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**Case Report**

# Pneumatosis intestinalis as a manifestation of chronic intestinal volvulus: case report

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## ABSTRACT

The pneumatosis cystoides intestinalis (PCI) is a rare disease characterized by presence of gaseous cysts containing nitrogen, hydrogen and carbon dioxide in the intestinal wall that can be idiopathic or associated with other diseases. There are 5 theories about the pathophysiology: mechanic (obstruction), immunosuppression (atrophy of Peyer's patches), bacterial (*Clostridium* spp.), pulmonary (chronic pulmonary disease) and chemical (exposure to trichloroethylene). The aim of this paper is to present a clinical case of a patient with rheumatic comorbidity admitted to the emergency service and diagnosed with acute abdomen secondary to intestinal volvulus associated with pneumatosis intestinalis.

**Keywords:** Volvulus, Pneumatosis, Acute abdomen

## INTRODUCTION

Intestinal pneumatosis is characterized by the presence of subserosal/submucosal gas-filled cysts in GI tract. Most common locations are colon (47%), small intestine (27%) and stomach (5%). Exact etiology is still unknown; it may occur in association with intestinal obstruction, intestinal ischemia, rheumatologic diseases, pulmonary pathology, steroid use, among others.[1-4] Primary form of cystic intestinal pneumatosis is asymptomatic, while secondary form may be associated with pulmonary/ intestinal pathology and, depending on presentation may/may not require urgent surgical management.

## CASE REPORT

### *Background*

The patient is a 67-year-old female with a history of systemic lupus erythematosus since 2012, managed with prednisone and hydroxychloroquine; rheumatoid arthritis since 2021, managed with meloxicam, tramadol, and paracetamol; and Sjögren's syndrome since 2023, managed with leflunomide and mycophenolate.

The patient reported the onset of symptoms on 22.04.23 with abdominal pain of intensity 10/10, localized in the epigastrium, exacerbated by physical activity. Initial management by a private physician showed poor improvement. After 24 hours, abdominal pain persisted, accompanied by anorexia and diaphoresis. The patient sought evaluation at hospital general de México, where laboratory tests revealed an elevation of acute-phase reactants. Abdominal standing X-rays showed free subdiaphragmatic air. The patient completed a pre-surgical protocol and underwent emergency surgery due to suspected gastric perforation (Figure 1).

### *Surgery*

An emergency exploratory laparotomy was performed, involving evisceration, revealing a mesentero-axial

intestinal volvulus with intestinal pneumatosis extending up to 120 cm distal to the Treitz angle. Retrograde dilation of jejunum and ileum loops, venous congestion, chylous ascites, and diffuse thickening of small intestine loops were identified. A 360-degree counter clockwise reduction of the intestinal volvulus was carried out. No disruptions in the abdominal wall or stenotic areas were identified. The intestines were repositioned using the Noble manoeuvre, and the procedure was concluded (Figures 2-4).

**Figure 1: Posteroanterior chest X-ray showing free subdiaphragmatic air suggestive of perforation at some level of the gastrointestinal tract. During the intraoperative period, no site of intestinal perforation was identified.**

**Figure 2: Jejunal loop showing intestinal wall edema, venous congestion, chylous ascites, and intestinal pneumatosis in the mesentery of the involved intestinal segment.**

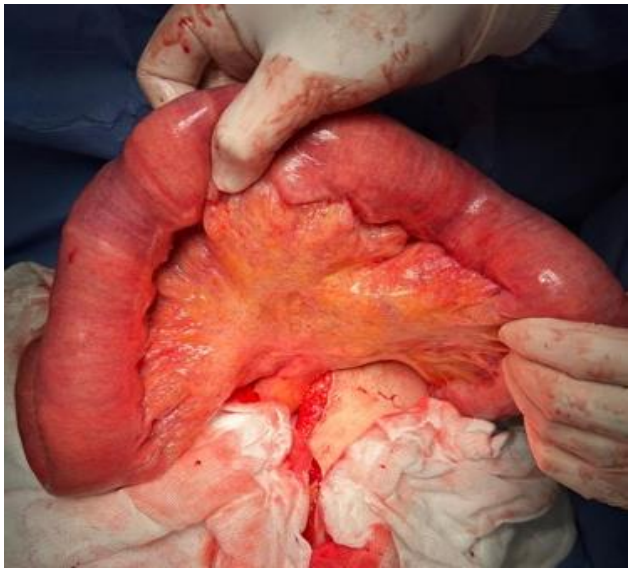

**Figure 3: Jejunal loop showing intestinal pneumatosis and pneumatosis in the mesentery.**

**Figure 4: Site of mesenteroaxial intestinal volvulus, managed with a 360-degree counter clockwise derotation.**

***Postoperative management***

The patient was kept fasting for 48 hours, ensuring adequate hydration and correcting any electrolyte imbalances. Analgesic management included nonsteroidal anti-inflammatory drugs, a COX-3 inhibitor, and gastroprotection. Oxygen therapy at 100% with a reservoir mask was initiated. A short course (5 days) of antibiotic therapy with ceftriaxone and metronidazole was prescribed due to the risk of bacterial translocation. Oral intake with a liquid diet was initiated once bowel sounds were auscultated, and parenteral nutrition support was provided for a week to meet nutritional requirements.

On May 3, 2023, an abdominal tomography was performed, confirming the resolution of intestinal

pneumatosis. Due to the patient's favourable clinical progress, a decision was made for discharge with follow-up through outpatient consultations.

## DISCUSSION

The first report of intestinal pneumatosis was made by Duvernoy in 1754 in France.[5] Cystic intestinal pneumatosis is a condition characterized by the presence of numerous cysts within the submucosal and subserosal layers of the intestine.[6] It can occur in benign conditions as well as in potentially life-threatening gastrointestinal diseases.[7] It is considered an indicative sign of advanced intestinal infarction.[4] In the patient in this clinical case, the presentation of intestinal pneumatosis was associated with rheumatologic comorbidity.

### *Epidemiology*

The prevalence of intestinal pneumatosis is 0.03% in the general population.[8] The average age of onset is 45 years with a range of 2-81 years, and the male-to-female ratio is 2.5-3.5:1.[2,3,5] The reported average disease course is 6 months.[5] The overall reported mortality rate is 20-30.4%, and the computed tomography (CT) finding most frequently associated is intestinal ischemia. The mortality of non-ischemic pneumatosis intestinal is 7.7%.[1] The high mortality associated with intestinal pneumatosis is attributed to its association with potentially lethal conditions such as intestinal ischemia. However, in this clinical case, the finding of pneumatosis was associated with mesenteroaxial intestinal volvulus, which had a subacute course.

### *Classification*

*Secondary intestinal pneumatosis*: The 85% of cases are associated with other pathologies. Life-threatening presentations warrant emergency surgical review and are related to underlying acute conditions such as intestinal ischemia.[5,9,10] Subacute presentations have been described in association with the following pathologies: chronic obstructive pulmonary disease, intraabdominal abscess, diverticular disease, intestinal obstruction, inflammatory bowel disease, acute gastric dilation, organ transplantation, barium enema, steroid use, volvulus, intestinal cancer, abdominal trauma, corrosive ingestion, haemodialysis, infection by parasites, bacteria or HIV, vasculitis, collagen diseases, and leukemia.[3,4,10]

*Primary or idiopathic intestinal pneumatosis*: represents 15% of intestinal pneumatosis cases, and typically, patients do not present with underlying pathology.[2,5,9]

### *Pathophysiology*

The pathophysiology is not fully understood but depends on the underlying cause[6]. Four theories have been proposed: mechanical, bacterial, pulmonary, and chemical. The theory of immunosuppression should be considered as an additional theory, related to the mechanical theory but with a different mechanism:

*Mechanical theory*: It argues that two conditions are necessary for the development of intestinal pneumatosis: distal obstruction that increases intraluminal pressure and mucosal damage that allows air insufflation through the damaged intestinal wall into the lymphatic channels and its subsequent distal diffusion secondary to peristalsis. Intestinal occlusion and volvulus are two conditions that fulfil the aforementioned theoretical definition, although any pathology that erodes the intestinal mucosa has the potential to develop intestinal pneumatosis.[1,2]

*Immunodeficiency theory*: It has been hypothesized that steroids and some antineoplastics cause lymphoid depletion, atrophy of Peyer's patches, and subsequent thinning of the intestinal mucosa, resulting in increased mucosal permeability that allows gas dissection into a non-inflamed abdominal wall. The increased permeability caused by defects in lymphoid tissue of the intestinal wall may facilitate the invasion of gas-producing aerogenic bacteria. Chemotherapy agents that have been associated with this include cyclophosphamide, cytarabine, vincristine, doxorubicin, daunorubicin, etoposide, docetaxel, irinotecan, and cisplatin.[7,9]

*Bacterial theory*: It is proposed that there is invasion of gas-producing bacteria in the intestinal submucosa, leading to the accumulation of intraparietal air.[1,2] The bacteria suggested to be involved include *Clostridium perfringens, Clostridium difficile, Escherichia coli, and Enterobacter aerogenes*. Intestinal pneumatosis has been successfully treated with metronidazole, indirectly supporting the bacterial theory.[8]

*Pulmonary theory*: Pulmonary diseases cause alveolar rupture, leading to mediastinal emphysema, and subsequently, air travels through the perivascular spaces of the mediastinum to the aorta and mesenteric vessels towards the intestinal wall.[1,2,5,9]

*Chemical theory*: It suggests that the cause is exposure to trichloroethylene.[2,9]

Based on the aforementioned concepts, it is possible that in the patient of this clinical case, the mechanical and immunosuppression theories were determining factors in the development of pneumatosis.

### *Clinical manifestations*

Dependiendo su patología subyacente y el modo de presentación puede ser asintomático en el 15% de los casos que corresponden a neumatosis intestinal primaria, la cual se descubre incidentalmente sobre todo en el momento de su abordaje cuando los síntomas son inespecíficos. Los quistes que contienen aire pueden causar neumoperitoneo que simula una perforación intestinal, que a menudo es de naturaleza benigna.[11] Los

pacientes pueden presentar síntomas abdominales inespecíficos como dolor abdominal (59%), diarrea (53%), náusea y emesis (14%), moco en las heces (12%) y hematoquezia (12%).[3,5] El 3% se presenta con complicaciones como neumoperitoneo, vólvulo, obstrucción intestinal e isquemia intestinal.[5]

***Laboratory tests***

Laboratory tests are normal in patients with primary pneumatosis, while when pneumatosis is associated with mesenteric ischemia or intestinal infarction, there is leukocytosis with a predominance of polymorphonuclear cells.[9]

***Imaging studies***

*X-rays*: The "fingerprints" sign, suggestive of intestinal wall edema, can be observed, and gas in the portal vein (branching in the liver) may also be visualized.[12]

*CT scan*: Numerous small cysts (grape cluster-like image) aligning with the intestinal wall can be observed.[2,5] The images are clearer when viewed in a lung window. Gas in the portal vein is reported in 25.5% of cases, increasing mortality to 50%.[5]

*Endoscopy*: The images present as protruding lesions. The differential diagnosis includes Crohn's disease, so a biopsy can be taken in case of diagnostic uncertainty. Endoscopic ultrasound has the capability to provide a definitive diagnosis.[5,10]

***Management***

The management is guided by the severity of clinical presentation, which can vary depending on the underlying pathology.[6] Attention is focused on the underlying disease responsible, as the majority of cases of intestinal pneumatosis are asymptomatic and secondary to an underlying condition.[8,10] Remission has been reported in 70% of patients with non-surgical management.[5] Antibiotic therapy is recommended until clinical or radiological resolution of pneumatosis, with metronidazole being the most commonly used antibiotic, inhibiting bacterial growth and thereby preventing hydrogen production. The use of parenteral nutrition has been reported to be effective.[9]

*Oxygen*: Oxygen at high flow and hyperbaric oxygen are treatment strategies for intestinal pneumatosis.[13] It has been suggested that the use of high-concentration oxygen induces a washout phenomenon of nitrogen and gas particle diffusion.[14] The rationale for recommending oxygen as a treatment modality in intestinal pneumatosis is the same as described for managing small pneumothoraxes (<30%), subcutaneous emphysema, and pneumomediastinum. Extrapulmonary air in the interstitial tissue is reabsorbed in the capillaries by diffusion following the pressure gradient caused by the sum of partial pressures exerted by water, $CO_2$, nitrogen, and oxygen. When breathing 100% oxygen, nitrogen is reabsorbed out of the blood, indirectly increasing the absorption gradient of oxygen at the tissue/capillary interface, which raises the absorption gradient of one gas (and indirectly of the others) and causes a 4-6 times increase in the gas reabsorption rate.[15,16]

*Surgical management*: Surgical management is the second line of treatment and is reserved when there is peritoneal irritation or persistent intestinal obstruction[9]. It is suggested to consider surgery for patients with symptoms of intestinal obstruction, leukocytosis, gas in the portal vein, those over 60 years of age, acidosis (pH <7.2, bicarbonate <20 ml/l, lactate >2 mmol/l), acute abdomen, acute kidney injury, or hypotension[5].

***Intestinal volvulus***

Intestinal volvulus is a torsion of the small intestine around its mesenteric axis. In adults, volvulus can be primary if there are no predisposing conditions or secondary if it is associated with underlying anatomical abnormalities (adhesions, tumors, previous stomas, pregnancy, intestinal diverticula). In 89% of cases, it presents acutely with intestinal ischemia and peritonitis. Computed tomography is the diagnostic method of choice, where the characteristic swirling pattern of the intestine can be observed, and signs of ischemia, infarction, or pneumatosis can also be identified. Management depends on the presentation; if it is acute, exploratory laparotomy is the preferred approach (BAUMAN).

## CONCLUSION

In acute abdomen, intestinal pneumatosis can be associated with cases of intestinal volvulus. In intestinal volvulus, the cause of pneumatosis follows the mechanical theory: obstruction plus damage to the intestinal mucosa. If there is no discontinuity in the intestine, the management of intestinal pneumatosis is conservative, involving adequate fluid and electrolyte support, high-flow oxygen, and antibiotic therapy. The management is guided by the severity of the clinical presentation, which can vary depending on the underlying pathology.

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