Case Report

Pneumatosis Cystoides Intestinalis: An Unusual Cause of Intestinal Ischemia and Pneumoperitoneum

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Pneumatosis cystoides intestinalis (PCI), with an unknown etiology, is an uncommon disease characterized by the presence of multiple gas-filled cysts within the submucosa or subserosa of the intestinal wall. Intestinal obstruction and/or perforation are relatively uncommon complications associated with PCI. The patients are often prone to misdiagnosis or mistreatment. The diagnosis of PCI is based on plain radiography or endoscopy. Multidetector computed tomography (MDCT) provides data on other intra-abdominal pathologies. Therefore, it is an important modality for the diagnosis of PCI. We present a case of PCI in a 58-year-old man affected by peritoneal free air with multidetector computed tomography imaging findings. We performed the plain film of the abdomen, and MDCT studies that showed numerous, diffuse, bubble-like intramural gas collections into the jejunum, ileum, and colon walls at the left-upper quadrant of the abdomen. MDCT findings were confirmed by surgical exploration.

Key words: Pneumatosis cystoides intestinalis – Intestinal ischemia – Pneumoperitoneum – Imaging

Pneumatosis cystoides intestinalis (PCI) is a condition in which polycystic air has entered the submucosa or subserosa of the intestine. It is a rare and usually benign disorder.^{1,2} PCI may be

considered a surrogate marker for intestinal ischemia and impending perforation. The presenting clinical entity may be very heterogeneous and represent a challenge for the clinician. Multidetector

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Fig. 1 Plain film of the abdomen shows extensive cystic gas collections (black arrows), multiple air-fluid levels, and subdiaphragmatic free intraperitoneal airs (white arrows).

computed tomography (MDCT) may confirm the diagnosis, give some additional information, provide differential diagnosis, and help in determining the primary cause or some possible coexistent complications.²⁻⁴ In this paper, we report the PCI case studied with MDCT imaging findings and confirmed by surgical exploration.

Case Report

A 58-year-old man with abdominal pain, nausea, and vomiting had abdominal distention and discomfort for 4 days. He was admitted to our radiology department for further evaluation of this abdominal distention. Plain film of the abdomen

was first performed and revealed extensive cystic gas collections and multiple air-fluid levels (Fig. 1). Due to suspicion of perforation or PCI, the patient subsequently underwent MDCT procedure in order to evaluate the intestinal walls and the extra-intestinal compartments. MDCT imaging was performed using a second-generation 256-slice CT scanner (SOMATOM Definition Flash; Siemens Healthcare, Forchheim, Germany) with 50 mL of iopromide (Ultravist 370 mg/mL; Bayer Schering Pharma, Berlin, Germany) infused intravenously at a rate of 4 mL/second. The contrast-enhanced MDCT images showed the presence of numerous gas collections within the wall of the small bowel, consisting mainly of isolated bubbles, free intraperitoneal airs, multiple air-fluid levels, and distention in the small bowel (Figs. 2A, 2B). Because a diagnosis of obstruction or perforation was suspected, surgical exploration was performed in order to confirm the diagnosis. It revealed the characteristic features of pneumatosis cystoides intestinalis: air-filled cysts of various sizes in the subserosa of the small bowel (Fig. 3). Necrosis and marked ischemia were also observed in the intestine (Fig. 4). There was no perforation of the bowel wall in the surgical exploration. Thus, we thought that free intraperitoneal air was associated with spontaneous rupture of air-containing cysts within the wall of the small bowel. The patient was treated surgically.

Discussion

Pneumatosis intestinalis was defined by Lerner and Gazin in 1946, as the presence of gas in an abnormal site of the body.⁵ PCI is a rare condition characterized by multilocular gas-filled cysts localized in the submucosa and subserosa of the gastrointestinal tract.^{1,6} The incidence of PCI is unknown. Symptoms, if any, are usually secondary to an underlying

Fig. 2 Virtual endoscopic view (A) and axial contrast-enhanced MDCT scan (B) show the presence of numerous gas collections (arrows) within the wall of the small bowel.

Fig. 3 The surgical exploration shows the characteristic features of pneumatosis cystoides intestinalis: air-filled cysts of various sizes in the subserosa of the small bowel.

disease. Nonspecific symptoms, such as abdominal discomfort, diarrhea, constipation, rectal bleeding, tenesmus, or weight loss; and severe complications, including volvulus, intestinal obstruction, tension pneumoperitoneum, bleeding, intussusception, and intestinal perforation, may be seen.^{3,4} Pneumoperitoneum and pneumoretroperitoneum can be rare complications due to rupture of the cysts.⁷ In our case, free intraperitoneal air was secondary to rupture of the cysts.

Radiological tools are important for diagnosing PCI. These include plain radiographs, ultrasonography, barium series, MDCT, MDCT colonoscopy, and magnetic resonance imaging.⁷ The x-ray is of great

importance because it is readily available in every emergency room. Cysts usually appear as radiolucent shadows, similar to a bunch of grapes, close to the intestinal lumen on radiographs.⁷ MDCT allows for consequential acquisition of images with higher quality and accuracy because of the advancement of multidetector technology.⁸ It is the most useful modality for diagnosing PCI and is important because it provides data on other intra-abdominal pathologies.^{6,10} Surgical exploration is quite useful to confirm a PCI diagnosis, if the physical examination and imaging findings are suspicious.^{7,9,10}

Conservative approaches, including nasogastric decompression, intestinal rest, antibiotic therapy, and oxygen, are recommended for patients with positive examination findings and normal biochemical parameters who are confirmed radiologically to have no intestinal ischemia or perforation. An urgent laparotomy is necessary in cases of intestinal ischemia, obstruction, intestinal bleeding, or peritonitis.^{10,11} Definitive surgery should be performed during laparotomy if necrosis, perforation, or marked ischemia is observed in the intestine, as in our patient.

In conclusion, as illustrated by this case, the correct diagnosis and management of PCI is based on the results of clinical assessment and imaging techniques. PCI also should be kept in mind as a rare cause of pneumoperitoneum. Concordantly, it is very important for the radiologist to recognize the abnormal findings on the MDCT or MRI studies for differentiating between medical and surgical causes of PCI.

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Fig. 4 Surgical specimen reveals necrosis and marked ischemia in the intestine.

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