A Case of Pneumatosis Cytoides Intestinalis Successfully Treated by Inhalation of High Concentration Oxygen

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Abstract
A 44-year-old man was referred to our hospital because of a positive fecal occult blood test. A barium enema study revealed numerous oval-shaped, elevated lesions with smooth surfaces in the region including the ascending colon. He was admitted to our hospital for investigation and therapy. Based on colonoscopic examination, we diagnosed him as having pneumatosis cytoides intestinalis (PCI). He was treated with oxygen (5L/min × 5 hours/day for 14 days) via a nasal cannula. Most of the multiple cysts diminished and some changed into white scars. The simplicity of oxygen therapy supports its use as a first-line treatment.

Key words  Pneumatosis cytoides interstinalis, High-concentration oxygen inhalation

Introduction
Pneumatosis cystoides intestinalis (PCI) is a relatively rare condition in which numerous gas-filled cysts mainly containing nitrogen are formed within the intestinal wall. While idiopathic and secondary cases are known, the latter may result from exposure to trichloroethylene, chronic respiratory diseases such as pulmonary emphysema, and collagen disease. We report our experience with a case of PCI, which was detected by workup following a positive fecal occult blood test, including literature-based discussion of this disease.

Case
Patient: 44-year-old male.
Chief complaint: None.
Medical history: Not remarkable.

Family history: Not remarkable.
Past exposure: No past exposure to trichloroethylene.
Present illness: The patient was found to be positive for fecal occult blood on a health screening, and was referred to our hospital for detailed examination. An outpatient barium enema study revealed numerous oval-shaped, elevated lesions with smooth surfaces in the region from the hepatic flexure to the ascending colon. The patient was hospitalized for investigation and therapy.

Condition at the time of hospitalization:
Height 165 cm, body weight 67 kg, blood pressure 136/74 mmHg, heart rate 72/min, regular pulse, no signs of anemia in palpebral conjunctiva, no jaundice in bulbar conjunctiva, no abnormal findings in cardiopulmonary auscultation, no palpable hepatosplenomegaly or masses in the abdomen.

Laboratory findings at the time of hospital-
increase in total bilirubin (2.24). Antinuclear antibodies, anti-RNP antibodies, anti-Scl-70 antibodies, and anti-Jo-1 antibodies were negative. Blood gas analyses were within normal ranges, including pH 7.412, PCO₂ 40 mmHg, PO₂ 90.9 mmHg, BE 0.9, and SaO₂ 98.2%.

Abdominal X-ray at the time of hospitalization (Fig. 1): Aggregation of numerous round-shaped transparencies was seen in the region from the hepatic flexure to the ascending colon.

Abdominal CT at the time of hospitalization (Fig. 2): Abnormalities were not found in general hematology tests. Blood chemistry tests showed no abnormalities except for a slight

<table>
<thead>
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<th>Table 1 Laboratory findings at the time of hospitalization</th>
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<td>RBC 552 × 10⁴/mm³</td>
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<td>Hb 17.0 g/dl</td>
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<td>Ht 48.5%</td>
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<td>Plt 18.9 × 10⁴/mm³</td>
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<td>WBC 5280/μm³</td>
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<tr>
<td>TP 7.3 g/dL</td>
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<td>Alb 4.3 g/dL</td>
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<tr>
<td>T. Bil 2.24 mg/dL</td>
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<tr>
<td>AST 22 U/L</td>
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<td>ALT 31 U/L</td>
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<td>ALP 226 U/L</td>
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<td>LAP 127 mU/ml</td>
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<td>γ-GTP 17 U/L</td>
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<td>CHE 333 U/L</td>
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<td>LDH 135 U/L</td>
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<td>CPK 90 U/L</td>
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Fig. 1 Abdominal X-ray

Fig. 2 Abdominal CT
PCI SUCCESSFULLY TREATED BY INHALATION OF HIGH CONCENTRATION OXYGEN

the patient as having PCI and commenced intermittent high-concentration oxygen inhalation. Oxygen was administered via a nasal cannula at the rate of 5 L/min, 5 hours/day for 2 weeks.

Colonoscopy on the day after the end of oxygen therapy (Fig. 5): Most of the multiple cysts had diminished, although some small reddish elevated lesions remained.

Colonoscopy 8 weeks after the end of oxygen therapy (Fig. 6): Multiple cysts had largely disappeared and only white scars were observed.

(Fig. 2): Although air was depicted in the intestinal wall in the hepatic flexure, no wall thickening or other signs of inflammation were seen.

Barium enema X-ray at the time of hospitalization (Fig. 3): Diffusely distributed elevated lesions with smooth surfaces were seen in the region from the hepatic flexure to the ascending colon.

Colonoscopic findings (Fig. 4): Numerous multiple cysts were seen in the region from the ascending colon to the hepatic flexure.

Based on the above findings, we diagnosed
Discussion

PCI was first described by Du Vernoi in 1730. The first report of this disease in Japan was made by Miwa in 1901, and according to Matsuoka et al., there were 509 cases reported by 1997. Although it was previously considered to occur preferentially in the ileum, recent reports suggest a high occurrence in the large intestine, in particular the sigmoid colon. While older reports indicated higher prevalence in males than females, recent cases include more females than males. No symptoms specific to PCI are known. Cases involving the small intestine often show abdominal distention, abdominal pain, and flatulence, while those involving the large intestine usually present bloody mucous stool, melena, and diarrhea.

The entity of PCI is the presence of gas-filled cysts in intestinal walls. An overwhelming majority of PCI cases have some underlying disease, as 15% of all cases are idiopathic and 85% are secondary to underlying disease.

The etiology of PCI has not been well clarified. Bacterial, mechanical, and chemical mechanisms have been proposed. The bacterial theory depends on the fact that necrotizing enterocolitis in children is often complicated with PCI. It presumes that the gas-generating bacteria in the intestines invade into the intestinal wall and form cysts. However, the bacterial theory seems to be disproved by the fact that gas-generating bacteria mainly produce hydrogen and methane, while the gas in PCI lesions consists of 90% nitrogen. At present, the mechanical theory is considered plausible. There are 2 possible mechanical processes causing PCI. One is the mechanism depending on intestinal factors, where the elevation of pressure in the intestinal lumen due to intestinal obstruction, surgery, endoscopy, or trauma is considered to cause infiltration of intestinal gas through minute cracks in the mucosa. The other possibility, assuming the causal involvement of the lungs, is that painless rupture of pulmonary alveolar walls may develop in such conditions as chronic obstructive pulmonary disease and the gas may enter the intestinal walls via the mediastinum, retroperitoneal tissues, and the tissues around mesenteric arteries. With respect to the chemical theory, Yamaguchi et al. reported a high occurrence of PCI among persons chronically exposed to trichloroethylene and the detection of trichloroethylene in the gas-filled cysts. In addition, there have been reports of PCI accompanying collagen disease such as systemic scleroderma. Because our case had no history of surgery or trauma, showed no abnormality in chest X-ray and blood gas analyses, and lacked a history of exposure to trichloroethylene, we considered that this case was idiopathic.

It is difficult to diagnose PCI solely based on clinical symptoms, but a physician with the knowledge of this disease can relatively easily identify this disease based on the findings from abdominal X-ray, barium enema X-ray, and endoscopy. PCI most frequently develops in the submucous tissues, and the development of PCI within the muscular layer is considered rare. In cases where the gas-filled cysts are located predominantly in the submucous layer, abdominal X-ray depicts numerous oval-shaped bubbles of various sizes like bunches of grapes along the intestinal wall, and barium enema study reveals multiple filling defects resembling holly leaves. If there are gas-filled cysts in the subserous layer, lesions are identified as froth-like clustering of small transparencies or linear transparencies along the outer perimeter of the intestines.

PCI can be treated either with surgical removal or with a conservative method using oxygen inhalation. According to the compilation by Sakashita et al., surgery has been selected for the cases showing severe melena and other symptoms, cases with suspected digestive tract perforation due to pneumoperitoneum, cases developing ileus, cases in which malignancy could not be ruled out, and cases that actually developed digestive tract perforation. With respect to oxygen therapy, Forgacs reported successful disappearance of pneumatosis after continuous inhalation of 75% high-concentration oxygen for 6 days in 1973. In Japan, Daitoku et al. reported the effectiveness of this therapy in 1980, followed by reports of high-concentration oxygen therapy using a non-breathing face mask and nasal catheter, high-pressure oxygen therapy, and a number of other methods. The mechanism of this treatment has been explained as follows: Because the gas in the cysts mainly consists of nitrogen, the elevation of arterial oxygen partial pressure caused by high-concentration oxygen...
inhalation may result in replacement of the nitrogen in the cysts with oxygen and the oxygen may gradually be absorbed in adjacent tissues, to the effect of the disappearance of gas-filled cysts. Many reports on oxygen inhalation therapy indicate that methods such as high-pressure oxygen therapy, oxygen mask, and oxygen cannula are appropriate, provided that a 200–300 mmHg elevation of PaO_2 can be achieved, and that the administration of oxygen for 5 or 6 hours per day over the period of 1 or 2 weeks provides largely satisfactory results.\textsuperscript{20,21} In our case, nearly complete disappearance of multiple cysts was achieved by oxygen administration via a nasal cannula at the rate of 5 L/min, 5 hours/day for 2 weeks.

Conservative treatment such as oxygen inhalation is considered the first-line therapy for this disease. However, patients must be followed with careful observation, since some cases may develop melena or digestive tract perforation. According to Senoo et al.,\textsuperscript{21} the disease recurred in 26\% of the patients receiving oxygen inhalation therapy. This fact emphasizes the need for long-term follow-up after treatment.

\textbf{Conclusion}

We experienced a case of idiopathic PCI in the ascending colon. High-concentration oxygen therapy for 2 weeks was effective in this case.

\textbf{References}