

Primary Pneumatosis Cystoides Intestinalis

Chun-Hsiung Liu, MD, Hong Haw Chen¹, MD, Wan-Ting Huang², MD

We describe an unusual case of pneumatosis cystoides intestinalis (PCI) in a patient with chronic pulmonary disease. A 79-year-old woman was hospitalized due to abdominal fullness and bowel habit change. Colonoscopy revealed numerous round cystic lesions in the sigmoid colon, which bled easily on contact. Due to persistent local peritonitis, a left hemicolectomy with primary anastomosis was performed. The patient has done well in the 12 months following surgery. (*Chang Gung Med J* 2003;26:144-7)

Key words: pneumatosis cystoides intestinalis.

Pneumatosis cystoides intestinalis (PCI) is a rare disorder, characterized by the presence of gas-filled cysts in the submucosa and subserosa of the bowel. The cysts contain nitrogen, hydrogen, and carbon dioxide but not methane.^(1,2) The pathogenesis of PCI remains obscure. The primary mode of therapy is conservative treatment with antibiotics, an elemental diet,⁽³⁾ and inhalation of gases with a high concentration of oxygen. There is a high rate of recurrence (50%-78%)⁽⁴⁾ within months to years. We report a case of PCI in an elderly woman with chronic pulmonary disease who received operative resection for local peritonitis.

CASE REPORT

A 79-year-old woman was admitted due to left lower quadrant pain and abdominal fullness for 3 days. She had developed a bowel habit change with mucoid diarrhea for several months. She had previously been diagnosed as having congestive heart failure, bilateral pulmonary emphysematous lesions, and a suspicious tuberculous lesion over the right upper lobe of the lung. On gross physical examination, she was malnourished. The abdomen was distended and tympanic to percussion. The bowel sounds were

hypoactive. On palpation, no palpable mass was noted. Tenderness over the left lower abdominal quadrant was also noted. Laboratory values on admission were as follows: hemoglobin 13.7 g/dl, hematocrit 41.6%, white cell blood count (WBC) 4400/mm³, neutrophils 69.0%, monocytes 11.4%, lymphocytes 18.5%, alkaline phosphatase 30 U/L, albumin 2.7 g%, and carcinoembryonic antigen (CEA) 2.98 ng/dl. Abdominal computed tomography showed segmental narrowing of the sigmoid colon. A lower gastrointestinal series revealed long segmental multiple radiolucent cystic filling defects in the sigmoid colon. Colonoscopic examination revealed numerous round cystic lesions in the sigmoid colon, which bled easily on contact (Fig. 1). A biopsy was not performed due to concern about the local peritonitis. Due to persistent local peritonitis and the symptom occurring off and on for several months, she did not receive high-flow oxygen inhalation therapy. An exploratory laparotomy showed multiple cystic lesions protruding into the serosal layer of the sigmoid and descending colon, with an air-tight sensation. The mesentery was not involved.

A left hemicolectomy with primary anastomosis was carried out. Pathological analysis revealed mul-

From the Department of General Surgery, Tainan General Hospital, Department of Health, the Executive Yuan, Tainan; ¹Department of Colon and Rectal Surgery, ²Department of Pathology, Chang Gung Memorial Hospital, Kaohsiung.

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Address for reprints: Dr. Hong Haw Chen, Department of Colon and Rectal Surgery, Chang Gung Memorial Hospital, 123, Ta-Pei Road, Niasung, Kaohsiung 833, Taiwan, R.O.C. Tel.: 886-7-7317123 ext. 8094; Fax: 886-7-7328229

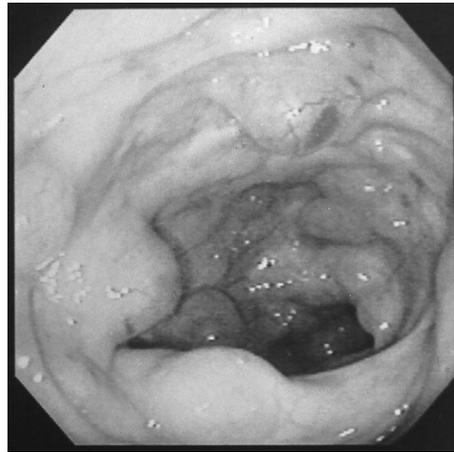
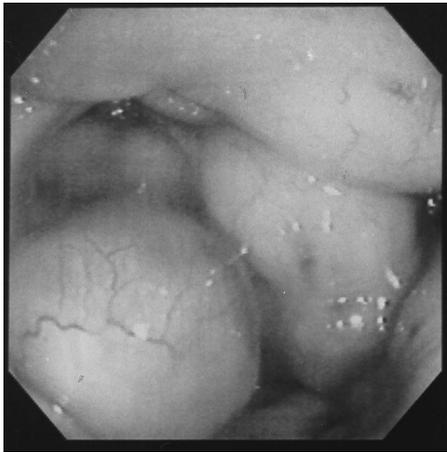


Fig. 1 Colonoscopy showing numerous cystic lesions of various sizes in the sigmoid colon.



Fig. 2 Gross appearance of the resected colon revealing multiple cobblestone-like lesions with gas-filled cysts distributed within the bowel wall.

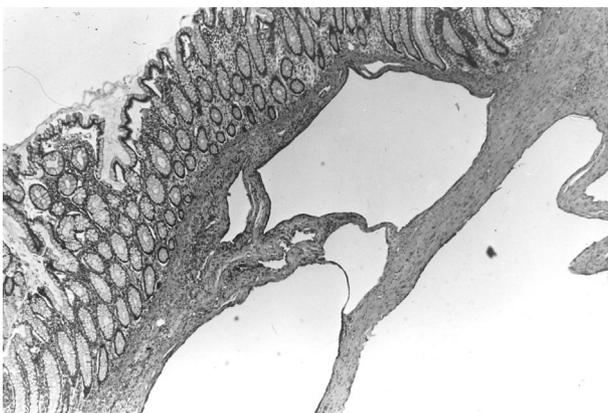


Fig. 3 Histology showing irregular dilated cysts distributed within the muscularis propria and submucosal layer, and which are lined by multinucleated giant cells. (hematoxylin & eosin, $\times 10$)

multiple cobblestone-like lesions apparently associated with gas-filled cysts distributed within the mucosa and submucosa (Fig. 2). Microscopically, the cysts were mainly distributed within the submucosa and muscularis propria and were lined by multinucleated giant cells. The overlying mucosa revealed chronic inflammatory cell infiltration in the lamina propria (Fig. 3). The postoperative course was uneventful, and there has been no recurrence during the 12-month follow-up period.

DISCUSSION

PCI is characterized by the presence of gas-filled cysts in the wall of the small bowel or colon, and the left side of the colon is usually affected. Clinical manifestations are non-specific with various presenting symptoms that include: diarrhea, mucoid discharge, rectal bleeding, and constipation. The etiology of PCI is not clear, and several theories have been proposed, among which the mechanical hypothesis may have been the most likely in our case. Increased intraluminal air pressure pushes through tears in the mucosa, and air is entrapped within the lymphatic channel and bowel wall. These events might occur during partial colonic obstruction, colonoscopic complications,⁽⁵⁾ or with excessive colonic gas production in patients with chronic obstructive pulmonary disease who are unable to excrete gas via the lungs.

Plain abdominal films may show the "stripe sign" due to overlapping of the cyst walls, producing a thin

line of increased density in the middle of the empty bowel lumen.⁽⁶⁾ Barium enema examination of the lower gastrointestinal portion of the abdomen in PCI usually reveals clusters of translucent lesions along the margin of the bowel wall or multiple, smooth, filling defects throughout the colon wall that mimic either polyps or cancer. Visualization of the gas-filled cysts on colonoscopy often confirms the diagnosis of PCI in the hands of an experienced endoscopist. The disease is benign and can be managed conservatively since some lesions disappear spontaneously within months or years. The high recurrence rate (50%-78%)⁽⁴⁾ may be due to an underlying disease such as psychiatric disorders, chronic pulmonary diseases, colitis, or the inability to completely eradicate anaerobic bacteria within the colonic mucosa.⁽²⁾ Since the mortality is very high (up to 75%)⁽⁷⁾ once these patients develop bowel obstruction or ischemia, it is advisable to perform surgical resection of the involved segment of the bowel before such conditions develop.

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原發性腸氣囊病

劉俊雄 陳鴻華¹ 黃琬婷²

腸氣囊病是很少見的疾病，其特徵是在腸子的黏膜下層及漿膜下層出現充滿氣體的囊泡。在囊泡中的氣體可能是氮氣、氫氣或二氧化碳。但造成此種現象的原因仍然未明。我們在此報告一例：79歲慢性肺氣腫婦人，因為左下腹脹氣及大便習慣改變。在大腸鏡下呈現“一碰即出血般”的囊泡變化，病人接受切除有病變的氣囊腸段，並且順利出院。回顧文獻，腸氣囊病以吸高濃度氧氣治療為主，但其復發率報告可高達 50-78%，因此我們認為老年人或有局部腹膜炎現象病人，在其病情惡化或復發前，手術切除將是另一種選擇方式。(長庚醫誌 2003;26:144-7)

關鍵字：腸氣囊病。

行政院衛生署台南醫院 一般外科，長庚紀念醫院 高雄院區 外科部大腸直腸外科，²病理部

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索取抽印本處：陳鴻華醫師，長庚紀念醫院 大腸直腸外科。高雄縣833鳥松鄉大埤路123號。Tel.: (07)7317123轉8094; Fax: (07)7328229