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Pneumatosis Cystics Intestinalis in Adults Revealed by Peritonitis Due to Intestinal Perforation: A Case Report

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Abstract **Case Report**

Pneumatosis cystics intestinalis (PCI) is a rare condition characterized by the presence of gas within the wall of the gastrointestinal tract. It must not be confused with pseudo-pneumatosis (intra-luminal gas trapped between feces and the digestive wall). Its presence alone is not indicative of severity. Suspected pathophysiology (mechanical or bacterial), localization, and radiological characteristics (linear, bubbly, cystic) can guide towards different etiologies, including vascular (mesenteric ischemia), pulmonary, digestive, infectious, or autoimmune causes. In the absence of poor prognostic factors, surgical intervention is not always necessary.

Keywords: Cystic Intestinal Pneumatosis, Mesenteric Ischemia, Colon, Small Intestine, Acute Peritonitis, Bowel Obstruction, Pneumoperitoneum.

INTRODUCTION

Intestinal pneumatosis (IP) refers to the presence of gas in the digestive wall, a relatively rare radiological finding. Numerous causes, ranging from benign to life-threatening, may be responsible. However, it is often associated with severe conditions, such as mesenteric ischemia, requiring urgent medical or surgical intervention [1]. Two main forms of IP are identifiable: linear IP and cystic IP, though a mixed form is also possible. The type of IP does not determine its severity, and it is crucial to correlate imaging findings with clinical and biological data, in close collaboration with the clinician [2–4]. It is essential to distinguish [3-5]:

- Chronic cystic intestinal pneumatosis (CCIP), a stable, long-lasting condition, often secondary to a wide range of digestive or extra-digestive conditions but frequently idiopathic and not typically associated with emergencies like necrosis, intestinal perforation, or acute peritonitis.
- Intestinal pneumatosis or intestinal emphysema, which is always secondary and observed during acute infectious, ischemic, or toxic conditions of the gastrointestinal tract.

CASE REPORT

A 67-year-old male, with no known medical or surgical history, presented to the emergency department with acute, progressively worsening right iliac fossa pain for 5 days, which became generalized and was accompanied by vomiting, cessation of stool and gas passage, fever, and deterioration of his general condition.

On questioning, the patient reported a vague history of moderate-intensity abdominal pain. Clinical examination revealed an unstable patient, tachycardic (120 beats per minute), tachypneic (24 breaths per minute), and hypotensive (83/49 mmHg). Abdominal examination showed diffuse tenderness with guarding, especially localized in the right iliac fossa. Laboratory results indicated leukocytosis (white blood cell count of 20,000) with neutrophil predominance and an elevated C-reactive protein (CRP) level of 300 mg/L. Renal function was slightly impaired, but the rest of the results were unremarkable.

Abdominal X-ray revealed free air (pneumoperitoneum) in the hepato-diaphragmatic region. Given the patient's clinical presentation and imaging findings, after a brief period of preparation, the

patient was taken to the operating room. Surgical exploration found approximately 3 liters of purulent fluid and a small bowel perforation located 60 cm from the ileocecal valve and the presence of Pneumatosis cystics intestinalis. The procedure consisted of converting the perforation into an ileostomy, followed by lavage and drainage. The patient was discharged after 5 days.

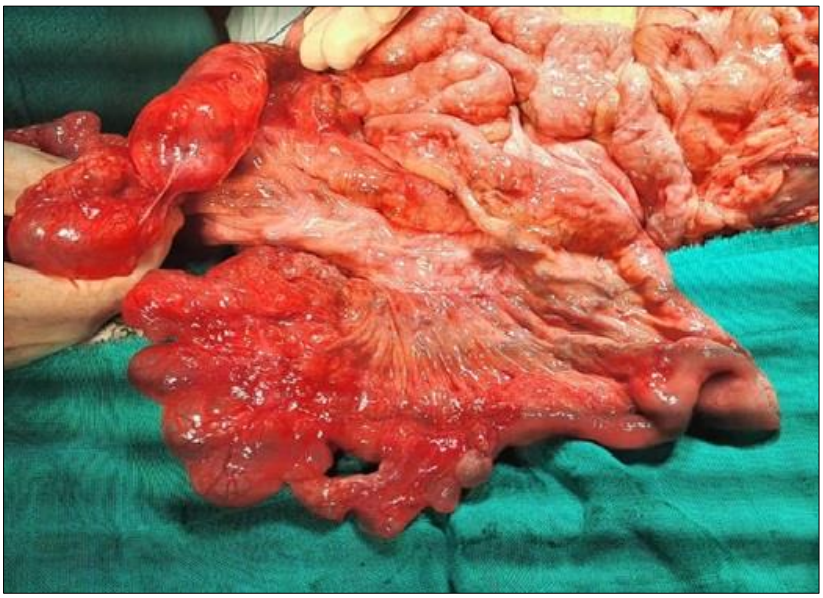

Figure A: Intraoperative image showing Pneumatosis cystics intestinalis

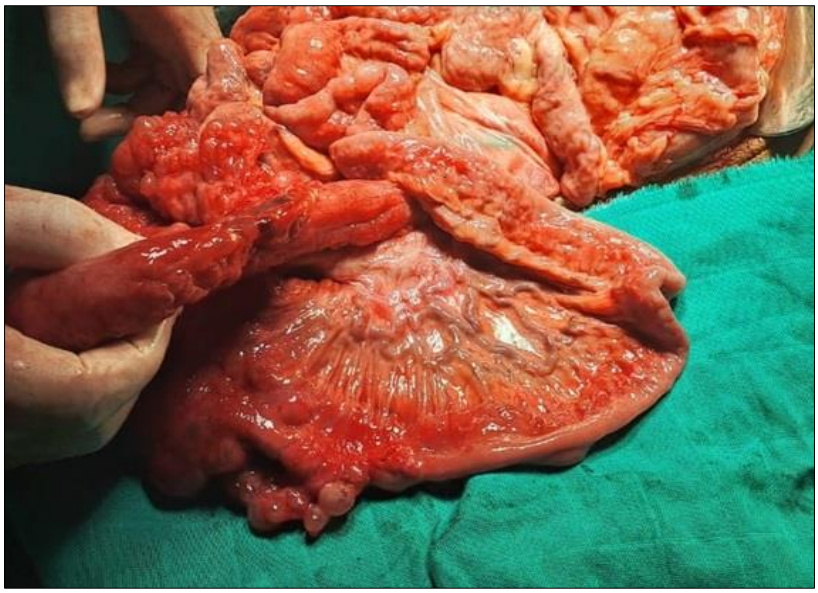

Figure B: Intraoperative image showing Pneumatosis cystics intestinalis

Pathophysiology

Although the exact cause of intestinal pneumatosis remains unclear, three main theories have been proposed in the medical literature:

1. Mechanical Theory:

Gas dissects into the intestinal wall either from the intestinal lumen or the lungs via the mediastinum due to a pressure-increasing mechanism (e.g., intestinal obstruction or emphysema). Gas dissects through mucosal tears due to local intra-luminal pressure secondary to digestive obstruction [3].

2. Pulmonary Theory:

Gas diffuses from the lungs to the digestive serosae via intra-thoracic hyperpressure, following peri-vascular or peri-lymphatic pathways through the mediastinum. During respiratory diseases such as chronic obstructive pulmonary disease (COPD) or asthma, coughing could be responsible for gaseous dissection along vessels, from the mediastinum to the mesentery, and eventually to the intestines [5].

3. Bacterial Theory:

Gas-producing bacteria penetrate the submucosa through mucosal tears or increased mucosal

permeability, producing gas within the intestinal wall [6]. Anaerobic bacteria produce hydrogen, which accumulates in the submucosa. Four primary gases are present in the intestinal lumen: carbon dioxide, oxygen, nitrogen, and hydrogen. Carbon dioxide and oxygen are highly diffusible, while nitrogen and hydrogen are less so. Persistent elevation of hydrogen partial pressure leads to gas accumulation in the digestive wall, preventing nitrogen diffusion into the lumen. Hydrogen accumulation is responsible for the formation of cysts in pneumatosis. Patients with cystic intestinal pneumatosis (CIP) are thought to be high hydrogen producers with insufficient aerobic flora to convert hydrogen into methane.

DISCUSSION

Cystic intestinal pneumatosis (CIP) is a rare condition characterized by the presence of gas cysts in the intestinal wall. It can affect the entire digestive tract, with a predilection for the small intestine and colon, and is more common in men over 50 [7]. CIP can be idiopathic or more frequently secondary to various diseases.

Primary forms predominantly affect the left colon, with mainly submucosal gas cysts, while secondary CIP affects the small intestine, with subserosal gas cysts.

The origin of this condition is multifactorial. The long list of pathological associations has led to the development of various etiopathogenic theories, likely combining multiple mechanisms. Often asymptomatic and incidentally discovered, CIP can also present with mucous-bloody emissions, abdominal pain, or diarrhea. Endoscopic and radiological examinations easily confirm the diagnosis [4], avoiding unnecessary laparotomies. The pathophysiology remains poorly understood, with three current theories [2-4]:

- Formation of cysts due to local intra-luminal hyperpressure or a procedure-related cause.
- Thoracic hyperpressure with gas diffusion to the digestive serosae.
- Microbial fermentation by gas-producing bacteria.

The treatment varies depending on the etiology. For primary forms, antibiotics aimed at reducing hydrogen-producing colonic flora are the first-line treatment [8]. If this fails, oxygen therapy (via mask or hyperbaric) may be attempted to replace hydrogen with oxygen. For secondary forms, treatment targets the underlying condition. In most cases, CIP is asymptomatic, requiring no treatment. Surgery should be reserved for particularly severe or complicated cases.

CONCLUSION

Cystic intestinal pneumatosis is a rare, generally benign condition, with no specific symptoms, and its pathophysiology is still debated. Its diagnosis is based on imaging data, and it is most often associated with a benign prognosis.

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Conflict of Interest Declaration: The authors declare no conflicts of interest.

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