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Case Report

A rare case of extensive pneumatosis cystoides intestinalis with intestinal malrotation: Case report ☆

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ABSTRACT

Intramural bowel gas (Pneumatosis intestinalis) refers to the radiological or clinical evidence of gas within the wall of the bowel lumen. While intramural gas could be secondary to life-threatening pathologies such as mesenteric ischemia in adults and necrotizing enterocolitis in neonates, it could also occur as a rare benign sub-type called Pneumatosis cystoides intestinalis, which is characterized by multiple gas-filled cysts in the submucosa and/or subserosal of the gastrointestinal tract. Distinguishing between life-threatening Pneumatosis intestinalis and its benign subtypes requires careful clinical and imaging evaluation. This involves identifying additional findings that could indicate potentially concerning causes of Pneumatosis intestinalis. Recognizing these signs is essential for effectively managing the patient because conservative management is preferred for Pneumatosis cystoides intestinalis. In this case study, we describe a patient presenting to our hospital with chronic intermittent abdominal pain persisting for about 2 years, accompanied by episodic vomiting. An abdominal CT scan revealed the presence of multiple air-filled cysts within the wall of the mal-rotated cecal bowel loop, which is abnormally located in the right upper quadrant. Associated with this pneumoperitoneum is seen in the peritoneal cavity. No other significant findings were observed on the scan. To our knowledge, this is the first case of pneumatosis cystoid interstinalis occurring in a mal-rotated gut. We also delve into the potential etiologies and management strategies for Pneumatosis cystoides intestinalis, as well as differentiating signs from the life-threatening intramural gas variant.

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Introduction

Pneumatosis cystoides intestinalis (PCI) represents a benign subtype within the spectrum of Pneumatosis intestinalis. While it can manifest anywhere along the gastrointestinal (GI) tract, studies indicate a predisposition to the colon, accounting for approximately 46% of cases, followed by the small intestine, which comprises approximately 27% of cases [1,2]. Intestinal malrotation arises as a congenital anomaly, originating from an aberrant rotation of the gut during its return to the abdominal cavity in embryogenesis.

Case presentation

A 24-year-old male patient presented to our hospital with a 2-year history of diffuse, intermittent abdominal pain accompanied by episodic vomiting. Despite seeking medical attention at multiple clinics and receiving treatment for dyspepsia, the symptoms persisted without significant improvement. Further investigation included upper gastrointestinal (GI) endoscopy, which was indicated due to unresponsiveness to proton pump inhibitors treatment for dyspepsia, but the results were normal. The patient denied experiencing diarrhea, fever, or any significant weight changes. On physical examination, the vital signs were in normal range and no abnormality was detected. Laboratory investigations, including full blood count, liver function tests, and renal function tests, were within normal limits. Stool microscopy findings were also unremarkable. Following initial assessments, the patient underwent both an abdominal ultrasound and a CT scan for further evaluation. The abdominal ultrasound was unremarkable, limited by extensive gaseous bowel. However, the CT scan demonstrated extensive variable-sized air-filled cysts (Fig. 1) primarily arising from the walls of the cecum located in the right upper quadrant (Fig. 2). The right lower quadrant is occupied with small bowel loops (Fig. 3). Additionally, large cysts were observed to extend between the liver and the right hemidiaphragm. Free intra-abdominal air collection was also detected, notably between the left lobe of the liver and the anterior abdominal wall, as well as in the gastro-hepatic ligament (Fig. 2b).

Discussion

Pneumatosis intestinalis (PI) is a condition characterized by the presence of gas in the bowel wall. It can be classified into 2 main forms: primary and secondary. The most common and often life-threatening type is the secondary, which is caused by an obstructed and/or necrotic bowel. It accounts for 85% of the cases. This form is often accompanied by symptoms and can be a sign of a more severe underlying condition. Clinicians should often rule out this form before diagnosing the primary/benign type of Pneumatosis intestinalis [3]. Primary Pneumatosis intestinalis, also recognized as Pneumatosis cystoides intestinalis (PCI), was initially documented by Du Vernoi in 1730 during a cadaver dissection [4]. It accounts for 15% of the cases with an estimated incidence of ~0.03%. Although predominantly observed in adults in their 5th to 8th decades of life, it can occur in individuals of all age groups [1,5]. This form is usually asymptomatic and may be incidentally discovered through radiography or endoscopy. Among symptomatic patients around two-third have diffuse or localized chronic abdominal pain other associated symptoms include diarrhea, vomiting and abdominal distension [6].

The pathogenesis of Pneumatosis cystoides intestinalis has long been a subject of debate, with various theories suggested to explain its underlying mechanisms. Among these, 3 main theories have emerged as central to understanding the condition: mechanical, bacterial, and pulmonary causes. The mechanical theory proposes that an elevation in intraluminal pressure triggers mechanical damage and mucosal rupture within the intestinal wall, facilitating the migration of gas from the gastrointestinal cavity into the intestinal wall [2,4]. Bacterial theory suggests that air-producing bacteria breach the intestinal mucosal barrier, instigating gas production that becomes trapped within the submucosa and lymphatic channels. This idea is backed by experiments showing that introducing bacteria into the gut wall causes pneumatosis, with the cysts containing hydrogen. There are also reports of the gas disappearing after treatment with antibiotics [7–10]. Pulmonary theory suggests that chronic lung conditions like chronic obstructive pulmonary disease (COPD), asthma, and interstitial pneumonia can cause alveolar (lung air sac) rupture. This rupture can lead to the escape of air into the mediastinum, and from there, the air can travel along the aorta and mesenteric blood vessels, eventually reaching the intestinal wall [11]. PCI may also arise from secondary causes, including inflammatory bowel disease (such as Crohn's disease and ulcerative colitis), sigmoid volvulus, and progressive systemic sclerosis [5,12,13].

When evaluating Pneumatosis intestinalis, it is crucial to ascertain whether the cause is life-threatening or non-life-threatening. Clinically, elevated lactate levels, tachycardia, and abdominal tenderness are suggestive of pathological Pneumatosis intestinalis [14]. CT scans are the most sensitive diagnostic tool. In Pneumatosis cystoides intestinalis, the cysts may rupture, leading to the production of pneumoperitoneum, which is often benign. Therefore, pneumoperitoneum alone cannot be used as a differentiating clue [15]. Secondary CT signs are indeed important in differentiating between benign and worrisome intraluminal gas. A retrospective study involving CT scans of 84 patients treated at a tertiary cancer center revealed that imaging features can aid in this differentiation. Findings such as bowel wall thickening, mesenteric stranding, ascites, bowel dilatation, and portomesenteric venous gas show a significant correlation with life-threatening Pneumatosis intestinalis. Conversely, if the gas is confined to the colon alone, it suggests a benign origin [16]. We suggest that assessing for these secondary signs is crucial when evaluating patients with any intraluminal bowel gas.

Conservative management is the preferred management which includes observation and antibiotic therapy. Surgical management is reserved for complications or severe symptoms. PCI may be self-limited, and conservative treatments are effective in 90% of cases, consisting of oxygen or antibiotic therapy [17–19]. The proposed mechanism for hyperbaric

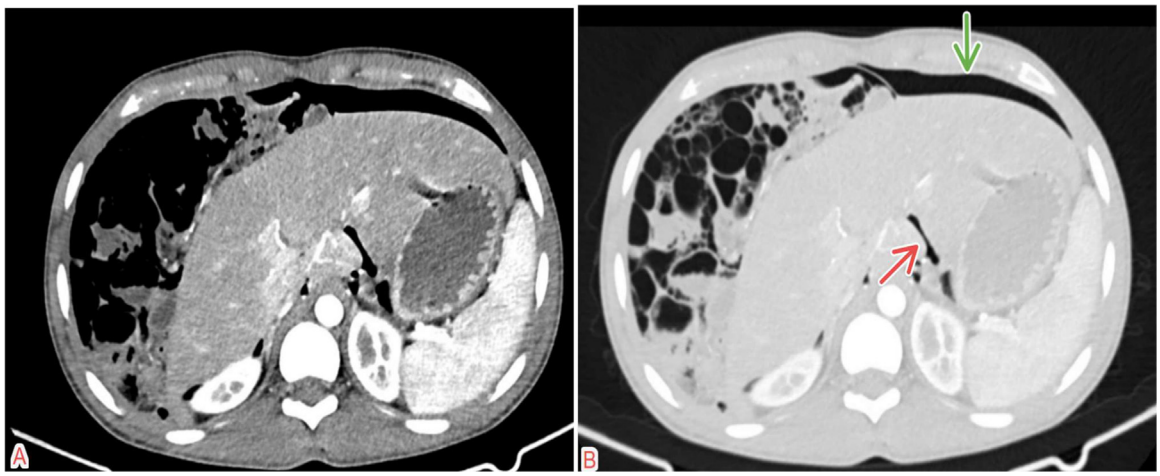

Fig. 1 – Axial post-contrast abdominal CT at the level of the liver (A) abdominal window (B) lung window. Shows multiple bubbly variable-sized air-filled cysts between the liver and right abdominal wall. The liver is displaced medially. On the lung window (B), free air was identified between the left lobe of the liver and the anterior abdominal wall (green arrow). Additionally, free air was also observed within the gastro-hepatic ligament (red arrow).

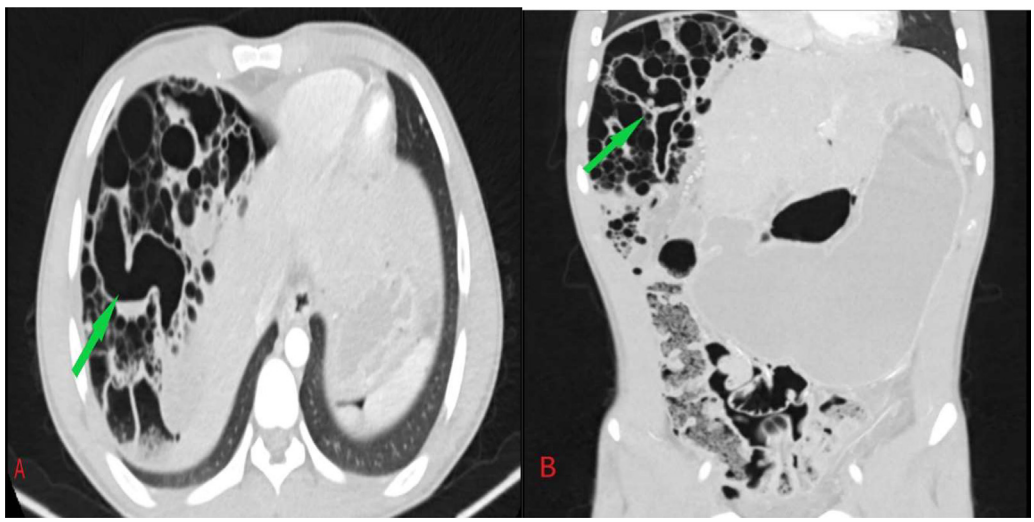

Fig. 2 – Axial (A) and coronal (B) abdominal CT in lung windows, shows the abnormally located cecum (in the right upper quadrant, green arrow with multiple variable size air-filled cysts seen arising from its wall.

oxygen therapy is it creates a high PaO2 gradient promoting gas outflow through the cyst wall, and it also produces a high amount of oxygen concentration lethal to gas-producing bacteria. However, its efficacy is not fully concluded with some authors suggesting up to 50% [20–22]. Antibiotic therapy primarily targets reducing anaerobic bowel flora to mitigate gas production, with metronidazole being a commonly used agent for this purpose. The duration of therapy typically continues until clinical and radiological resolution is achieved [23]. Surgical management of Pneumatosis cystoides intestinalis is typically reserved for cases where conservative treatment fails or when severe complications such as bowel necrosis, perforation, or persistent obstruction necessitate intervention based on the patient's clinical presentation [24–27]. In our case, Pneumatosis cystoides intestinalis occurred concomitantly in the malrotated bowel. Malrotation occurs when the process of intestinal rotation and fixation deviates from its typical course. This anomaly disrupts the normal positioning of the cecum and appendix. In malrotation, these structures will be located in the right upper quadrant, in a subhepatic location [28–30].

Due to recurrent vomiting in this case, surgical intervention was deemed necessary. The Ladd band was released, and cecoplexy was performed to address the mal-positioned cecum. During the procedure, extensive Pneumatosis cystoides intestinalis was discovered in the cecum, ascending colon, and surrounding mesentery.

We have not found any previous cases reported with these 2 occurring together, this might be because both conditions are rare and could be asymptomatic and it might be difficult to know to true incidence.

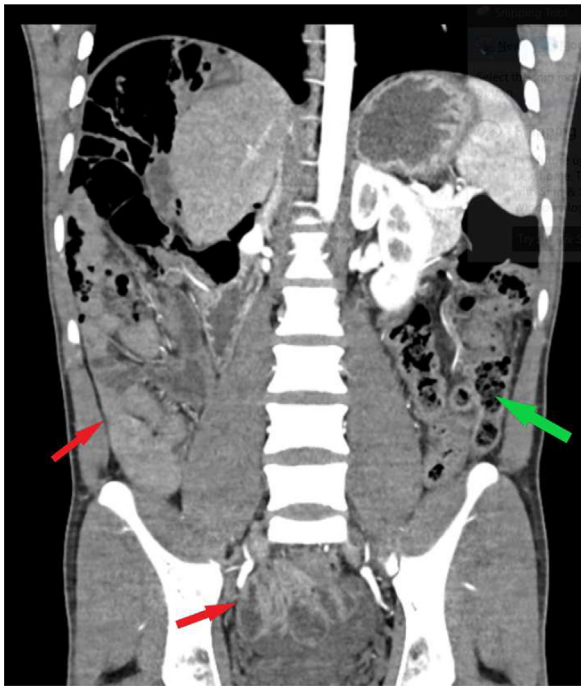

Fig. 3 – Post-contrast coronal CT of the abdomen shows small bowel loops aggregated in the right lower quadrant and pelvic cavity (red arrows). The large bowel is also seen within the left hemi-abdomen (green arrow).

Conclusion

Pneumatosis cystoides intestinalis is a benign form of intraluminal bowel gas. However, it is extremely important to differentiate it from life-threatening forms. Computed tomography (CT) scans play a pivotal role, with secondary signs such as bowel wall thickening, mesenteric stranding, ascites, and portomesenteric venous gas indicating a more worrisome pathology. Conservative measures, including observation, oxygen therapy, and antibiotics, are often preferred. However, surgical intervention may be warranted for severe symptoms or complications.

Patient consent

Complete written informed consent was obtained from the patient for the publication of this study and accompanying images.

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