Case Report

Pneumatosis Cystoides Intestinalis

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We herein present a case of pneumatosis cystoides intestinalis. A 56-year-old woman was admitted to Nagasaki Prefectural Shimabara Hospital with diffuse and mild abdominal pain. A plain abdominal X-ray revealed free air in the right subphrenic space, and computed tomography showed an extraluminal gas-filled lesion adjacent to the small intestine. With a tentative diagnosis of perforation of the small intestine, a laparotomy was performed, although she had little tenderness and no rigidity on physical examination. Upon opening the peritoneal cavity, multiple bullae-like cysts were noted on approximately one meter of the ileal serosa; however, no site of perforation was detected. Removal of the portion of what appeared to be the affected bowel was the procedure of choice. The resected specimens histologically showed pneumatosis cystoides intestinalis without any perforation. Her postoperative course was uneventful and she has been doing well with no evidence of recurrence as of the end of June 2005.

Keywords: Pneumatosis; Cystic lesion; Intestine

Introduction

Pneumatosis cystoides intestinalis (PCI) is a condition characterized by multiple gas cysts in the submucosa and/or the subserosa of the intestinal wall and occurs in both the colon and the small intestine. We herein report a case of PCI occurring in the ileum which we treated as a perforation of the small intestine, presenting as pneumoperitoneum.

Case report

A 56-year-old woman was admitted to Nagasaki Prefectural Shimabara Hospital with diffuse and mild abdominal pain. There was no history of chronic obstructive pulmonary disease. Physical examination revealed mild tenderness in the abdominal wall without rigidity. The results of the complete blood count were as follows: erythrocytes 363/mm³; leucocytes 9,200/mm³; hemoglobin 10.6 g/dL, C-reactive protein was 2.67 mg/dL (normal range: 0-0.3 mg/dL). Plain abdominal X-ray showed free subphrenic gas, although no perforations were found in the upper or lower gastrointestinal tracts. Computed tomography (CT) demonstrated an extraluminal gas-filled lesion of the small intestine without ascites (Figure 1). Portomesenteric venous gas was not present. With a tentative diagnosis of perforation of the small intestine, a laparotomy was performed on January 7, 2003. During surgery, multiple cysts, which looked like pulmonary bullae, were found at approximately one meter from the ileal end. Although a careful investigation was conducted, no perforations were detected in the gastrointestinal tract (Figure 2). Since cystic lesions were grossly noted in the ileum, this portion was considered to be the place where the perforation had occurred. Therefore, resection of the ileum was the procedure of choice in the present case. The resected specimens revealed numerous bullae-like lesions on the serosal surface; however, no perforative sites were identified (Figure 3). Histological examination revealed the bullae-like cysts distributed in the subserosa and intramuscular layer, and partly in the submucosal layer of the intestinal wall (Figure 4). These lining cells were not observed in the intestinal mucosa. The patient's postoperative course was uneventful and she was doing well as of the end of June 2005.

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Discussion

The present case was defined as primary PCI because of the absence of any underlying diseases such as acute or chronic inflammatory bowel diseases, collagen diseases, or chronic obstructive pulmonary diseases. The frequency of primary PCI has been reported to be approximately 15% of all cases of PCI. The main theories of etiology can be grouped as either mechanical or bacterial, although the etiology and pathogenesis of PCI are still unclear. In our case, there were no roentgenological or pathological findings supporting the mechanical theory. We did not conduct any bacterial studies.

Diagnosing PCI on plain abdominal X-ray is difficult, especially in the small intestine, although Boerner et al. reported that typical radiolucent findings such as grape-like clusters or honeycomb-shaped shadows along the contours of the bowel can be seen. However, CT is probably the most useful tool for diagnosis of small intestinal pneumatosis. Some authors have reported that information on the presence or absence of pneumatosis intestinalis could be obtained in the lung window setting, including information on the affected bowel segments.

For the treatment of PCI, oxygen therapy, which was first described by Forgacs et al. in 1973, has traditionally been recommended for the relief of pneumatosis. Patients with PCI were reported to have successfully been treated with continuously high concentrations of oxygen. Oxygen therapy has been immediately effective in the reduction of symptoms and disappearance of cysts, but recurrence has been reported in 50 to 78% of the patients thus treated.

Pneumoperitoneum in PCI does not necessitate an emergent
laparotomy.\textsuperscript{14,15} Pneumatosis on CT does not always indicate transmural necrosis of the bowel. In the present case, however, the patient had symptomatic free subphrenic air with positive C-reactive protein, although signs of neither panperitonitis nor leukocytosis were present. Under the suspected perforation of the small intestine, a decision was eventually made to perform an emergent laparotomy. Aside from the pertinence of the laparotomy for this case, it is important to be aware that free gas on abdominal X-ray can indicate pneumatosis-related pneumoperitoneum.\textsuperscript{14,15} If we had had such information about PCI, we might not have performed emergent laparotomy.

In conclusion, when extraluminal gas-filled cysts with pneumoperitoneum are present, physicians should take the possibility of the PCI rupture into account.

References