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Case Report

Idiopathic pneumatosis cystoides coli: An uncommon cause of pneumoperitoneum ☆

André Peixoto, MD, Rafaela Pereira, MD, João Leitão, MD*

Serviço de Imagiologia Geral, Centro Hospitalar Universitário Lisboa Norte, Avenida Professor Egas Moniz, Lisboa 1649-035, Portugal

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ABSTRACT

Pneumatosis cystoides intestinalis (PCI) is a rare entity characterized by the presence of gaseous cystic within the intestinal wall. The primary or idiopathic type represents 15% of cases and is a self-limited or chronic benign entity. The secondary type represents 85% of cases and is associated with various factors, such as surgery, pharmacotherapy, chemotherapy, autoimmune diseases, inflammatory diseases, and pulmonary illness. Pneumatosis cystoides intestinalis affects the colon (pneumatosis cystoides coli) in about half of the cases. The differential diagnosis of PCI includes potentially life-threatening diseases that cause pneumatosis intestinalis. The misdiagnosis of PCI is common and can lead to unnecessary treatments and surgical procedures. We describe an asymptomatic pneumoperitoneum incidentally seen on chest radiograph. The cause was pneumatosis cystoides coli, which did not require treatment.

Introduction

Pneumatosis cystoides intestinalis (PCI) is a rare entity, characterized by the presence of gaseous cystic within the intestinal wall [1,2].

There are 2 types of PCI. The primary or idiopathic type affects 15% of patients. The secondary type affects 85% of patients and is associated with various factors, such as surgery, pharmacotherapy, chemotherapy, autoimmune diseases, inflammatory diseases, and pulmonary illness [1,3].

Although the etiology and pathogenesis of PCI remain unclear, some hypotheses in the literature attribute the cause of this entity to predisposing factors, namely inflammation, physical damage to the intestinal mucosa, nutritional imbalance, dysbacteriosis, and immune dysfunction [1].

Despite PCI occurring from the esophagus to the rectum, it is more common in the colon, affecting about half of the patients. Involvement of the colon is called pneumatosis cystoides coli. PCI usually has a benign course and is a chronic or self-limited process [1–4].

☆ Competing Interests: The author declares that he has no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

* Corresponding author.
E-mail address: av_b14@hotmail.com (A. Peixoto).

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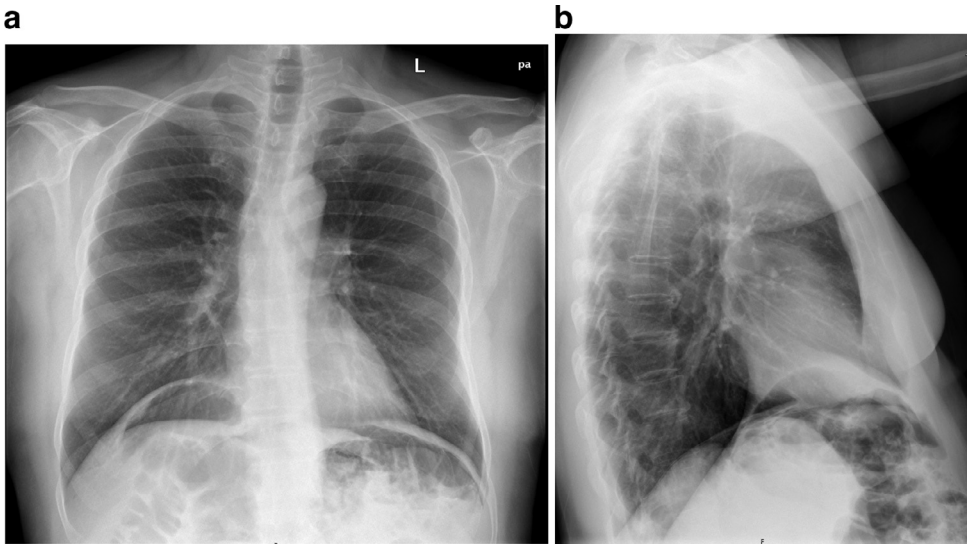

Fig. 1 – Chest radiograph, posteroanterior (A) and lateral (B) views show bilateral subdiaphragmatic free air (pneumoperitoneum). In the upper abdominal quadrant, along the intestine's wall, gas pockets radiolucency is noted. There were no signs of pulmonary infection.

The differential diagnosis of PCI includes potentially life-threatening diseases that cause pneumatosis intestinalis. PCI is a different entity from pneumatosis intestinalis. Pneumatosis intestinalis is characterized by the linear or bubbly appearance of intramural gas and is associated with severe diseases such as bowel ischemia [5–7].

We describe an asymptomatic pneumoperitoneum incidentally seen on chest radiograph. The cause was pneumatosis cystoides coli, which did not require treatment.

Case report

A 54-year-old male went to an appointment consultation with his general practitioner. Due to cough, a chest radiograph was ordered.

On chest radiograph, a pneumoperitoneum is noted (Fig. 1). There were no signs of pulmonary infection.

A review of 8-year-old abdominal radiograph demonstrated multiple gas pockets radiolucency, in the form of a grape-like cluster, along the intestine's wall, predominantly in the right upper abdominal quadrant (Fig. 2).

As the patient was asymptomatic, a computerized tomography (CT) scan was ordered and performed a few weeks later. The CT scan confirmed the presence of intestinal air cysts within the colon's wall, with a diffuse distribution, as well as the pneumoperitoneum (Fig. 3).

A follow-up CT scan 1 year later continued to demonstrate pneumoperitoneum and cystic gas along the colon's wall (Fig. 4).

The patient remained asymptomatic during follow-up and did not require any treatment.

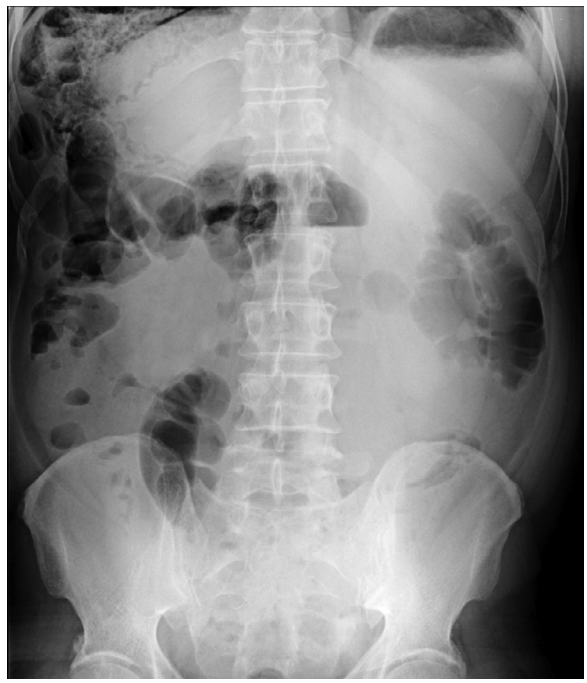

Fig. 2 – Old abdominal radiograph demonstrates multiple gas pockets radiolucency, in grape-like cluster or honeycomb-shaped appearance, along the intestine's wall, predominantly in the right upper abdominal quadrant.

Discussion

PCI is characterized by the presence of multiple gas-filled cysts in the wall (submucosa and subserosa) of the gastrointestinal tract, ranging from 0.5 to 2.0 cm in diameter [1,4,8].

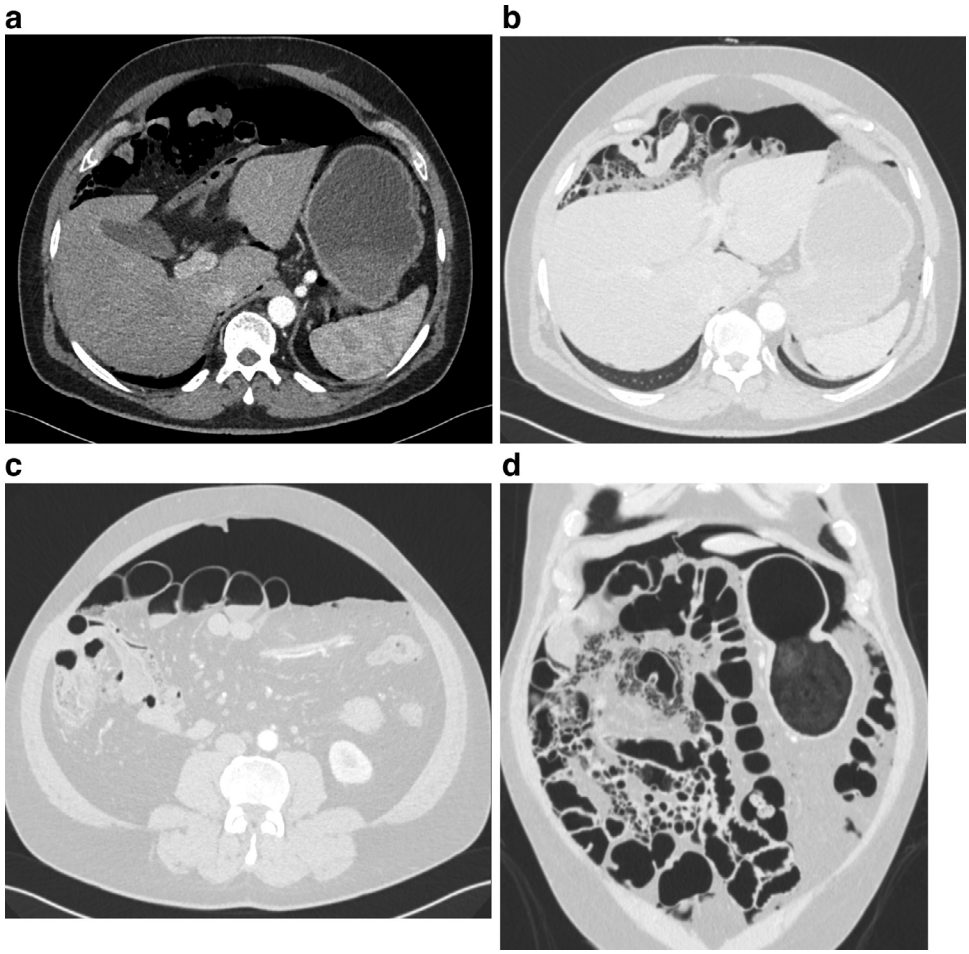

Fig. 3 – Enhanced computerized tomography, axial (A-C) and coronal (D) images show pneumoperitoneum and multiple intestinal air cysts within the colon's wall, with a diffuse distribution, more numerous on the right colon.

PCI usually affects adults in the fourth to sixth decades of life. In old cohort studies, there is a male predominance, while recent authors suggest that PCI affects males and females equally [3].

PCI is common in the colon (pneumatosis cystoides coli) and the small intestine [3,9].

Although the etiology of PCI is unknown, there are some hypotheses in the literature. The mechanical theory suggests that PCI is caused by increased endoluminal pressure due to intestinal obstruction, inflammatory bowel disease, ischemic bowel disease, bowel preparation or colonoscopy, and consequent gas migration from the lumen to the gastrointestinal wall. The pulmonary theory proposes that chronic lung pathology causes alveolar rupture and migration of the gas to mesenteric vessels and intestinal walls. The bacterial theory states that bacteria within intestinal mucosa produce gas and form cysts. The nutrition theory states that malnutrition prevents the digestion of carbohydrates, causing an increase in bacterial gas production and consequent submucosal dissection. However, none of the theories can entirely explain the pathologic process [3,4,8].

This is usually a chronic or self-limited benign disorder. The primary or idiopathic type affects 15% of patients. The secondary type affects 85% of patients and can be associated with several conditions, namely post-surgical, drugs (steroids, immunosuppressive drugs), trichloroethylene exposure, autoimmunity diseases (scleroderma), mechanic ventilation, gastrointestinal pathology (inflammatory bowel diseases, intestinal occlusion) and pulmonary pathology (chronic obstructive pulmonary disease, asthma) [6,10–12].

The vast majority of PCI are asymptomatic at diagnosis and incidentally detected. However, some patients complain of non-specific gastrointestinal symptoms, such as abdominal pain, abdominal distension, nausea, and diarrhea [6,8,10–13].

When misdiagnosed, PCI can lead to unnecessary examinations, treatments, and even surgical procedures [4,9].

CT scan is the most sensitive imaging modality for diagnosis. Endoscopy and endoscopic ultrasonography can help in the diagnosis [1,14,15].

On abdominal radiograph, there are radiolucent gas pockets, in grape-like clusters or honeycomb-shaped appearance, along the wall of the intestine [3,6,16].

CT scan shows intestinal air cysts within the submucosa or the subserosa of the colon. PCI may involve only a segment or be diffuse. Colonoscopy may show grape-like or beaded subepithelial gas cysts [2,6,15,16].

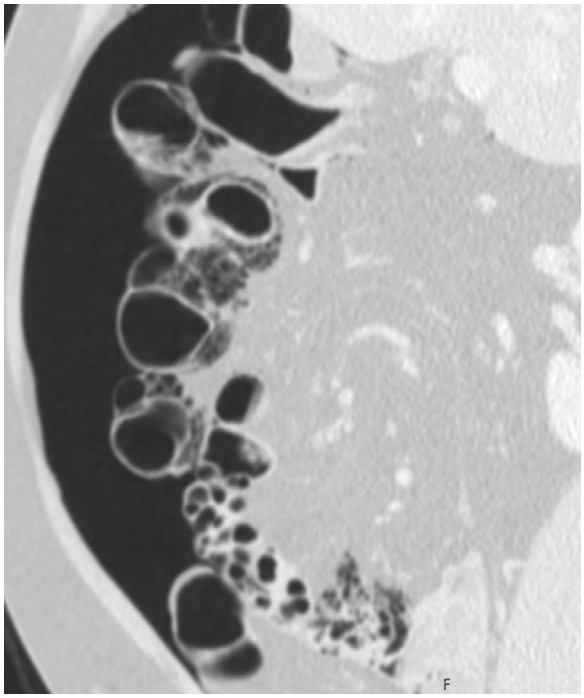

Fig. 4 – Follow-up enhanced computerized tomography, sagittal image continued to demonstrate pneumoperitoneum and colon with gas-filled cysts on the subserosa and submucosa.

The differential diagnosis includes potentially life-threatening conditions, such as bowel ischemia, infarction, necrotizing enterocolitis, neutropenic colitis, volvulus, and sepsis. The presence of ominous signs, symptoms of peritonitis, and laboratory abnormalities, like an elevated white blood cell count and acidosis, should raise a concern about these diseases. Furthermore, the detection of portal venous gas, bowel distension, ascites and decreased bowel wall enhancement is significantly associated with life-threatening causes [4,13,17,18].

Complications of PCI occur in 3 to 16% of patients. Intestinal perforation and intestinal obstruction are the most frequent. Large cysts can cause intrinsic or extrinsic compression, resulting in obstruction. Ruptured cysts can cause pneumoperitoneum, without signs of peritoneal irritation [1,3,9].

There is controversial data in the literature. A few authors report high rates of mortality. Nevertheless, the latter appears to be explained by the underlying pathology or misdiagnosis rather than the PCI itself. Some authors use the term "pneumatosis cystoides intestinalis" to describe the gas accumulation seen in pneumatosis intestinalis associated with severe diseases, such as bowel ischemia and necrosis. However, these cases do not have the typical cystic appearance of the benign disease type that we show in this case report [4–7,11–13,16–19].

PCI typically does not require treatment. If symptoms are pronounced, the treatment may include antibiotics, oxygen, bowel rest, gastrointestinal decompression, probiotics, and supportive care. If peritoneal irritation or bowel obstruction is suspected surgery must be considered [1–9,19–21].

Conclusion

PCI is a rare disease characterized by the presence of multiple gas-filled cysts in the intestinal wall. PCI is a chronic or self-limiting and benign entity. Colon (pneumatosis cystoides coli) is the most common location. It is usually asymptomatic, but can cause abdominal pain, abdominal distension, nausea and diarrhea. Cysts' rupture can lead to asymptomatic pneumoperitoneum. Therefore, when an unexpected pneumoperitoneum is seen, PCI should be considered in the differential diagnosis. Frequently, there is no need for treatment. In the presence of clinical and imaging worrisome signs, an alternative and potentially life-threatening disease must be considered.

Patient consent

The patient's informed consent for the publication of this case was granted.

Ethics statement

There are no ethical issues for the publication of this case report according to the standard of our institution.

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