Pneumatosis intestinalis as a presentation of Crohn’s disease: a case report

Neumatosis intestinal como presentación de la enfermedad de Crohn: reporte de un caso

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ABSTRACT
Pneumatosis intestinalis (PI) is defined as presence of gas within the intestinal wall. It is a rare condition, usually associated with a wide variety of pathologies. It requires a special diagnostic approach to determine underlying etiology. We present the case of a 18 year old woman with chronic abdominal pain, who presents with peritoneal signs and pneumoperitoneum. Laparoscopy showed NI. Thereafter, Crohn’s disease was diagnosed by balloon enteroscopy.

Keywords: Pneumatosis intestinalis; Pneumoperitoneum; Crohn disease; Diagnosis; Surgery (source: MeSH NLM).

RESUMEN
La neumatosis intestinal (NI) es la presencia de gas en la pared intestinal. Es un hallazgo infrecuente, generalmente presente en una gran variedad de patologías. Requiere de un abordaje diagnóstico detallado para determinar su etiología. Presentamos el caso de una mujer de 17 años con dolor abdominal crónico, quien se presenta con signos de irritación peritoneal y neumoperitoneo. Con hallazgo quirúrgico de NI en el intestino delgado, a quién mediante enteroscopia se le diagnóstica enfermedad de Crohn.

Palabras clave: Pneumatosis intestinalis; Neumoperitoneo; Enfermedad de Crohn; Diagnóstico; Cirugía (fuente: DeCS BIREME).

INTRODUCTION
Pneumatosis intestinalis (PI) is the presence of gas within the intestinal wall. It represents an imaging or surgical finding, and not a pathological entity per se [1].

It is classified as primary (15%) when no possible etiological factor is identified, and as secondary (85%) in association with a wide variety of conditions and pathologies, including asthma, trauma, intestinal ischemia, abdominal infections, medications, immunosuppression, and inflammatory/autoimmune diseases, among others [2].

Its pathogenesis is unclear. However, theories have been proposed to explain its formation. The proposed mechanisms include damage in the intestinal mucosa, gas generation by intestinal bacteria, and gas from the thorax in relation to pulmonary diseases [3].

It may be asymptomatic or have nonspecific gastrointestinal symptoms. Management depends on the benign or catastrophic nature of its clinical presentation [1-3].

CASE REPORT
Eighteen-year-old female patient with a history of intermittent episodes of chronic abdominal pain located in the right half of the abdomen, predominantly in the ipsilateral iliac fossa, and nausea since the age of 14. During the last 2 years, she presented increased frequency of abdominal pain, associated with abdominal distension, emesis and episodes of diarrhea without blood, lasting 2 days, which occurred 2-3 times a month, without identified triggers.

In the last 2 years she had undergone endoscopic studies of the upper and lower gastrointestinal tract, with no pathological findings; 18 months ago, she had an abdomen CT scan that showed thickening of the jejunal wall and dilation of the small intestine loops. She had not received any diagnostic intervention in this regard.

She was admitted to the emergency department of our institution for worsening of abdominal pain, increased abdominal distension, emesis of food content and absence of stool for 5 days. Her physical examination
showed normal vital signs, mild dehydration, abdominal distension, and positive Blumberg sign. Her low height and weight (BMI 16) attracted attention. Paraclinical tests showed mild leukocytosis; simple abdominal x-ray showed evidence of dilation of the small intestine loops, with “stack of coins” appearance, hydroaerial levels and pneumoperitoneum (Figure 1). Due to suspected intestinal obstruction with possible hollow visceral perforation, the patient underwent diagnostic laparoscopy. On entering the abdominal cavity, the exit of abundant gas and citrine peritoneal fluid was observed. Conversion to infraumbilical laparotomy was necessary due to technical difficulties, given the presence of dilated intestinal loops. Dilation of intestinal loops, PI of the jejunum, wall erythema and mesenteric adenitis were documented, and no perforations were found.

Contrast abdominal CT was performed showing segmental thickening of the jejunum and ileum walls, mesenteric adenitis, and PI of the jejunum. Due to suspected inflammatory bowel disease (IBD), ileocolonoscopy was performed without abnormal findings. It was decided to complement the diagnostic study with a magnetic resonance enterography (Entero-MRI) of the abdomen that revealed findings similar to those described in the CT (Figure 2); due to this, enteroscopy was performed that showed evidence of ulcerated ileitis (Figure 3). The histological study revealed flattening of the villi, abundant lymphocyte-dominated infiltration of the lamina propria with polymorphonuclear cells permeating the glandular epithelium. PAS, ZN stains were negative. PCR for cytomegalovirus (CMV), herpes, cultures for common germs and for tuberculosis mycobacteria were also negative. Fecal calprotectin level was elevated (1,196 mcg/g).

With all the clinical, imaging, endoscopic and histological findings, the diagnosis of Crohn’s disease (CD) IBD type was confirmed, and systemic steroid and immunomodulatory management was instituted. Four weeks after starting therapy, the patient reported 80% improvement in her symptoms, and with the intervention of clinical nutrition, she managed to gain 3
kg of weight. The patient is in plan of starting biological medication.

DISCUSSION

PI is defined as the presence of gas within the intestinal wall. It can be cystic, microvesicular or linear in appearance, and is located in the submucosal or subserosal planes. Cysts are filled with hydrogen, nitrogen, and carbon dioxide. It was first described in 1783 by Du Vernoi as a finding of pathological anatomy. It occurs in any age group and its incidence is approximately 0.03%, although case reports tend to increase, related to the wide use of CT in the emergency department and the improvement in sensitivity of the study. Although it can occur anywhere in the gastrointestinal tract, PI is most frequently located in the colon (46%), followed by the small intestine (27%), both in the colon and in the small intestine (7%), and in the stomach (5%).

The mechanism by which PI occurs is unclear. Proposed hypotheses include: 1) mechanical theory: intraluminal gas enters the intestinal wall due to mechanical forces through ruptures of the mucosa, 2) Bacterial theory: gas-forming bacteria (such as Clostridium) penetrate the intestinal mucous membrane, producing gas that is retained in the submucosa and lymphatic vessels, 3) pulmonary theory: chronic pulmonary diseases produce alveolar rupture and as a consequence, emphysema in mediastinum with air migration along the aorta and mesenteric vessels to the abdominal wall. In our patient, the chronic inflammatory process of the mucosa associated with Crohn’s disease contributed to the pathophysiological mechanism.

More than a pathological entity, PI is a radiological finding that has been described in association with a broad spectrum of pathologies or conditions (secondary) in 85% of cases; in the other 15% the etiology is not identified (primary or idiopathic) and implies a benign chronic condition.

Based on the evaluation of multiple case reports, Feuerstein documented more than 50 different etiologies, grouped into gastrointestinal, infectious, medicinal (iatrogenic), neoplastic, pulmonary, rheumatic and transplant-associated (Table 1).

Most cases of PI are asymptomatic, so they are diagnosed as an incidental radiological or endoscopic finding. Patients may present with nonspecific gastrointestinal symptoms: mild abdominal pain (59%), diarrhea (53%), weight loss (55%) nausea and vomiting (14%), mucus or blood in the stool (12%).

Generally, patients with primary PI have hemodynamic stability, normal lactate levels, and no signs of sepsis. In contrast, patients with life-threatening conditions often present with signs of peritoneal irritation. In general terms, PI symptoms are varied, and depend on the location of the pneumatosis and the underlying etiology.

CT is the most sensitive test for diagnosing PI. Some authors have described an association between the linear pattern of PI and intestinal ischemia compared to the benign nature of the cystic pattern. However, none of these patterns is pathognomonic of any underlying pathology, and they have not been correlated with its severity or etiology. CT also allows to detect findings that may suggest the underlying etiology, as well as potentially worrisome findings (“red flags”), such as thickening of the intestinal wall, decreased uptake of contrast material by the mucosa, dilation of intestinal loops, ascites, or presence of portal gas. Our patient’s abdominal CT showed thickening of the jejunum and ileum walls and dilation of the loops of the...
Table 1. Causes of pneumatosis intestinalis.

| Category                      | Cause                          |
|-------------------------------|--------------------------------|
| Gastroenterological           | Mesenteric infarction Intussusception |
| Celiac disease                | Mesenteric ischemia Lactose intolerance |
| Colorectal cancer             | Post-endoscopy status          |
| Constipation                  | Post-surgical status           |
| Crohn’s disease               | Pyloric stenosis               |
| Diverticulitis                | Ulcerative colitis             |
| Enteritis                     | Volvulus                        |
| Hirschprung’s disease         |                                |
| Intestinal obstruction        |                                |
| Intestinal pseudo-obstruction |                                |
| Infectious                    | Lactobacillus                  |
| Adenovirus                    | Tuberculosis                   |
| Clostridium                   | Salmonella                     |
| Cryptosporidium               | Rotavirus                      |
| Cytomegalovirus               | Varicella zoster               |
| Enterobacter                   | Whipple’s disease              |
| HIV                           |                                |
| Klebsiella                    |                                |
| Medicinal                     | Immunosuppression              |
| α-glucosidase inhibitors      | Lactose                        |
| Antacids                      | NSAIDS                         |
| Bowel Preparation             | Sorbitol                       |
| Chemotherapy                  | Trichloroethylene              |
| Hydrochloride                 |                                |
| Steroids                      |                                |
| Oncological                   |                                |
| Chemotherapy                  |                                |
| Graft-versus-host disease     |                                |
| Pulmonary                     | Positive pressure ventilation  |
| Asthma                        | Pulmonary fibrosis             |
| Bronchiectasis                | Sarcoidosis                    |
| Bronchitis                    |                                |
| COPD                          |                                |
| Cystic Fibrosis               |                                |
| Rheumatic                     |                                |
| Dermatomyositis               | Scleroderma                    |
| Juvenile rheumatoid arthritis | Systemic Lupus Erythematosus   |
| Mixed connective tissue disease| Systemic Sclerosis            |
| Polyarteritis nodosa          | Sjögren’s Syndrome             |
| Polymyositis                  |                                |
| Rheumatoid arthritis          |                                |
| Transplant                    |                                |
| Bone marrow                   | Hepatic                        |
| Cardiac                       | Pulmonary                      |
| Renal                         | Pancreatic                     |
| Primary - Idiopathic          |                                |

Taken from Feuerstein 2014. (1)

our patient’s case, acute abdominal pain with signs of peritoneal irritation, imaging compatible with intestinal obstruction and pneumoperitoneum, and leukocytosis, suggested an intra-abdominal catastrophe with indication for surgical management.
In conclusion, PI may occur in patients with Crohn’s disease; this is a rare radiologic finding that should be interpreted and studied in the context of its clinical presentation. Imaging studies have a role in identifying the underlying etiology and the “red flags” indicating surgical intervention. The management is directed to that of the underlying pathology. If no serious complications occur, the prognosis is good.

Conflict of interest: The authors declare no conflict of interest.

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