CLINICAL CASE
doi: 10.35366/109774

Cirujano General
July-September 2022
Vol. 44, no. 3 / p. 136-140

Spontaneous pneumoperitoneum secondary to intestinal pneumatosis: an uncommon cause of acute abdomen

Neumoperitoneo espontáneo secundario a neumatosis intestinal: una causa poco frecuente de abdomen agudo

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Keywords:
intestinal pneumatosis, pneumoperitoneum, acute abdomen, systemic sclerosis.

Palabras clave:
neumatosis intestinal, neumoperitoneo, abdomen agudo, esclerosis sistémica.

ABSTRACT

Intestinal pneumatosis is a rare entity characterized by the gas accumulation in the submucosa or subserosa of the walls of the digestive tract, usually associated with other diseases; primary presentation is rare. In most cases, the clinical manifestations are asymptomatic, infrequent the presentation as acute abdomen, secondary to spontaneous pneumoperitoneum. We report the case of a female patient with systemic sclerosis who presented a clinical picture of abdominal pain in the emergency department, with failure to conservative treatment and progression to the acute abdomen, for which surgical treatment was offered, resolving the spontaneous pneumoperitoneum secondary to intestinal pneumatosis involving the entire small intestine, by exploratory laparotomy and intestinal rest with excellent results.

RESUMEN

La neumatosis intestinal es una entidad poco frecuente que se caracteriza por mostrar acumulación de gas en la submucosa o subserosa de las paredes del tracto digestivo, por lo general se asocia con otras enfermedades, la presentación primaria es rara. Las manifestaciones clínicas en la mayoría de los casos son asintomáticas, siendo poco frecuente la presentación como abdomen agudo, secundario a neumoperitoneo espontáneo. Reportamos el caso de una paciente con esclerosis sistémica, la cual evidenció cuadro clínico de dolor abdominal en el servicio de urgencias, con falla al tratamiento conservador y con progresión a abdomen agudo, por lo cual se ofreció tratamiento quirúrgico, con lo que se resolvió el neumoperitoneo espontáneo secundario a neumatosis intestinal que comprometía todo el intestino delgado, mediante laparotomía exploratoria y reposo intestinal con excelente resultado.

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Received: 03/30/2021
Accepted: 12/23/2022

INTRODUCTION

Intestinal pneumatosis represents a rare clinical entity characterized by the gas accumulation in the submucosa or subserosa, forming cystic lesions within the gastrointestinal tract.[1] This pathology was first reported in 1730 by Du Vernoi and subsequently subcategorized by Koss in 1952.[2,3] The incidence of intestinal pneumatosis is still not precisely known. However, it is increasingly reported as a finding due to the frequent use of computed tomography in abdominal pathologies.[4] It can occur in any age group, from neonates to geriatrics,[3] with a slight male predominance and peak presentation between 30 and 50.[5,6] One autopsy series reported an incidence of 0.03% in the general population.[3] Intestinal pneumatosis can affect any segment of the digestive tract from the esophagus to the rectum, but most frequently occurs in the small intestine (42%) (60% in jejunum, 30% in duodenum, and 10% in ileum), followed by the colon (36%) or both (22%), according to reports in the literature.[3-5] These cysts contain a mixture in variable amounts of nitrogen,

How to cite: Salgado-Vives J, Chávez-Serna E, Yáñez-Herrera GG. Spontaneous pneumoperitoneum secondary to intestinal pneumatosis: an uncommon cause of acute abdomen. Cir Gen. 2022; 44 (3): 136-140. https://dx.doi.org/10.35366/109774

hydrogen, oxygen, carbon dioxide, butane, propane, methane, ethane, and argon.[5] In approximately 85% of patients, the pneumatosis is associated with chronic pulmonary pathology, rheumatologic diseases, immunosuppression, or gastrointestinal diseases, being classified as secondary intestinal pneumatosis; the remaining 15% have no underlying pathology, its etiology being idiopathic, so it is classified as primary pneumatosis.[7] Most cases are asymptomatic, although up to 30% may manifest as acute abdomen secondary to spontaneous pneumoperitoneum due to rupture of the bullae.[5]

PRESENTATION OF THE CASE

We present the case of a 61-year-old female patient who came to the emergency department with distension, intense and diffuse abdominal pain, intolerance to oral intake, and nausea leading to vomiting with food characteristics of a week's evolution with sudden exacerbation in the last day. As important personal history, she was diagnosed with systemic sclerosis in treatment with immunomodulators, primary hypothyroidism, and chronic liver disease under study, which required therapeutic paracentesis a year ago. The physical examination revealed an afebrile and conscious patient with a globose abdomen tympanic to percussion, decreased peristalsis, pain on deep palpation in the four quadrants, without evidence of peritoneal irritation; no masses or tumors were palpated, there was no ascites fluid under tension, and rectal examination showed no alterations. Laboratory studies were requested on admission without significant alterations, and an ultrasound did not show conclusive changes, with little perihepatic ascites fluid. Conservative management was started with fasting, antibiotic therapy with a double scheme (ciprofloxacin plus metronidazole), and placement of nasogastric tube without exit of food or fecal material through it, without data of improvement, with diaphoresis and progression of abdominal pain, so it was decided to perform a contrasted computerized tomography scan of the abdomen, where air and free liquid in the cavity with high suspicion of intestinal perforation was visualized *(Figures 1 and 2)*. The surgical findings were free air in the abdominal cavity with cystic intestinal pneumatosis of benign origin in the entire small intestine, scarce ascites fluid, and no associated intestinal perforation data *(Figure 3)*. Given this situation, the diagnosis of intestinal pneumatosis was proposed. The evolution was favorable after the three-day intervention with analgesic treatment, antibiotic therapy, and intestinal rest. After six days of remission of the clinical picture, without complications, the patient was discharged and sent to the general surgery outpatient clinic for continued monitoring.

DISCUSSION

Intestinal pneumatosis is rare in which gas-containing cysts form under the intestinal

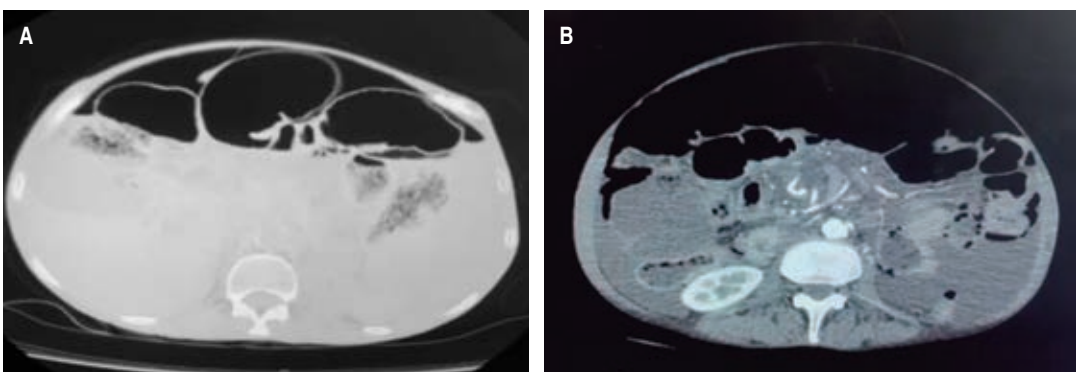

Figure 1: A) *Axial section at a hepatic level showing intestinal loops with significant dilatation, free air, and perihepatic fluid.* ***B)*** *Axial section at a renal level showing significant free air.*

mucosa and serosa. The exact pathophysiology of this disease is currently unknown, although several theories have been put forward.[7] Multiple pathologies are associated with this condition; the most common are those related to gastrointestinal, pulmonary, rheumatologic, infectious diseases, immunosuppressive treatments, and trauma secondary to endoscopic or laparoscopic processes. In our case, the patient presented rheumatologic pathology, possibly associated with a secondary cause.[2,3,5] Three theories have been described to explain etiopathogenesis: the mechanical theory, which attributes the presence of transmural air to a state of increased intraluminal pressure and mucosal damage, which causes gas to escape to the wall; the bacterial theory, which suggests that the air originates from the presence of gas-producing anaerobic bacteria (*Clostridium difficile* and *Clostridium perfringens*), and the pulmonary theory, in which the increase in intrathoracic pressure in patients with pulmonary pathology (asthma, COPD) causes rupture of the alveoli and gas extravasation reaching the mediastinum, the retroperitoneal space, the mesentery and finally, the intestinal serosa.[6]

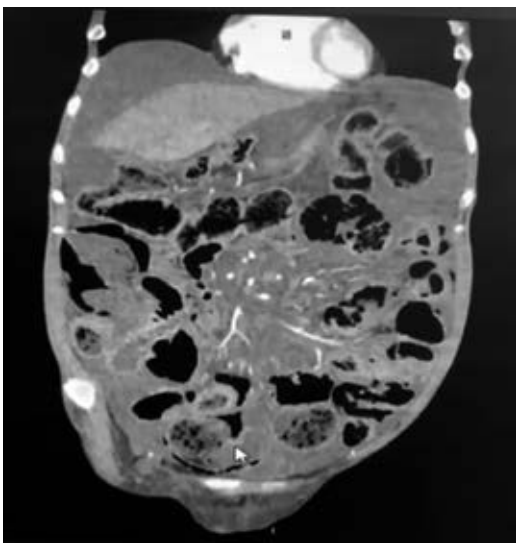

Figure 2: *Coronal section showing perihepatic and perisplenic fluid and diffuse intestinal involvement of intestinal pneumatosis.*

Most patients are asymptomatic, but when there are clinical manifestations in intestinal pneumatosis, they are nonspecific, such as abdominal pain (59%), diarrhea (53%), nausea and vomiting (14%), mucus in the stool (12%) and hematochezia (12%). Complications are infrequent but occur in approximately 30% of patients, with spontaneous pneumoperitoneum, volvulus, obstruction, and intestinal ischemia being frequent.[2,3,5,6]

The diagnosis of intestinal pneumatosis is made by exclusion, having ruled out other causes of abdominal pain, in general, by a computerized tomography scan or simple abdominal radiography. However, abdominal

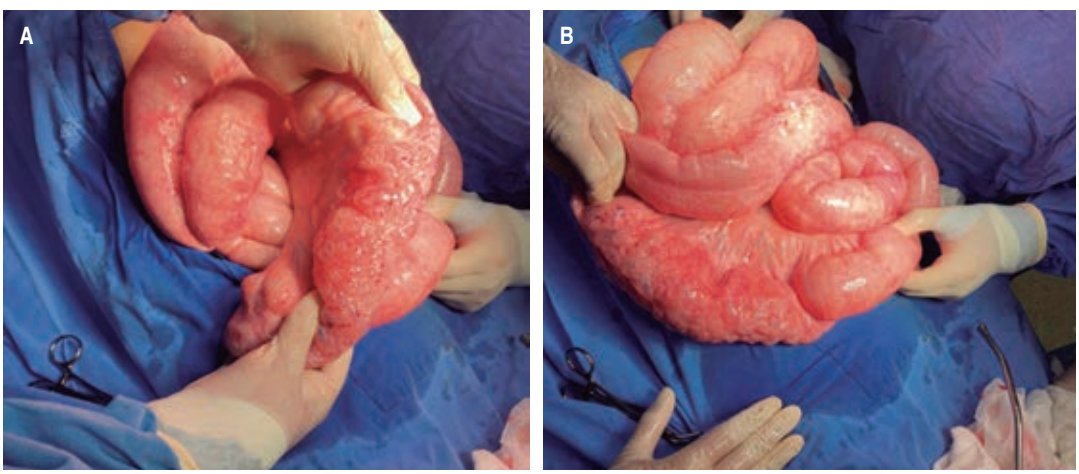

Figure 3: *Transoperative images showing intestinal pneumatosis affecting mainly the jejunum and ileum but involving the entire small intestine.*

CT scan is the most sensitive and is considered the study of choice. In our case, the patient presented with tension abdomen, but without frank data of acute abdomen on admission, and due to the history of chronic liver disease, the CT scan helped rule out other more frequent causes of abdominal pain.[4] Cysts can be described as linear, circular, or bubbly, with no direct relation to the severity of the pathology.[7] However, confusing them with intestinal polyps, cancer, inflammatory bowel disease, and necrotizing enterocolitis is easy.[2]

The differential diagnosis includes mainly visceral perforation when it starts with spontaneous pneumoperitoneum, which occurs in 30% of patients with this pathology.[7]

Laboratory abnormalities are usually the result of the underlying disease causing intestinal pneumatosis. The main clinical predictors of intestinal necrosis and mortality in patients with intestinal pneumatosis include the following: pH less than 7.3, bicarbonate level of less than 20 mEq/l, lactate level of more than two mmol/l, amylase level of more than 200 U/l, and laboratory test results consistent with disseminated intravascular coagulation (prolonged prothrombin time, decreased fibrinogen level, elevated fibrinogen degradation products, and elevated D-dimer level).[3,8,9]

The treatment of intestinal pneumatosis must be individualized according to the patient's clinical conditions. Asymptomatic patients do not require any specific treatment; to indicate conservative treatment, we must have a high diagnostic suspicion of this disease,[6,7] while in patients with mild symptoms, conservative treatment can be initiated with intravenous antibiotic therapy (metronidazole is considered the antibiotic of choice, and is used for intraluminal bacteria, thus reducing anaerobic gas production), nasogastric decompression, sclerotherapy and bowel rest (decreases the availability of substrates for bacteria) with a success rate of up to 93%.[3,6,10]

In patients with severe symptoms, with suspicion of related complications (perforation, obstruction, hemorrhage, intestinal volvulus, or portal pneumatosis) and if there are predictors of mortality in the laboratory results, emergency surgical intervention will be indicated, with exploratory laparotomy being successful in most of these cases. In this case, we opted for surgical treatment due to the progression of the patient's symptoms, the poor response to conservative treatment in the first hours, and the findings obtained from the tomographic study, where there was a high suspicion of perforation due to air and free fluid.[1,3,6]

High-flow oxygen therapy and hyperbaric oxygen have long been recognized as effective therapy for intestinal pneumatosis, leading to cyst regression on imaging and resolution of symptoms. It is currently an alternative to conservative treatment that has shown excellent results.[3] The accumulation of oxygen in the cysts increases the partial pressure of hydrogen in the cysts, which causes high-pressure diffusion of hydrogen out of the cyst into the bloodstream; cyst resolution follows with oxygen reabsorption for use in cellular metabolism. Increased oxygenation at the tissue level may facilitate phagocytic activity and directly target gas-producing organisms. To date, no complications have been reported with the use of this therapy, with a reported improvement of symptoms in 89% of patients.[3,9]

CONCLUSION

Intestinal pneumatosis is a rare entity; in most cases, it is associated with other pathologies, it is usually asymptomatic, and it is diagnosed as a finding with imaging studies; however, it can manifest as acute abdomen secondary to spontaneous pneumoperitoneum, where the presence of perforation of the hollow viscera should be ruled out as a first option. Intestinal pneumatosis should be considered a differential diagnosis causing acute abdomen, mainly when the patient has associated pathologies such as rheumatologic diseases. Currently, there is no standardized regimen for treating this pathology within conservative management; oxygen therapy seems to be an alternative with promising results that should be considered; however, each patient should be individualized to avoid complications. Surgical treatment is reserved for cases presenting acute abdomen, high suspicion of complications, and failure of conservative treatment.

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Ethical considerations and responsibility: the authors declare that they followed the protocols of their work center on the publication of patient data, safeguarding their right to privacy through the confidentiality of their data.

Funding: no financial support was received for this work.

Disclosure: the authors declare that there is no conflict of interest in carrying out the work.

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