A Case Report on Pneumatosis Intestinalis complicated by pneumoperitoneum

Abstract:

Background
Pneumatosis intestinalis (PI) is a rare radiological finding characterized by gas within the bowel wall, often linked to conditions such as ischemia, infection, or inflammation. When complicated by pneumoperitoneum, it suggests a potentially life-threatening condition, such as bowel perforation, and may necessitate surgical intervention.
Case Summary
A 43-year-old male patient presented with 5 days of diffuse abdominal pain, distention, and vomiting of gastric contents. He had a remote history of peptic ulcer disease. On physical examination, he appeared sick, dehydrated, and cachexic. His vital signs were stable, but his BMI was low (16.2 kg/m2). His abdomen was distended with diffused tenderness and had no guarding or bowel sounds. There was a radiological evidence of pneumoperitoneum, pneumatosis intestinalis, and free fluid concerning hollow viscus perforation. However, small bowel biopsy showed polypoid, grape-like lesions with cystic spaces lined by multinucleated giant cells and surrounded by inflammatory cells and hemosiderin pigment. Findings were consistent with Pneumatosis Cystoides Intestinalis, with no atypia or malignancy. Following fluid resuscitation, the patient underwent laparotomy exploration which did not demonstrate any bowel injury but scarred the 1st part of the duodenum. The pneumoperitoneum was attributed to the rupture of gas-filled cysts located along the bowel wall but not within it. He had an uncomplicated postoperative course. The endoscopy was done on follow-up, revealing a small Forest III pre-pyloric ulcer with a stenotic pyloric ring managed by balloon dilatation and PPI.
Conclusion
Pneumatosis intestinalis is a rare finding and its significance cannot be determined outside of a clinical contest. Surgery may be performed occasionally when pneumoperitoneum is present, raising concerns about the risk of hollow viscus injury.

Key Words: Pneumatosis intestinalis, pneumoperitoneum, exploratory laparotomy, bowel perforation

Introduction:

Pneumatosis intestinalis (PI) is a rare condition where you can find gas-filled cysts in the intestines(1). It can be linked to many different conditions, from harmless and self-resolving ones to serious situations that need immediate surgery(2). PI is just a sign, not a disease, so we need to look at it in the bigger picture of the whole clinical context(1). Even though it seems like there are many factors contributing to PI, the specific cause is still not clear(3). There are a lot of different theories in the medical literature regarding where gas in the intestinal wall comes from, including bacterial, mechanical, and pulmonary sources(4). To understand the development, we need to look at two key points: the origin of the gas and the process of how it arrived there. Three possibilities have been suggested as the source of gas: (1) intraluminal GI gas, (2) bacterial production of gas, and (3) pulmonary gas (2)(4). PI is classified into two categories based on the factors believed to contribute to its development: idiopathic, which accounts for 15%, and secondary, which makes up 85%(5). The secondary classification is linked to various diseases, including digestive tract stenosis, obstructive pulmonary disease, abdominal external injury or surgery, and malnutrition(5).

Case Presentation:

This case discusses a patient who needed surgery because of the presence of pneumoperitoneum. This case shows an example of PI that was identified through radiology and required surgery. A 43-year-old man came to the emergency department with a 5-day history of upper abdominal pain, abdominal distention, and vomiting. The patient reported that there were no other changes in his bowel movements, and his medical history included recurrent upper abdominal pain and vomiting, for which he received treatment with PPIs. During the presentation, he appeared unwell, dehydrated, and cachectic, but his vital signs were stable. The physical exam showed that there was some swelling in the abdomen, tenderness in the upper stomach area, and slight guarding, but there were no signs of rebound tenderness, masses, or enlarged organs. Bowel sounds were hypoactive. His labs were significant for Hematocrit: 39, WBC: 6.800 /µL, CRP: 18, Serum

Potassium (K): 3.7, Sodium (Na): 133, Chloride (Cl): 87, BUN: 24, Serum Creatinine: 0.9, Bicarbonate:37, Amylase: 123, ALT: 12, AST:17, Albumin: 3.7.

A plain abdominal radiograph demonstrated extensive pneumoperitoneum (Figure 1).

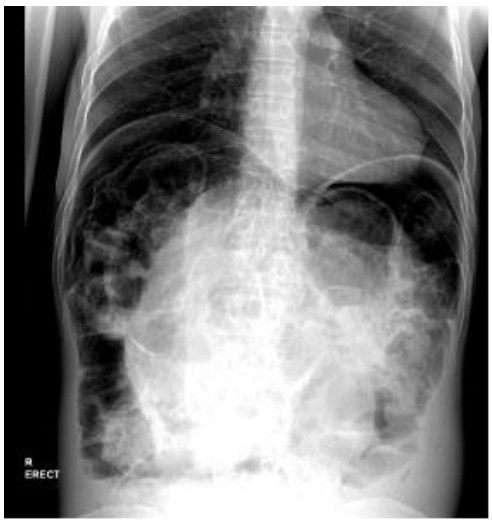

Figure 1: Abdominal radiograph demonstrating a significant amount of free intraperitoneal free air with dilated bowel loops

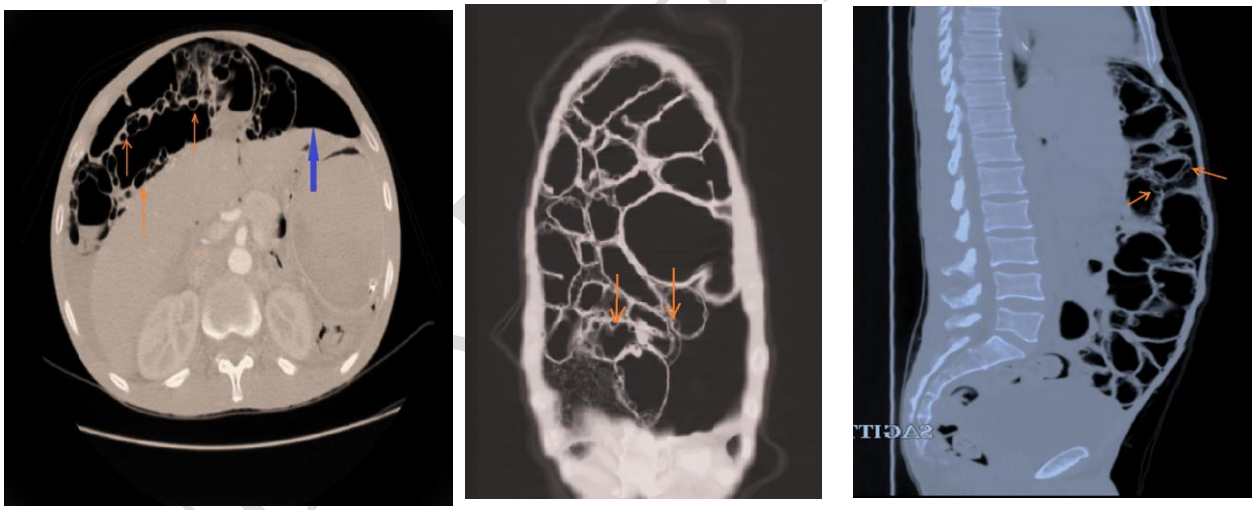

Figure 2: CT scan was obtained, which showed a significant amount of free intraperitoneal air and fluid. A focal area of bowel wall thickening was noted at the terminal duodenum and proximal jejunum. And pneumatosis of small bowel. It is showing circumferential cyst-like structures located through the small bowel (orange arrows), and intra-abdominal free air (blue arrows).

The excisional biopsy from the small bowel revealed a polypoid, grape-like lesion characterized by multiple thin, transparent cystic spaces. Microscopic examination showed fibrofatty tissue predominantly replaced by large thin-walled cystic spaces lined with multinucleated giant cells, surrounded by mixed inflammatory cells, focal hemosiderin pigments, and hemorrhagic blood in some cysts. These findings were consistent with Pneumatosis Cystoides Intestinalis. No atypia or malignancy was noted

Following resuscitation, the patient was immediately taken to the operating room over concerns of internal organ injury. The patient underwent a non-therapeutic surgical procedure that initially commenced laparoscopically. However, extensive distention of the small bowel hindered proper visualization, leading to a

conversion of an open-through upper midline incision. Intraoperatively, we noted a large amount of free fluid, with dilated small bowel and gas-filled cysts involving most of the bowel of different sizes. The cysts were located within the wall and subserosa, and adherent to the serosa & some were floating and adherent to the parietal peritoneum in the upper abdomen (Figure 3). Exploration revealed a thickened wall of 1st part of the duodenum with scarring, but no free perforation was found. We looked at the small bowel from the ligament of Treitz to the terminal ileum, and there was no sign of bowel issues, just observing the multiple emphysematous protrusions. The colon appeared normal. It seemed that a lot of the extra air thought to be outside the bowel was actually from the blebs, and the free air seen on the imaging was likely caused by a ruptured bleb. We filled the stomach and duodenum with air, and there was no bubbling of the irrigation in the abdomen. We also took the samples of cysts from the wall for histopathology examination.

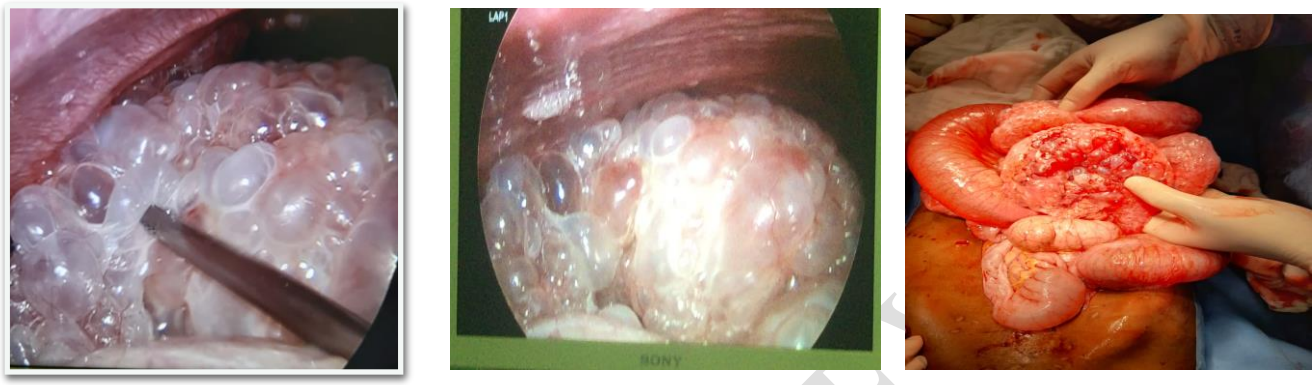

Figure 3: Intra-operative findings: numerous gas-filled thin-walled cysts in the bowel wall, of various sizes, some of them adherent to the parietal peritoneum

Postoperatively, the patient initially did well and was able to tolerate an oral diet. He was discharged from the hospital on the 5th postoperative day. However, he returned after two weeks with symptoms of epigastric fullness and vomiting. The patient was readmitted, and a nasogastric tube (NGT) was inserted for fluid resuscitation and correction of electrolyte imbalance. He was discharged after his improvement in treatment. Eight months later, he returned with the same complaints and was readmitted. He underwent resuscitation, and an upper endoscopy was performed, which revealed superficial erosions in and around the antrum, as well as a small Forest III pre-pyloric ulcer. A stenotic pyloric ring was also observed, for which balloon dilatation was applied. After the procedure, the patient improved and was discharged on a proton pump inhibitor (PPI) and ondansetron PRN. Despite this, he continues to experience intermittent abdominal discomfort.

Discussion

Pneumatosis intestinalis (PI) is a rare pathological condition characterized by gas-filled cysts in the intestinal submucosa and subserosa(3). Even though it seems like there are many factors involved in PI, we still don't really know what the exact cause is(6). There are two main theories that have been suggested in medical research(7). A mechanical theory suggests that gas can enter the bowel wall from the intestinal lumen through breaches in the mucosa or from the lungs through the mediastinum(8). After entering the bowel wall, gas can travel along the mesentery to other areas(8). This explains the link between PI and conditions that compromise mucosal integrity, including intestinal infections, IBD, necrotizing enterocolitis, caustic ingestions and intestinal ischemia(9). This theory talks about how PI is linked to obstructive pulmonary disease. Coughing in these patients might lead to alveolar rupture. Air can then move along blood vessels into the mediastinum, pass through the diaphragm, and finally reach the mesenteric root and the mesenteric blood vessels that go through the bowel(10). In bacterial theory, PI happens when gas-forming bacteria get into the submucosa through tears in the mucosa or when the mucosa becomes more permeable, leading to gas production within the bowel wall(11). The response of the PI to therapy with antibiotics supports this theory. A mix of both theories could definitely make sense. Bacterial overgrowth in the gastrointestinal tract can happen for various reasons and may result in too much hydrogen gas being produced, causing the bowels to swell and leading to the dissection of hydrogen gas into the bowel wall. Patients taking alpha-

glucosidase inhibitors have been reported to experience PI, which is linked to increased intestinal gas production.

Pneumatosis intestinalis can impact any section of the gastrointestinal tract. About 42% of PI cases affect the small bowel, 36% involve the colon, and 22% include both the small bowel and colon(12). The cysts can be located in the mucosa, submucosa, subserosa, or they might affect all three layers. Subserosal cysts are usually found in small intestinal pneumatosis, whereas submucosal cysts tend to occur more frequently in colonic pneumatosis(1).

Many patients who have pneumatosis intestinalis don't show any symptoms, particularly when it's linked to lung disease(13). Patients who seek clinical help show symptoms linked to either the presence of PI, like bleeding, obstruction, or abdominal pain because of the underlying disorder connected to PI(14). A key characteristic of PI is the presence of pneumoperitoneum without any signs of peritoneal irritation due to a cyst rupture(14). On the other hand, some cases might show air retention that leads to acute abdominal findings(6). The symptoms related to PI vary based on which part of the intestine is impacted(15). The most common symptoms in patients with colonic pneumatosis include constipation, diarrhea, distention, abdominal pain and hematochezia(15). Some other symptoms are flatulence, a decrease in appetite and tenesmus (16). Physical exams are usually normal unless there are signs of peritoneal issues from intestinal perforation in cases of PI caused by serious conditions.

Different imaging techniques are being used to diagnose PI. Abdominal radiography and CT scans are the most commonly used methods for diagnosing PI. CT shows greater sensitivity compared to radiography when it comes to detecting PI(17). CT is known to be more sensitive in detecting hepatic portal and portomesenteric venous gas, which can indicate the possibility of PI from serious causes(18). In both CT scans and radiographs , PI typically shows up as a low-density linear or bubbly gas pattern within the bowel wall. It might be a mix of both bubbly linear gas in the bowel wall. There might also be circular gas collections in the bowel wall(18). The findings from the colonoscopy might resemble multiple polyposis or groups of submucosal tumors, but subserous pneumatosis could potentially be overlooked(19). Laparoscopic exploration is really helpful for confirming a PCI diagnosis when the physical exam results seem suspicious, especially in cases that weren't diagnosed before surgery using the radiological methods mentioned earlier(20). Diagnostic laparoscopy is useful because it allows for an easy switch to open surgery if needed and helps confirm the diagnosis. Some patients with PI need urgent surgery, while others can be treated with conservative methods(21). Emergent exploratory laparotomy is necessary if there are any of the following: signs of peritonitis during the physical exam, metabolic acidosis, lactate levels greater than 2.0 mmol/L, or the presence of portal venous gas(22). Surgery might not be necessary for pneumoperitoneum caused by PI if there aren't any clinical signs indicating an underlying acute abdominal emergency(22).

Treatment needs to focus on the underlying cause of PI, even if there are no symptoms present. Stopping the medication that caused the issue, like alpha-glucosidase inhibitors or targeted therapy, might lead to the resolution of PI(23). Patients who are asymptomatic typically don’t need any extra treatment since gas cysts generally go away on their own after some time(24). For patients who have mild symptoms, using a combination of antibiotics and an elemental diet has been effective in resolving PI, likely by changing the colonic microflora(23). Inhalational oxygen or hyperbaric oxygen therapy is considered the best treatment for severe cases of PI(25). This is because high-flow oxygen therapy increases the venous oxygen concentration, which is thought to lower the partial pressure of nitrogenous gases(25). This process helps to reduce gaseous cysts by releasing the gases inside them, which are then replaced with oxygen(25). The oxygen is metabolized, leading to resolution and eliminating anaerobic gut bacteria due to its toxic effects on those bacteria(25). Surgery is something that should be considered for patients with PI who still have symptoms even after trying medical treatments or who face complications from PI like necrotic bowel, peritonitis, perforation and bowel obstruction(26).

Conclusion

PI is an uncommon condition of poorly understood etiology. When coincident with pneumoperitoneum, which may be due to rupture of emphysematous blebs without bowel wall injury, it can mimic perforated hollow viscus and surgical intervention might be performed to establish the diagnosis. More conservative management has been proposed even in the case of concomitant pneumoperitoneum.

Disclaimer (Artificial intelligence)

Author(s) hereby declare that NO generative AI technologies such as Large Language Models (ChatGPT, COPILOT, etc.) and text-to-image generators have been used during the writing or editing of this manuscript.

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APPENDIX

Table A1. Causes of Pneumatosis Intestinalis in the Adult: Benign and Life-Threatening Causes and Associations

A. Benign causes	**Medications**
Pulmonary	• Corticosteroids
• Asthma	• Chemotherapeutic agents
• Bronchitis	• Lactulose
• Emphysema	• Sorbitol
• Pulmonary fibrosis	• Voglibose
• Positive end-expiratory pressure (PEEP)	Organ transplantation
• Cystic fibrosis	• Bone marrow
Systemic disease	• Kidney
• Scleroderma	• Liver
• Systemic lupus	• Cardiac
• AIDS	• Lung
Intestinal causes	• Graft versus host
• Pyloric stenosis	Primary pneumatosis
• Intestinal pseudoobstruction	• Idiopathic (up to 15% of cases and usually involves the colon)
• Enteritis	• Pneumatosis cystoid intestinalis
• Peptic ulcers	**B. Life-threatening causes**
• Bowel obstruction	Intestinal ischemia
• Adynamic ileus	Mesenteric vascular disease
• Inflammatory bowel disease	Intestinal obstruction (especially strangulation)
• Ulcerative colitis	Enteritis

• Crohn's disease	Colitis
• Leukemia	Ingestion of corrosive agents
• Perforated jejunal diverticulum	Toxic megacolon
• Whipple's disease	Trauma
• Intestinal parasites	Organ transplantation (especially bone marrow transplants)
• Collagen vascular disease (especially scleroderma)	Collagen vascular disease
• Diverticulitis	
Iatrogenic	
• Barium enema	
• Jejunoileal bypass	
• Jejunostomy tubes	
• Postsurgical anastomosis	
• Endoscopy	

Note—A number of causes and associations occur under both benign and life-threatening categories.