



Pneumatosis Cystoids Intestinalis as Cause of Unnecessary Surgery: Clinical Case

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ABSTRACT

Background: The causes of PCI vary; 85% are secondary to other conditions such as abdominal pain from trauma, intestinal obstruction, inflammatory bowel disease, malignancies, chemo radiotherapy, chronic lung diseases, and connective tissue diseases. Diagnosis is challenging due to asymptomatic or atypical symptoms. Endoscopic ultrasonography (EUS) and endoscopic fine-needle aspiration are helpful for the diagnosis and treatment of PCI. **Clinical Case:** A 54-year-old female patient income for abdominal pain. Background: rheumatoid arthritis treated with prednisone for five years, surgical: cesarean section 11 years ago, Hartmann procedure for complicated diverticular disease in November 2020, intestinal transit restoration surgery in April 2021, referred allergy to aminoglycosides. We performed an exploratory laparotomy, which obtained the following findings:

"scarce intra-abdominal free air, ten mL of inflammatory reaction fluid in the cavity, diverticula among 2-3 cm in diameter. Cystic formations in the mesenteric border of approximately 5 mm in diameter at the level of the jejunum and proximal ileum with cystic pneumatosis in the mesentery of the jejunum and proximal ileum 150 cm to 170 cm from the angle of Treitz and 80 cm from the ileocecal valve. Discussion: PCI is associated with a wide variety of underlying conditions such as chemotherapy; underlying malignancy, and graft-versus-host disease (GVHD), long-term steroid treatment has also been reported to be associated with PCI. Gastrointestinal GVHD occurs infrequently in patients suffering from chronic GVHD after successful treatment of bone marrow transplantation (BMT) or stem cell transplantation which can lead to mucosal damage and increased apoptosis between mucosal cells and cells inside the intestinal wall. These alterations can result in diarrhea, abdominal pain, malnutrition, and pseudo-obstruction, which can increase extraintestinal pressure and cause PCI.

Keywords: Pneumatosis cystoids intestinalis, Pneumoperitoneum, Abdominal imaging, acute abdomen, oxygen therapy, Surgery.

INTRODUCTION

Cystic pneumatosis of the intestine (PCI) is a condition characterized by the presence of gas-filled cysts in the intestinal wall. Duo Vernet was described in autopsy specimens in 1730 [1] and was recognized radiologically in 1946 by Lerner and Gazin [2]. The causes of PCI vary; 85% are secondary to other conditions such as abdominal pain from trauma, intestinal obstruction, inflammatory bowel disease, malignancies, chemo radiotherapy, chronic lung diseases, and connective tissue diseases. Diagnosis is challenging due to asymptomatic or atypical symptoms. Endoscopic ultrasonography (EUS) and endoscopic fine-needle aspiration have to be helpful for the diagnosis and treatment of PCI.

This entity is classified into primary (15%) and secondary (85%) types. The secondary type occurs with diseases such as strictures of the digestive tract, obstructive pulmonary disease, external abdominal injury or surgery, and malnutrition. There are three hypotheses for its etiopathogenesis: 1) mechanical theory: involving an increase in intraluminal pressure that causes damage and rupture of the mucosa of the intestinal wall, which leads to the migration of gas from the digestive tract towards the intestinal wall; 2) pulmonary theory: chronic lung diseases such as chronic obstructive pulmonary disease, asthma, and interstitial pneumonia lead to alveolar rupture, causing mediastinal emphysema and gas release along the aorta and mesenteric blood vessels in the intestinal wall; and 3) bacterial theory: aerogenic bacteria penetrate the intestinal mucosal barrier, ferment in the intestinal wall and produce gas (3).

PCI is a disease that can occur at all ages, commonly presenting at 45 ± 15 years of age. Non-surgical treatment results in remission in 70% of patients. However, PCI can be confused with other serious diseases due to the "serious" imaging findings; they can lead to surgical procedures, as in our case (4).

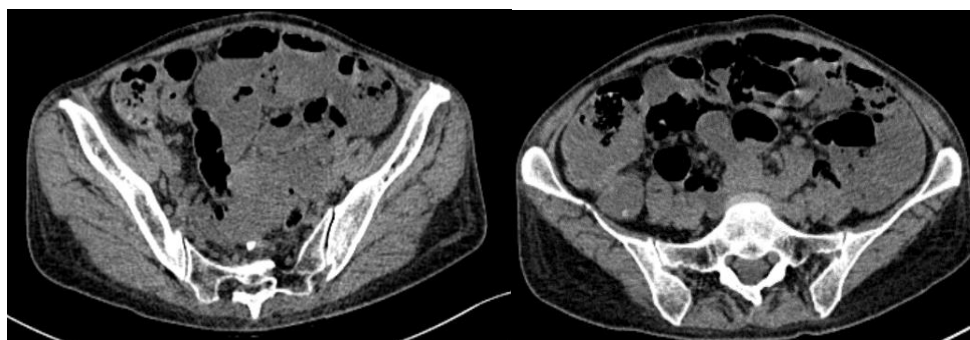
CLINICAL CASE

A 54-year-old female patient income for abdominal pain. Background: rheumatoid arthritis treated with prednisone for five years, surgical: cesarean section 11 years ago, Hartmann procedure for complicated diverticular disease in November 2020, intestinal transit restoration surgery in April 2021, referred allergy to aminoglycosides.

His condition began a month prior to admission with the presence of abdominal pain located in the epigastrium, left iliac fossa, and hypogastrium, with intensity 6/10 of colic type; he referred diarrheal bowel movements interspersed with periods of constipation, which is why he went to a gastroenterologist who indicated management. Doctor with partial improvement; due to persistent pain predominantly in the right iliac fossa and right flank triggered after food intake, radiating to the lumbar region, without improvement after ingesting paracetamol, associated with abdominal distension.

On physical examination, a globose abdomen at the expense of the adipose panniculus and abdominal distension, with the presence of multiple hypertrophic scars secondary to previous surgical procedures, soft, depressible, decreased peristalsis in intensity and frequency, painful on deep palpation in the right flank and right iliac fossa, negative rebound, with voluntary muscular resistance, without other alterations.

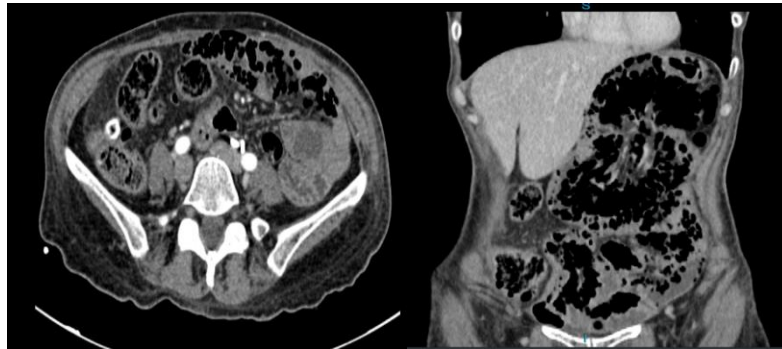
It was performed on admission with standard laboratories, and a simple abdominal tomography revealed dilation of small bowel loops, air-fluid levels, intestinal pneumatosis, and diverticulosis.



Figures 1, 2. Simple tomography upon admission with intestinal pneumatosis.

Due to stable vital signs, moderate pain, and the absence of clinical and laboratory data justifying emergency surgical treatment, admission to the general surgery floor was indicated for surveillance.

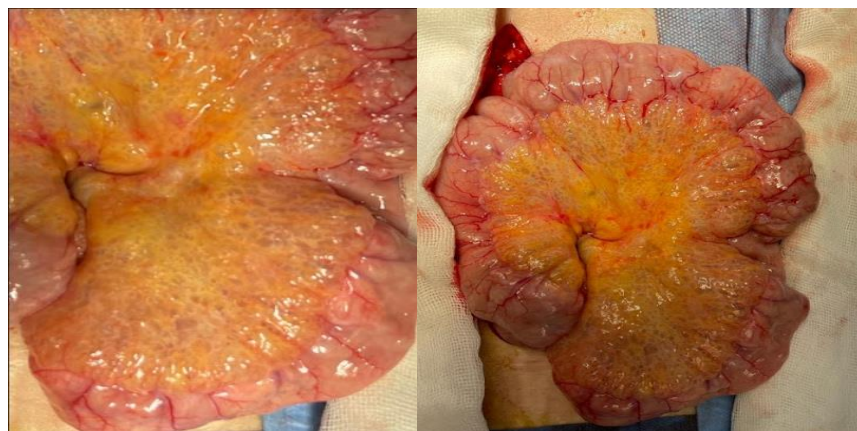
At 48 h, a control abdominal tomography was performed, which reported: "small bowel loops up to 35 mm in diameter with air-fluid levels, intestinal pneumatosis, with a significant increase in density and striation of the adjacent fat; as well as interasa fluid, pelvic cavity, and right iliac fossa; identified free fluid and air in the abdominal cavity.



Figures 3, 4. CT scan at 48 h of hospital stay. Watch intestinal pneumatosis and dilation of small bowel loops.

We decided to perform an exploratory laparotomy, in which we observed the following findings: "scarce intra-abdominal free air, ten cc of inflammatory reaction fluid in the cavity, diverticula among 2-3 cm in diameter. Cystic formations in the mesenteric border of approximately 5 mm in diameter at the level of the jejunum and proximal ileum with cystic pneumatosis in the mesentery of the jejunum and proximal ileum 150 to 170 cm from the angle of Treitz and 80 cm from the ileocecal valve."

The peritoneal cavity was without finding other abnormalities, so the patient was re-admitted to the general surgery floor, where she progressed favorably with metronidazole until she was discharged from the hospital.



Figures 5, 6. Cystic formations with air content in the mesenteric border of the jejunum and ileum.

DISCUSSION

PCI has a wide variety of underlying conditions such as chemotherapy; underlying malignancy, and graft-versus-host disease (GVHD), long-term steroid treatment has also been reported to be associated with PCI. Gastrointestinal GVHD occurs infrequently in patients suffering from chronic GVHD after successful treatment of bone marrow transplantation (BMT) or stem cell transplantation which can lead to mucosal damage and increased apoptosis between mucosal cells and cells inside the intestinal wall. These alterations can result in diarrhea, abdominal pain, malnutrition, and pseudo-obstruction, which can increase extraintestinal pressure and cause PCI (5).

Its clinical importance may vary depending on the underlying pathology that causes it. Traditionally, pneumatosis intestinalis is considered a sign of intestinal necrosis requiring urgent surgical management. When ischemia occurs, the patient can deteriorate rapidly, leading to up to 85% mortality rates. However, many cases not related to ischemia follow a more favorable course. In these scenarios, timely clinical surveillance is necessary. Some causes associated with this condition include traumatic events (endoscopy, ulcer, and anastomosis); inflammatory diseases or autoimmune diseases (Crohn's disease, diverticular disease, and lupus); infectious diseases (HIV, mycobacterium, and adenovirus), lung disorders (asthma, COPD and cystic fibrosis); drug-related (corticosteroids, lactulose, and alpha-glycosidase inhibitors) and other rare etiologies (sclerosis and hemodialysis) (6)

PCI is a challenging and rare condition and should be suspected in patients with pneumatosis of the intestines and pneumoperitoneum associated with mild symptoms and insignificant laboratory evaluation. CT can also make a diagnosis. Recognition of PCI is essential to avoid unnecessary surgical procedures (7).

When presenting in the setting of trauma, PCI can make management difficult. To our knowledge, there are no evidence-based guidelines for setting PCI in blunt abdominal trauma with pneumoperitoneum. Asymptomatic patients, without signs of clinically worrying disease and with adequate examination, probably will not benefit from surgery and can be managed medically (8).

The pneumoperitoneum refers to the presence of free gas in the peritoneal cavity. A visceral perforation accounts for 85 to 95% of all cases and generally indicates a surgical emergency. However, in approximately 10% of cases, a variety of benign conditions could also lead to a spontaneous pneumoperitoneum that does not require surgical intervention. In these cases, the pneumoperitoneum is asymptomatic and incidentally detected by radiological investigations. Sometimes, computed tomography can also reveal a false image of intraperitoneal free air leading to a misleading diagnosis of pneumoperitoneum (9).

PCI can be associated with multiple disease entities, such as intestinal pseudo-obstruction, and the pathological mechanisms have not been elucidated. Systemic sclerosis, characterized by progressive sclerotic changes of the skin and internal organs, is a refractory collagen disease frequently associated with digestive disorders (10); connective tissue disease is also a significant cause of secondary PCI. In addition, the use of prednisolone and some immunosuppressants, as well as the presence of diseases such as diabetes mellitus, constipation, and lung diseases, are involved in the development of PCI (11); PCI in adult dermatomyositis (DM) is rarely described. Liu et al. (12) observed six patients with DM (four with anti-MDA5 antibodies, one with anti-SAE antibodies, and one with anti-TIF-1g antibodies). The remaining five patients were asymptomatic except for one with transient abdominal pain. PCI is a known but rare complication of scleroderma; Shing et al. (13) present a case with eight months of weight loss, urgency, diarrhea, bloating, and cramping abdominal pain. The patient was diagnosed with systemic scleroderma's limited cutaneous variant (CREST). Yang et al. (14) reports a patient with abdominal distension and constipation after chemotherapy for hematological malignancy.

Yuan et al. (15) present a case of PCI in a 90-year-old man. After emergency surgery, the patient was hospitalized due to an acute abdomen and diagnosed with perforation of the sigmoid colon by PCI with Schistosomiasis. PCI can be associated with volvulus (16), intussusception (17), and pneumoperitoneum (18).

Regarding the management of this entity, we have hyperbaric oxygen and conservative treatment in mild cases until surgery. Hyperbaric oxygen, in combination with a personalized diet and antibiotics, may be beneficial for moderate to severe PCI in patients without an indication for emergency exploratory laparotomy (19). Hamada et al. (20) treated a case with low-dose oxygen therapy to prevent acute exacerbation of interstitial pneumonia; the oxygen dose was adjusted so that the PaO₂ value was approximately 100 mmHg. After seven days of treatment, the colonic gas had disappeared, and no acute exacerbation of interstitial pneumonia. At a PaO₂ value of about 100 mmHg, it is effective for PCI without inducing an acute exacerbation of interstitial pneumonia.

The treatment modalities range from conservative management, bowel rest, antibiotic therapy, and enteral nutrition to emergency surgery, bowel resection, and stoma formation.

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