for a safe anastomosis.

### Conclusion

Although reports on Crohn's disease associated with PCI are rare, this condition must be kept in mind. The resection margin should be determined based on the distribution of CD lesions and the degree of intestinal edema for a safe anastomosis.

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## 腸管のう胞状気腫症を併発したクローン病の1例

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腸管のう胞状気腫症（以下、PCI）は腸管壁内にガスが貯留する病態で、発症機序は不明であるが炎症性腸疾患をはじめ様々な病態に合併することが知られている。しかしながら、実臨床でクローン病（以下、CD）に合併することは比較的稀である。回盲部CD狭窄の肛門側にPCIが発生し、CDによる狭窄のため手術を施行した症例を経験した。腸管切除手術を考慮する際に切除断端の決定に迷うことも推測されるため本症例を報告する。症例は61歳、女性。2001年他院にてCDと診断され副腎皮質ステロイド5mg/日および5-アミノサリチル酸の内服で寛解状態であった。他の併存症や薬物暴露の既往歴はない。2008年8月、腹痛が増強したため当院入院となる。注腸造影ではCDによる回盲部狭窄とその肛門側のPCIの所見が認められた。CTでは腸管壁の肥厚と内腔のバブル状のガス像を認めた。栄養療法を併用して保存的に加療したが、4ヵ月後に通過障害をきたす回盲部CD病変のため腹腔鏡下手術となった。手術所見では、回盲部に著明な狭窄があり口側回腸は軽度拡張していた。狭窄の肛門側の上行結腸では、CD病変に連続したPCIとその境界が漿膜面から可能であった。回盲部切除を行って機能的端々吻合で再建した。切除標本では、回盲部狭窄の非乾酪性肉芽腫を伴うCD病変とその肛門側の粘膜下に連続するPCIを認めた。粘膜面に潰瘍形成や瘻孔形成は認めなかった。術後経過は良好に退院となり現在までPCIの再発は認めていない。PCIの成因には腸管内圧の上昇に起因する器械説と細菌説などが推測されている。本症例ではCD狭窄病変に連続して肛門側腸管に発生していることからCDによる粘膜破綻からの器械説が疑われる。CDに併発したPCIは稀であるが、本症を念頭に診療にあたる必要がある。腸管切除縁の決定はCDの病変範囲と腸管浮腫の状況を考慮して判断すべきである。