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CASE REPORT

Pneumatosis cystoides intestinalis

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pneumatosis cystoides intestinalis

Summary Pneumatosis cystoides intestinalis is a rare disease with an acute or subacute clinical presentation. Here, we present two cases seen at our institution over a 5-year period. The first patient underwent emergent laparotomy due to abdominal extraluminal free air caused by ruptured pneumatosis cystoides intestinalis, which may be related to the use of alpha-glucosidase inhibitors. No perforation of the ascending colon was identified during the operation. Multiple sigmoid pneumocysts mimicking colonic polyposis were found in the second patient, who underwent sigmoidectomy for excluding colonic malignancy. We emphasize the importance of correct diagnosis of pneumatosis cystoides intestinalis and optimal management to prevent unnecessary surgical treatment.

1. Introduction

Pneumatosis cystoides intestinalis is a rare condition characterized by gas-filled cysts within the mucosa, submucosa, or subserosa of the bowel wall.

It typically presents in the 5th to 8th decade, and is idiopathic (15%) or secondary (85%), manifesting itself in a wide variety of gastrointestinal (e.g., pyloric stenosis, sigmoid volvulus, ulcerative colitis, and ischemic bowel) and non-gastrointestinal (e.g., chronic obstructive pulmonary disease, depression, and systemic sclerosis) diseases.[1]

Pneumatosis cystoides intestinalis is usually a benign disease, and its etiology is still obscure. Precise knowledge of the disease with regard to its differential diagnosis, course, and treatment modalities is essential in providing optimal care for patients who present with this condition.

2. Case 1

A 76-year-old woman took an alpha-glucosidase inhibitor for diabetes mellitus type II for more than 5 years. She was admitted to the hospital with dull discomfort in the right lower chest and dyspnea on exertion for 1 day. Progressive

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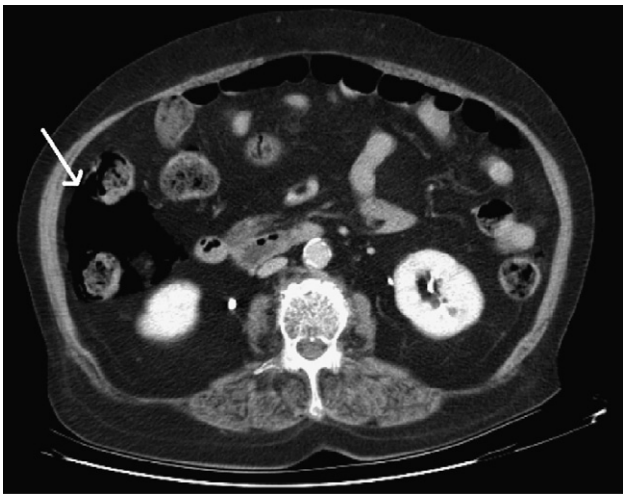

Figure 1 Abdominal computer tomography exhibiting extraluminal free air near the ascending colon without fluid accumulation (white arrow).

dull pain of the whole abdominal also occurred. Physical examination in the emergency room revealed a tender obese abdomen and muscle guarding. Laboratory examination showed leukocytosis. Abdominal computer tomography exhibited extraluminal free air accumulated near the ascending colon without obvious surrounding fat stranding or intraperitoneal fluid (Fig. 1). At laparotomy, multiple encysted gases of the ascending colon were found, without any perforation or fecal leakage. Right hemicolectomy with end-to-side anastomosis was performed. A histopathologic examination revealed features of pneumatosis cystoides intestinalis with varying-sized empty cysts in the submucosa (Figs. 2 and 3). Although focal mucosal hemorrhage and mucosal swelling were identified, no tumor, perforation, or purulent exudate on the serosa was seen. The patient was discharged uneventfully, and there has been no recurrence of pneumatosis cystoides intestinalis.

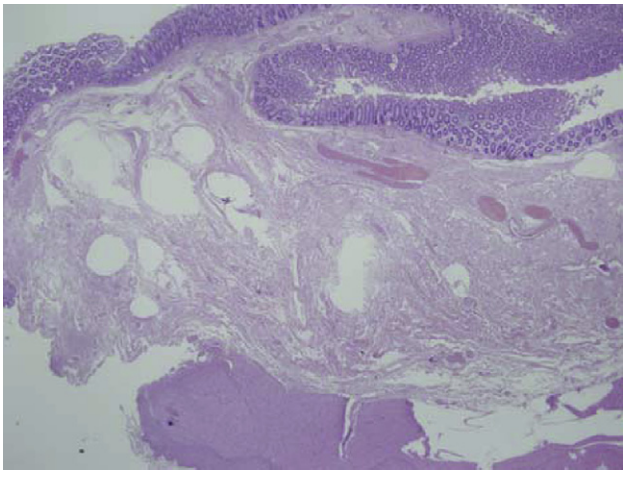

Figure 2 Photomicrograph of varying-sized empty cysts in the submucosa (white arrow) (hematoxylin & eosin, original magnification: 20×).

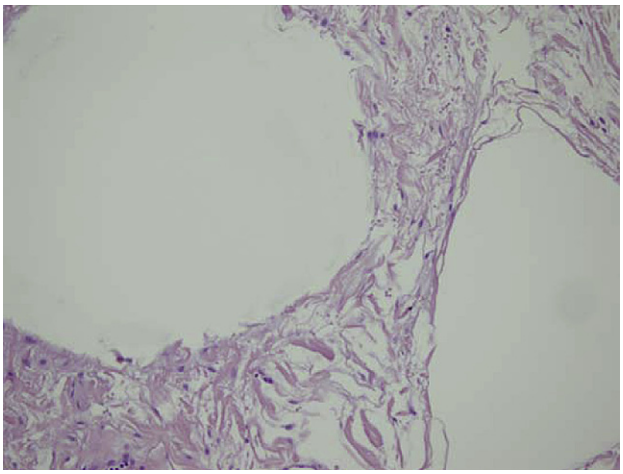

Figure 3 Cysts lacking an epithelial lining, surrounded by flattened cells or multinucleated histiocytes (hematoxylin & eosin, original magnification: 200×).

3. Case 2

A 78-year-old woman was seen at the outpatient department owing to loose stools at a frequency of five times daily for 6 months. She had no family history of colon tumor. She did not complain of body weight loss, hematochezia, abdominal fullness, or abdominal pain. Abdominal examination revealed a soft abdomen without distention. No rectal tumor was found by digital examination. Her stool occult blood test was positive. A barium enema examination revealed multiple polypoid lesions at the sigmoid and rectosigmoid junctions (Fig. 4). A colonoscopy examination also showed multiple elevated lesions with a smooth surface and transparent lesions, 3—6 mm in size, in the sigmoid colon (Fig. 5). A biopsy was not done to prevent iatrogenic colonic perforation. No specific finding was identified by computer tomography. Sigmoidectomy was performed for excluding sigmoid malignancy and persistent diarrhea. Pneumatosis cystoides intestinalis was confirmed histologically (Fig. 6). The patient had a complete recovery, and no recurrent pneumocyst was found by the follow-up colonoscopy.

4. Discussion

Pneumatosis cystoides intestinalis is a rare disease with an acute or subacute clinical presentation. The mechanisms of pathogenesis and etiology of pneumatosis cystoides intestinalis are not fully understood. The disease is probably caused by a combination of associated diseases exerting elevated intraluminal pressure and inflicting mucosal damage, allowing gas-forming microorganisms to enter the bowel wall, thus forming cysts.[2]

Case reports have documented pneumatosis cystoides intestinalis in patients taking alpha-glucosidase inhibitors, which involve intestinal gas production through fermentation by the intestinal flora of carbohydrates, of which absorption is inhibited by alpha-glucosidase inhibitors.[3]

This factor, along with peristaltic hypofunction associated with diabetic autonomic neuropathy, may lead to

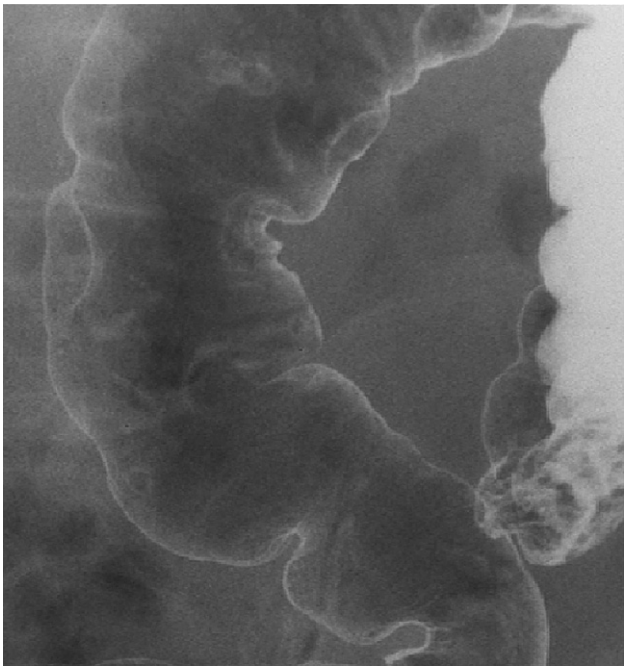

Figure 4 Fluoroscopic and film examinations with double contrast barium enema of the lower gastrointestinal tract showing multiple bowel filling defects at the sigmoid colon and smooth passage of barium without obstruction.

raised intraluminal pressure, allowing the gas-producing bacteria to invade the colonic mucosa through mucosal breaks, forming pneumocysts.[4]

Our first patient had abdominal extraluminal free air caused by ruptured pneumatosis cystoides intestinalis, which was considered to be probably related to the use of alpha-glucosidase inhibitors after we excluded other possible factors inducing pneumatosis cystoides intestinalis.

Histologically, the cysts of pneumatosis cystoides intestinalis are pseudocysts because they lack an epithelial lining. However, they may become surrounded by a rim of histiocytes, multinuclear giant cells, lymphocytes, neutrophils, eosinophils, granulomas, and fibrosis, especially after they collapse.[5]

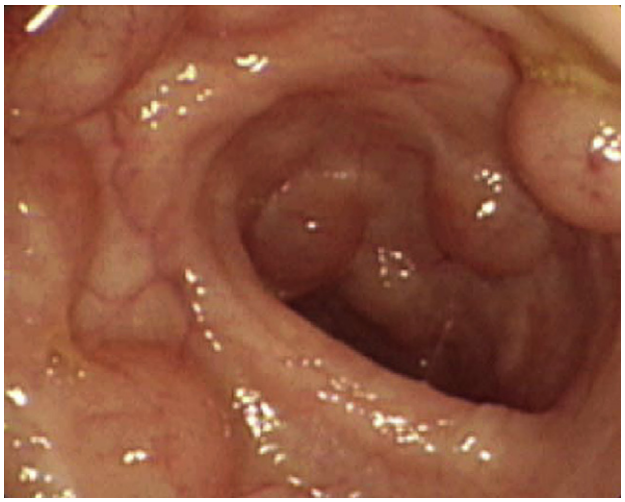

Figure 5 Colonoscopy showing multiple submucosal cysts with smooth surface at the sigmoid colon.

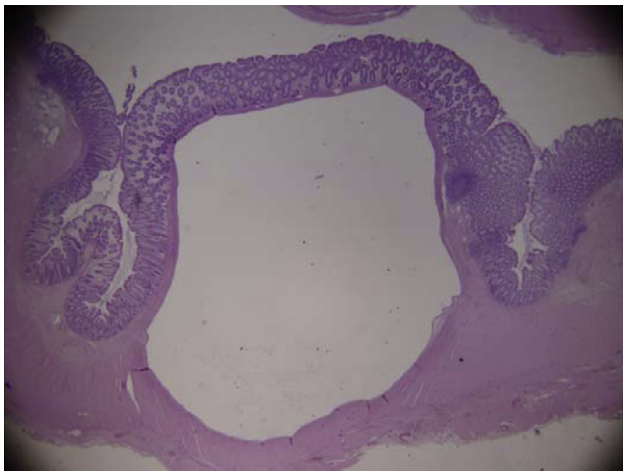

Figure 6 A submucosal cyst lined by a rim of histiocytes (hematoxylin & eosin, original magnification: 20×).

Depending on the location of the gas-filled cysts, the symptoms in each patient may vary enormously. This disease can present with profound disturbances of bowel function, including diarrhea, constipation, rectal bleeding, passage of mucus, vague abdominal discomfort, weight loss, and excessive flatus.

Generally, pneumatosis cystoides intestinalis can be asymptomatic and incidentally found at endoscopy or laparotomy.

Typically, plain abdominal radiographs may disclose intramural gas collections and even free intraperitoneal air if the cysts are perforated.

An endoscopic examination could exhibit "polypoid" intraluminal masses corresponding to pneumocysts, which should not be misdiagnosed for a diffuse polyposis. Pneumatosis cystoides intestinalis remains a challenging diagnostic problem on biopsy material, except when a deep biopsy allows differential diagnosis with colonic polyposis. Because the cysts may rapidly deflate after the biopsy, this presentation provides an important clue for diagnosis.[6] Contrast studies of the gastrointestinal tract can demonstrate filling defects within the intestinal lumen corresponding to protrusion of submucosal gas-filled cysts. Characteristic findings of pneumatosis cystoides intestinalis by computer tomography include circumferential collections of air adjacent to the lumen of the bowel that run in parallel with the wall of the bowel. However, those findings also mimic transmural infarction of the bowel in patients with intestinal ischemia.[7] Patients with life-threatening bowel ischemia or perforation may present with metabolic acidosis, systemic inflammatory response syndrome, and hepatic portal vein gas. Consideration of both image findings and clinical presentation could help in the differential diagnosis.

Because the illness is usually a self-limited condition, and intramural gas cysts may disappear spontaneously over time, no specific therapy is, in most cases, needed, other than treatment of the underlying disease of pneumatosis cystoides intestinalis for patients who are asymptomatic or minimally symptomatic. Symptomatic pneumatosis cystoides

intestinalis often responds to a conservative management, for example, oxygen therapy, administration of an elemental diet, antibiotic therapy, or discontinuation of the alpha-glucosidase inhibitor. Hyperbaric oxygen could be used to reduce the duration of oxygen administration and the potential risk of oxygen toxicity to the lung. When needed, hyperbaric oxygen therapy is the treatment of choice if there is no contraindication.[8]

Failure in recognition and treatment of the underlying disease of pneumatosis cystoides intestinalis will result in a high recurrence rate (50—78%).[9]

It is also important to consider this disease in the context of acute and subacute abdominal symptoms as it sometimes presents as a pneumoperitoneum due to ruptured cysts. Patients with free intraperitoneal air usually undergo emergency surgery, such as the first reported case here. However, some of these patients will have no identifiable perforation, for example, in spite of the presence of pneumatosis cystoides intestinalis.

Failure to recognize the specific colonoscopic feature of pneumatosis cystoides intestinalis in the second patient led to an unnecessary colectomy.

Therefore, surgery should be avoided unless there are signs of severe inflammation, sepsis, metabolic acidosis, or portal venous gas, which are indicators of bowel ischemia or bowel perforation. However, in patients who present with equivocal symptoms, or have indistinct image findings, a correct preoperative diagnosis of pneumatosis cystoides intestinalis is very difficult.

The most important tasks of the physician include: (1) full recognition of the entity of pneumatosis cystoides intestinalis so that patients are not misdiagnosed and mismanaged as having malignancy or polyposis; and (2) proper differentiation of the benign noncomplicated cases from life-threatening forms (bowel necrosis, perforation, and infections), in which immediate surgery is necessary, and which are associated with high mortality.

In conclusion, pneumatosis cystoides intestinalis is a rare clinical condition, usually benign, in which surgery should be avoided. The treatment is effective with oxygen therapy and other conservative management. Further care should be directed at identifying, treating associated diseases, discontinuing certain medications, and excluding causes, such as bowel ischemia. However, we must emphasize that the correct preoperative diagnosis of pneumatosis cystoides intestinalis is often very difficult, sometimes necessitating surgery for a definitive diagnosis.

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