CASE REPORT

Recurrent extensive idiopathic pneumatosis intestinalis of the small intestine with pneumoperitoneum: Effective treatment with the surgical intervention

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Abstract

A 72-year-old man complained of a 7-days' history of abdominal distension, diarrhea, and anorexia. The chest and abdominal plain films, and the abdominal computed tomography (CT) showed a large amount of intra-abdominal free air, and the gas in the wall of the small intestine and the mesentery. Viewing the CT images with the lung window clearly revealed the presence of a huge gas in the small intestinal wall and the mesentery, the intramural gas spreading approximately 1 m in the length. There were no signs of inflammation. He was diagnosed to have extensive idiopathic pneumatosis intestinalis with pneumoperitoneum. The symptoms were relieved by the conservative treatments. The intestinal pneumatosis, however, recurred shortly after starting the oral feeding. The pneumatosis appeared to spread from the localized region of the jejunum. Then, the laparoscopy-assisted partial resection of the jejunum (30 cm in the length) was performed. Pathological examination of the removed specimen revealed the presence of multiple gas-filled cysts in the jejunal wall, but no breaks on the mucosal surface. The cysts were not lined by epithelial cells, but were surrounded by many macrophages. He became completely free of the pneumatosis after the surgery. The findings indicate that the surgical intervention should be considered in certain patients with recurrent extensive idiopathic pneumatosis intestinalis.

Key words

Pneumatosis intestinalis, Pneumoperitoneum, Surgical intervention

1 Introduction

Pneumatosis intestinalis (PI) is a rare disorder characterized by the presence of multiple gas-filled cysts within the wall of the small or large intestine [1, 2]. PI is often detected on the abdominal plain films, approximately two-thirds of affected patients being positive on the plain films [2]. The abdominal computed tomography (CT) is more sensitive than the plain films to detect the presence of PI, and is also helpful to detect the underlying disorders. The intra-abdominal free air

(pneumoperitoneum) associated with PI may result from the rupture of subserosal blebs, or may be associated with the life-threatening conditions such as bowel necrosis and perforation [3]. Although the surgical intervention is not required for most patients with idiopathic PI, it should be considered in certain patients, who remain symptomatic despite the conservative treatments or suffer from recurrent PI [1, 4]. Here we report a case with recurrent extensive idiopathic PI of the small intestine with pneumoperitoneum, who was effectively treated by the surgical intervention (partial resection of the jejunum). We also show that viewing the abdominal CT images with the lung window, but not the abdominal window, clearly revealed the presence of gas in the small intestinal wall and the mesentery.

2 Case report

A 72-year-old man visited his family doctor, complaining of a 7-days' history of abdominal distension, diarrhea, and anorexia. The chest and abdominal plain films showed a large amount of intra-abdominal free air, suggesting possible perforation of the gastrointestinal tract. Then, he was referred to our hospital. On presentation, his body temperature was 36.6°C, blood pressure 102/72 mmHg, heart rate 72/min, and oxygen saturation 98% while he was breathing ambient air. His lungs were clear to auscultation. His cardiac examination was normal, without a murmur or rub. His abdomen was distended, but there were no tenderness, rebound tenderness, or muscular guarding. He had a history of cerebral infarction, benign prostatic hypertrophy, gastric cancer (the early stage) treated by the endoscopic submucosal dissection 2 years ago, and cholecystolithiasis treated by laparoscopic cholecystectomy 1 year ago. He had a medication with warfarin, tamsulosin, frosemide, and spironolactone.

Red blood cell count was $4.37 \times 10^6/mm^3$, hemoglobin level 14.0 g/dL, hematocrit 40.8%, platelet count $159{,}000/mm^3$, and white cell count $4{,}300/mm^3$. Prothrombin time was 15.6 s (international normalized ratio 1.37), activated partial thrombin time was 38.0 s, and fibrinogen level was 295 mg/dL. The serum C-reactive protein level was 0.88 mg/dL (normal < 0.3 mg/dL), blood urea nitrogen 20.2 mg/dL, creatinine 1.06 mg/dL, sodium 137 mmol/L, potassium 4.6 mmol/L, chloride 102 mmol/L, calcium 8.5 mg/dL, total protein 7.1 g/dL, albumin 3.9 g/dL, aspartate aminotransferase 28 U/L, alanine aminotransferase 19 U/L, alkaline phosphatase 423 U/L, and total bilirubin 1.1 mg/dL.

The chest and abdominal plain films showed a large amount of intra-abdominal free air under the diaphragm bilaterally (see Figure 1a and 1b). The abdominal plain film also showed dilatation of the small intestine and circular collections of the gas in the intestinal wall (see Figure 1b). Viewing the abdominal CT images with the abdominal window showed a large amount of intra-abdominal free air (see Figure 1 c and e), whereas viewing them with the lung window clearly revealed the presence of a huge gas in the small intestinal wall and the mesentery (see Figure 1 d and f). The intramural gas in the small intestine appeared to spread approximately 1 m in the length. The small intestine was dilated, and the air-fluid level was detected in the lumen of the small intestine (see Figure 1 c-f). No portal venous gas was detected (see Figure 1g). A small amount of ascites was found in the pelvic cavity (see Figure 1h).

He was diagnosed to have extensive, presumably idiopathic, PI with pneumoperitoneum, because there were no signs or symptoms strongly indicating perforation of the gastrointestinal tract and peritonitis. Then, he received the conservative therapy with intravenous administration of fluids for 13 days and antibiotics (cefmetazole) for 2 days, respectively [4, 5].

Oral feeding was allowed on day 7 after his admission, since the intra-abdominal free air as well as the intramural and intra-mesenteric gas was significantly decreased (see Figure 2a). No worsening of the symptoms or signs was observed during 5 days of oral feeding (see Figure 2b). The clinical course was uneventful, and he was discharged free of any symptoms.

Two months after his discharge, he visited our hospital again, complaining of abdominal distention and weight loss over the preceding several weeks. The abdominal plain film and the CT images revealed the recurrence of the small intestinal pneumatosis (see Figure 3a). He also complained of diarrhea. The culture of feces, however, did not detect any pathologic

bacteria responsible for diarrhea. These findings and no signs of inflammation taken together suggest that diarrhea results from the intestinal pneumatosis but not intestinal infections. The recurrence of PI after the short duration suggests that there might be certain lesions in the small intestine, which might be responsible for the recurrence of PI. It is possible that the intra-luminal gas may penetrate into the intestinal wall through the breaks on the mucosal surface, leading to extensive intestinal pneumatosis. He was readmitted on the diagnosis of recurrent PI, and the surgical intervention was considered. He was initially treated with the elemental diet and the central venous nutrition. On day 10 after his admission, the intramural gas of the small intestine completely disappeared with a small amount of gas remaining in the mesentery on CT (see Figure 3b), making it difficult to determine the primary lesion that should be removed by the surgical intervention. Then, oral feeding was allowed. On day 7 after starting the oral feeding, the intramural gas was detected again in the localized region of the small intestine (see Figure 3c). On day 9 after starting the oral feeding, the gas spread into the mesentery of the small intestine (see Figure 3d), and the laparoscopy-assisted operation was performed.

The multiple gas-filled cysts of approximately 1 mm in diameter were found on the surface of the mesentery of the jejunum (see Figure 4a), whereas the cysts were not found on the jejunal surface. The jejunum, however, showed diffuse edema, and the jejunum adjacent to the involved mesentery felt like the tissue paper or Rice Krispies, strongly indicating the presence of gas in the jejunal wall. The jejunum of 30 cm in the length (the involved jejunum of 15 cm and the healthy jejunum of 15 cm, respectively) was removed (see Figure 4b). The postoperative course was uneventful, and he became completely free of PI after the surgery.

Pathological examination of the removed specimen revealed the presence of multiple gas-filled cysts in the submucosa, the muscularis propria, and the subserosa of the jejunum (see Figure 4c). The cysts did not communicate with the lumen of the intestinal wall, and no breaks were found on the mucosal surface. The cysts were not lined by epithelial cells, but were surrounded by many macrophages (see Figure 4d). Multinuclear giant cells were also found, but no bacteria were detected in the specimen. The possible primary lesions such as the breaks on the mucosal surface, which could cause PI, were not detected by the pathological examination. Then, the diagnosis of idiopathic PI was finally made on the basis of the clinical and pathological findings.

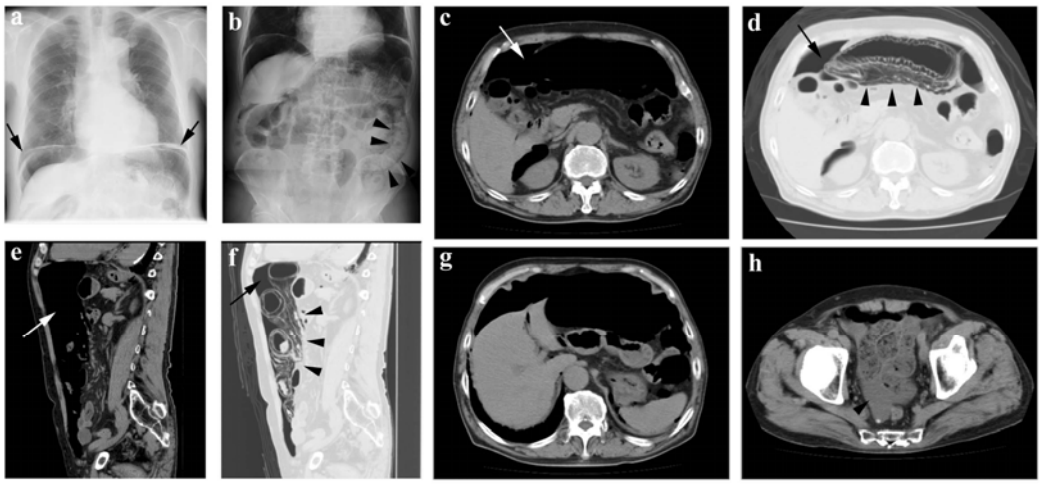

Figure 1. (a, b) The plain films showed a large amount of intra-abdominal free air under the diaphragm bilaterally (arrow), and circular collections of the gas in the intestinal wall (arrowhead). The axial and sagittal CT images viewed with (c, e) the abdominal window and (d, f) the lung window showed a large amount of intra-abdominal free air (arrow), and the gas within the small intestinal wall and the mesentery (arrowhead). The small intestine was dilated, and the air-fluid level was detected in the lumen of the small intestine. (g) No portal venous gas was detected. (h) A small amount of ascites was found in the pelvic cavity (arrowhead)

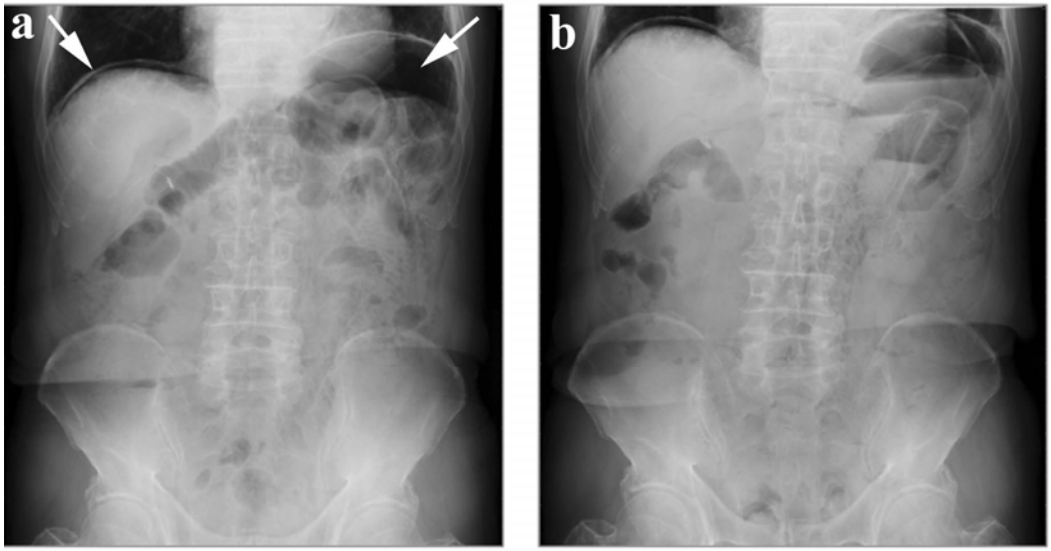

Figure 2. (a) The plain film on day 7 after his admission. The intra-abdominal free air as well as the intramural and intra-mesenteric gas was significantly decreased (arrow). (b) The plain film on day 5 after starting the oral feeding

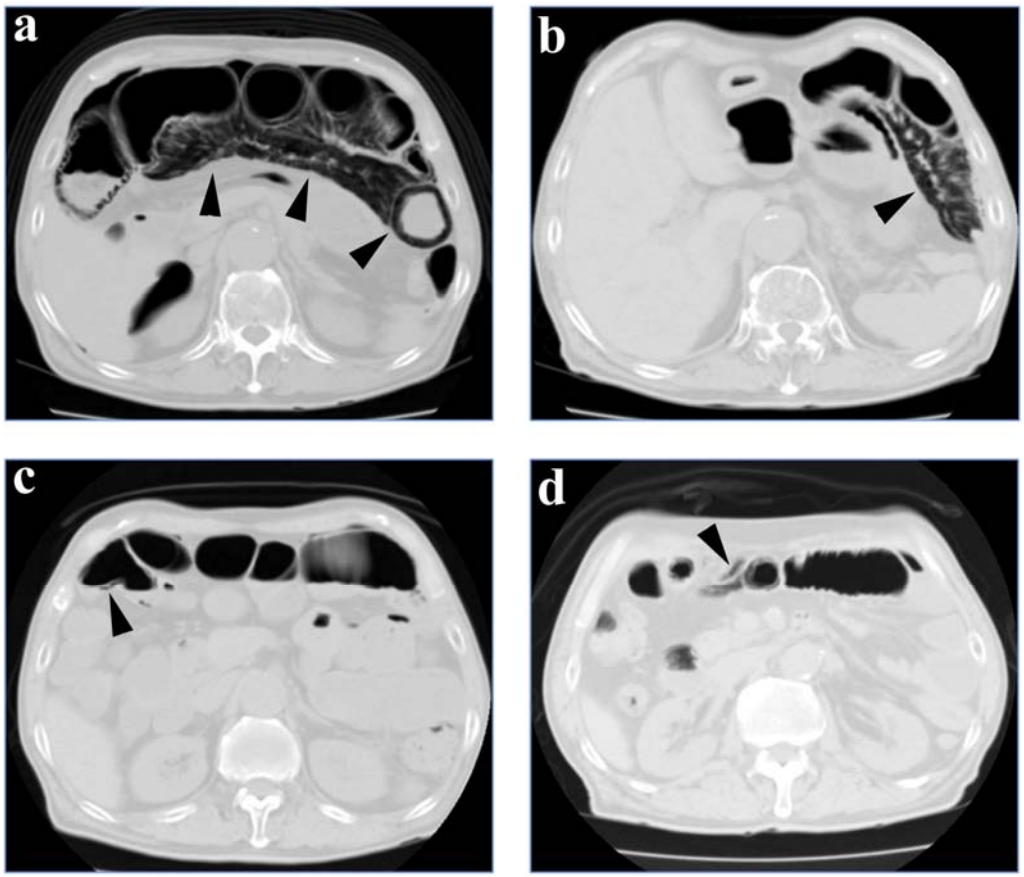

Figure 3. (a) The axial CT images on the day of his readmission showed a large amount of gas in the small intestinal wall and the mesentery (arrowhead). (b) On day 10 after his readmission, the intramural gas of the small intestine completely disappeared with a small amount of gas remaining in the mesentery (arrowhead). (c) On day 7 after starting the oral feeding, the intramural gas was detected again in the localized region of the small intestine (arrowhead). (d) On day 9 after starting the oral feeding, the gas (arrowhead) spread into the mesentery of the small intestine

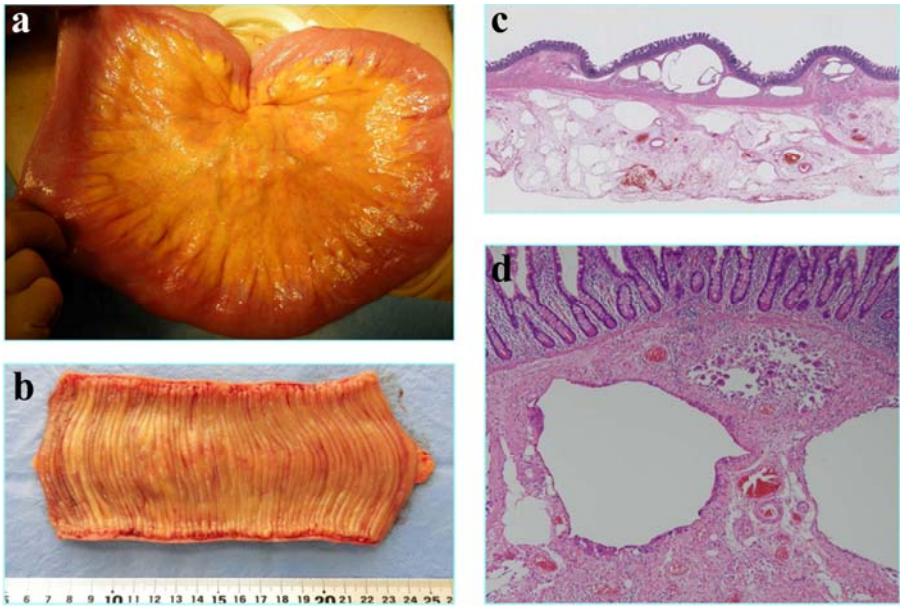

Figure 4. (a) The multiple gas-filled cysts of approximately 1 mm in diameter were found on the surface of the mesentery of the jejunum. (b) The jejunum of 30 cm in the length was removed. No breaks were found on the mucosal surface. (c) Pathological examination revealed the presence of multiple gas-filled cysts in the submucosa, the muscularis propria, and the subserosa of the jejunum. (d) The cysts were not lined by epithelial cells, but were surrounded by many macrophages

3 Discussion

PI may occur in infants and adults. Most cases in infants are secondary to necrotizing enterocolitis, whereas PI in adults is often observed in the fifth to eighth decade, and is idiopathic (15%) or secondary (85%) to various disorders [6, 7]. Mortality rates are high in the cases of PI associated with the disorders that lead to bowel necrosis or perforation. In these cases, urgent surgical intervention is required. By contrast, the prognosis is generally good in patients with idiopathic PI, and the surgical intervention is not required in most cases [1, 4-9]. The present case shows extensive idiopathic PI with pneumoperitoneum, which recurs shortly after starting the oral feeding. He became completely free of PI after partial resection of the jejunum. In this case, the surgical intervention was highly effective, supporting the notion that the surgical intervention should be considered in certain cases with recurrent extensive idiopathic PI.

The pathogenesis of PI is poorly understood, and several hypotheses, including the mechanical, bacterial, and biochemical theories, have been proposed to explain accumulation of the gas in the intestinal wall [1, 10, 11]. PI could be caused by the gas-producing anaerobic bacteria when they gain access to the submucosa through the breaks on the mucosal surface. The present case showed no signs or symptoms indicating bacterial infections or inflammation of the intestine, and no breaks were found on the mucosal surface, precluding the possibility of the bacterial theory. It has been proposed that the intra-luminal bacteria produce excessive amounts of hydrogen gas through fermentation of carbohydrates, and the gas may be forced directly through the mucosa for accumulation of the gas in the intestinal wall. Such a case has been reported in some patients with diabetes mellitus taking α-glucosidase inhibitors [12]. The present case did not take any medications that might cause PI, precluding the possibility of the biochemical theory. The present case was effectively treated by partial resection (30 cm in the length) of the jejunum, suggesting that PI may be caused by the localized pathological condition of the jejunum; for example, the penetration of the intra-luminal gas into the jejunal wall through the injured mucosa, a notion that may support the mechanical theory. The intra-luminal gas could penetrate into the intestinal wall under certain conditions such as intestinal ischemia, intestinal infections, and inflammatory bowel diseases, in which the mucosal

integrity is disrupted [13]. Although we could not find out any breaks on the mucosal surface of removed specimen, we cannot exclude the possibility that the disrupted mucosal integrity was already repaired in the removed specimen.

Viewing the abdominal CT images with the lung window, but not the abdominal window, clearly revealed the presence of gas in the intestinal wall and the mesentery. The intramural gas showed circumferential collections of air adjacent to the intestinal lumen that run in parallel with the intestinal wall on CT. These findings indicate that the abdominal CT images should be viewed with the lung window when PI is suspected. The intramural gas shows a low-density linear or bubbly pattern on the plain films [14]. Complications such as pneumoperitoneum, secondary bowel obstruction, volvulus, intussusceptions, and hemorrhage are reported to occur in approximately 3% of PI patients [11]. It should be noted that the emergent surgical intervention is not necessarily required even for the patients exhibiting the intra-abdominal free air, and the conservative therapy is generally recommended for most patients with idiopathic PI [1, 3]. The surgical intervention, however, should be considered in certain patients with recurrent extensive idiopathic PI, such as the current case, as well as the patients with life-threatening conditions.

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