Pneumatosis Cystoides Intestinalis Accompanied by Schistosomiasis: A Case Report

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Case Report

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Abstract

Background: Pneumatosis cystoides intestinalis (PCI) is a rare disease that occurs frequently in the large and small intestine and has no obvious clinical symptoms. It is difficult to preoperatively diagnose and is easily undiagnosed or misdiagnosed.

The main pathological features are air-bearing cysts in the intestinal submucosa, intermuscular wall and subserous membrane.

Case presentation: PCI is a rare disease, which often occurs in the large intestine and small intestine without obvious clinical symptoms. It is difficult to diagnose before operation and is easy to be missed and misdiagnosed. Duvernoi first described the disease in 1730. The main pathological features were air-bearing cysts in intestinal submucosa, intermuscular wall and subserous membrane. Conservative treatment is the first choice when there are no serious complications, and timely surgical treatment is needed for serious and life-threatening complications. This report presents the clinical and pathological analysis of PCI in a 92-year-old man. The patient was admitted to hospital because of acute peritonitis, and was diagnosed as PCI with schistosomiasis after emergency operation. Postoperative follow-up is generally in good condition.

Conclusions: The incidence of PCI is low. Because of a lack of specificity in clinical manifestations and endoscopic findings, PCI is difficult to diagnose preoperatively and is easily missed and misdiagnosed. Conservative treatment is the first choice when there are no serious complications; however, timely surgical treatment is required for serious and life-threatening complications. PCI treatment is effective and convenient, and the prognosis is optimistic. Therefore, clinicians should increase their awareness of PCI to avoid unnecessary misdiagnosis.

Background

Pneumatosis cystoides intestinalis (PCI), which was first described by Duvernoi in 1730,¹ is a rare disease, without obvious clinical symptoms, occurring frequently in the large and small intestine, respectively. It is difficult to preoperatively diagnose PCI, and may be easily missed or misdiagnosed. Conservative treatment is the first choice when there are no serious complications; however, timely surgical treatment is required for life-threatening complications. The main pathological features are air-bearing cysts in the intestinal submucosa, intermuscular wall, and subserous membrane, respectively.

Case Presentation

A 92-year-old man presented with sudden abdominal pain for more than 1 day, was admitted to the emergency department of our hospital. No obvious etiologies were noted, and the symptom persistently caused sexual aggravation, without any relief in any position or after eating. The patient experienced nausea without vomiting. Abdominal computed tomography (CT) indicated a gas-free abdominal cavity

and a digestive tract perforation (Fig. 1). He was diagnosed with peritonitis with digestive tract perforation and was admitted for further treatment. His medical history included coronary heart disease and long-term diabetes for which he was medicated with aspirin and sugar-lowering drugs (Metformin Hydrochloride Sustained Release Tablets), but reported no similar abdominal pain, gallstone disease, or kidney stones. Results of the physical examination indicated a flat abdomen, no abdominal breathing, pain in the upper and right lower abdomen, muscle guard, anti-jump pain, and a weakened intestinal tone. There was no obvious mass in the abdomen. Auxiliary examination via abdominal CT indicated the presence of free gas in the abdominal cavity, suggesting a digestive tract perforation. The small intestine had expanded due to gas and fluid accumulation; minimal oozing of the lower right abdominal tract tube was detected, and schistosomiasis intestinal disease was diagnosed. As the CT showed acute peritonitis caused by a gastrointestinal perforation, a laparotomy was performed. During the operation, at the end of the reflux, the colon was irregular and slightly thickened, with inflammatory changes. Air cysts under the mucous membrane and pulp membrane were observed approximately 15 cm from the middle sigmoid colon. The sizes varied, with a maximum diameter of approximately 2 cm and a 0.5-cm perforation surrounded by pus. No metastatic nodules were found in the pelvic cavity, liver, stomach, abdominal aorta, or the abdominal wall, respectively.

Pathological examination

The intestinal mucous membrane surface was covered with soap-bubble-like small cysts. The cyst cavities with a diameter range of 0.5–2 cm were visible on the surface mucosa, muscle wall and pulp film, respectively (Fig. 2). Mirror examination results indicated different sizes of round or fissure-like cystic cavities with a clear boundary under the mucous membrane emphysema, with mucous membrane thinning. For the most part, the sac wall was not lined with the upper skin, and a part of it was covered with flat or cubic epithelial cells (Fig. 3). The sac wall contained multi-core giant cells. There was a schistosome egg deposition in the intestinal wall (Fig. 4).

Immunohistochemical staining

Endothelial cell markers CD34, CD31 and D2-40 were expressed in the lining epithelium of the cyst wall by immunolabeling. The polynuclear giant cell CD68 was positive.

Pathological examination indicated colon sigmoideum PCI with schistosomiasis.

Postoperative follow-up

There was no recurrence, and the general condition was good. Postoperative follow-up results were good after 6 months.

Discussion And Conclusions

PCI is characterized by benign lesions. The age of onset is considerably varied, and according to age, the disease can be classified as infant- or adult-type PCI. Infant-type PCI is observed among infants within 2 years of age, while adult-type PCI is observed among patients from 40 to 70 years of age. According to the pathogenesis, the disease is divided into primary and secondary types, accounting for 15% and 85% of the total cases, respectively; the disease is attributed to various intestinal diseases and injuries³. Our case is PCI with schistosomiasis. Thus, it is a secondary type of PCI.

Though it remains unknown, the pathogenesis of PCI has been hypothesized as follows⁴: (1) The mechanical theory involves an increase in intraluminal pressure that causes mechanical damage and mucosal rupture of the intestinal wall, leading to the migration of gas from the gastrointestinal cavity to the intestinal wall; (2) the pulmonary theory considers that chronic lung diseases such as chronic obstructive pulmonary disease, asthma, and interstitial pneumonia lead to alveolar rupture, causing mediastinal emphysema and the release of gas along the aorta and mesenteric blood vessels into the intestinal wall; and (3) the bacterial theory proposes that aerogenic bacteria penetrate the intestinal mucosal barrier, ferment in the intestinal wall, and produce gas.

PCI can occur in any part of the digestive tract,⁵⁻⁸ such as the small intestine, colon, esophagus, intestinal membrane, liver and stomach ligaments, abdominal lymph nodes, and vaginal walls, but most commonly in the colon sigmoideum. Given that the clinical symptoms of the disease are not specific, pre-operative diagnosis is difficult and may lead to undiagnosed or misdiagnosed PCI. In recent years, with the improvement of imaging and endoscopy technology, imaging and endoscopic examinations have become effective means of aiding in pre-operative diagnosis, and pathological histology examination is the direct basis for diagnosis. The imaging findings of the disease have been reported as follows⁹: (1) Abdominal flat sheets show a wave-like cystic permeable area of the intestinal edge; when the cyst ruptures, free gas under the diaphragm can be seen. Endoscopic examination is the most intuitive, showing a submucosal polyp-shaped emphysema protruding into the cavity. However, endoscopy can be used only in observing the mucous membrane cyst and not the mucosa muscle layer and the plasma membrane layer of the cyst. As such, pre-operative diagnosis should be combined with imaging and endoscopy to provide a reasonable treatment. PCI involves benign lesions and has a good prognosis. The principle of treatment is conservative-based and endoscopic. Surgical treatment is considered as a supplement to comprehensive treatment. If the condition does not lead to intestinal obstruction, perforation, bleeding and other life-threatening complications, surgical treatment is not required. Previously, PCI was often managed via conservative therapy, which changed in recent years; Wang et al.⁵ used waterless alcohol to make tissue cells dehydrated, enabling protein solidification, blood vessel contraction, endothelial cell degeneration and other characteristics. After aspirating the gas in the cyst with an injection needle and injecting 0.5-1.0 mL of anhydrous alcohol, the gas cyst collapsed. This method successfully treated PCI five cases, with no bleeding or perforation. Surgery is performed under the following conditions¹¹: (1) Patients failed to improve after conservative and endoscopic treatment; (2) a wide range of lesions were present in the intestines; (3) patients had polyps and/or tumors; or (4) infection, hemorrhage, perforation, obstruction, or torsion in the digestive tract was present. Our patient

was treated by performing an intestinal perforation emergency surgery with high efficacy and satisfactory effects. Follow-up results were good at 6 months postoperatively.

Surgical treatment should not be always the first course of treatment if the disease can be managed via conservative treatment. In our case, surgery was necessary because of an acute abdomen caused by an intestinal perforation. Surgical treatment should strictly provide a better scope for improvement rather than other options and should not be performed only to avoid conservative treatment. In short, the best treatment scheme should be selected according to age, history, symptoms, lesion scope and possible complications in the clinical treatment of PCI. Physicians should raise awareness of the disease because early detection, diagnosis, and treatment are crucial to avoid missed diagnosis and misdiagnosis.

Abbreviations

PCI Pneumatosis cystoides intestinalis

CT Computed tomography

Declarations

Ethics approval and consent to participate:

Not applicable.

Consent for publication:

Consent was obtained from the patient in question for publication of this case report and accompanying images. A copy of the written consent is available upon request.

Competing interests:

The authors declare that they have no conflicts of interest.

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Authors' contributions:

Chunyan Yuan and Zhongyue Huang contributed equally to this work.

Chuanyan Yuan wrote the manuscript and acquired information of the patient.

Zhongyue Huang performed literature review and followed-up.

Xia Sheng revised the manuscript.

Xuming Wang revised the manuscript.

All the authors read and approved the final manuscript.

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Authors' information:

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Figures

Figure 1

Imaging findings of PCI

Figure 2

Intestinal mucous membrane surface covered with soap bubble-like small cysts

Figure 3

Part of the sac wall was covered with flat or cubic epithelial cells

Figure 4

Schistosome egg deposition in the intestinal wall

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