

Pneumatosis Intestinalis: Cause of Intraabominal Free Gas as no Urgent Surgical Management. Case Report and Review of Literature

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Abstract: Pneumatosis intestinalis is a rare disease by the presence of multiple gas-filled cysts in the intestinal wall, which eventually could burst and release the gas into de abdominal cavity that can simulate and acute abdomen event. The etiology is not well known but it is attributable to rheumatologic diseases. To correctly manage the patients affected with this pathology is important to differentiate the candidates to conservatively versus surgical management.

Keywords: Pneumatosis Intestinalis, Rheumatologic Diseases, Surgical Management, Intraabominal Free Gas.

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INTRODUCTION

Pneumatosis intestinalis (PI) is defined by the presence of gas-filled cysts in submucosa, subserosa in the bowl wall or in the mesentrium [1]. Since 1730 Du Vernoy described in autopsy specimens but until 1825 Mayer named as pneumatosis cystoides intestinalis (PCI) [2, 3].

The worldwide incidence reported is 0.3-1.2%, and the prevalence of PCI is difficult to determine because mostly of the patients are asymptomatic [4], few studies have reported the incidence by intestinal segment and showed 46% in colon, 27% in small intestine and 7% colon and small intestine combined [5].

We present a 47-year-old female with history of hypertension and rheumatoid arthritis (RA) with Raynaud's phenomenon programed for elective surgery of umbilical hernioplasty, but in her preoperative protocol, infradiaphragmatic free gas in asymptomatic patient was found, the day of the surgery the thorax radiography was repeated and was corroborated.

CASE REPORT

A 47-years-old female with history of RA and Raynaud's phenomenon in treatment with prednisone and methothrexate, with last acute event 6 months before surgery date. At the Cardiologist examination infradiaphragmatic free gas was observed in the thorax x-ray, no irritative peritoneal signs was found and was discharged to home. The day of the surgery an urgent x-ray was requested, and the massive free gas was corroborated (Figure 1), and Raynaud's phenomenon was identified by Rheumatologist. (Figure 2) even so, the planned surgery continued. At physical examination we observed distended abdominal wall and a diastasis recti-like. (Figure 2) Under regional anesthesia, a supraumbilical incision was selected, dissected until abdominal cavity and we found 3x3 cm abdominal wall defect, incidental air expulsion in the moment of hernia sac opened, (Figure 4) scarce reactive intraperitoneal fluid, no intestinal content in cavity and at least 150 cm of yeyunum-ileon full covered with cysts. (Figure 5) Finally tensional hernioplasty was performed with interrupted Smead Jones closure with absorbable monofilament suture.

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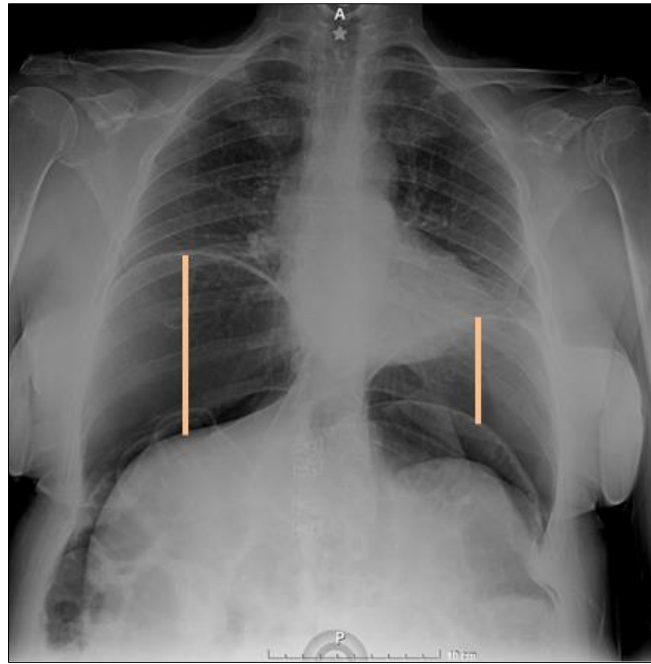


Figure 1: Massive intraabdominal free gas. Lines points out the space between diaphragm and intraabdominal organs



Figure 2: Raynaud's phenomenon



Figure 3: A: Abdomen in rest, evident umbilical defect. B: Abdomen in valsalva maneuver

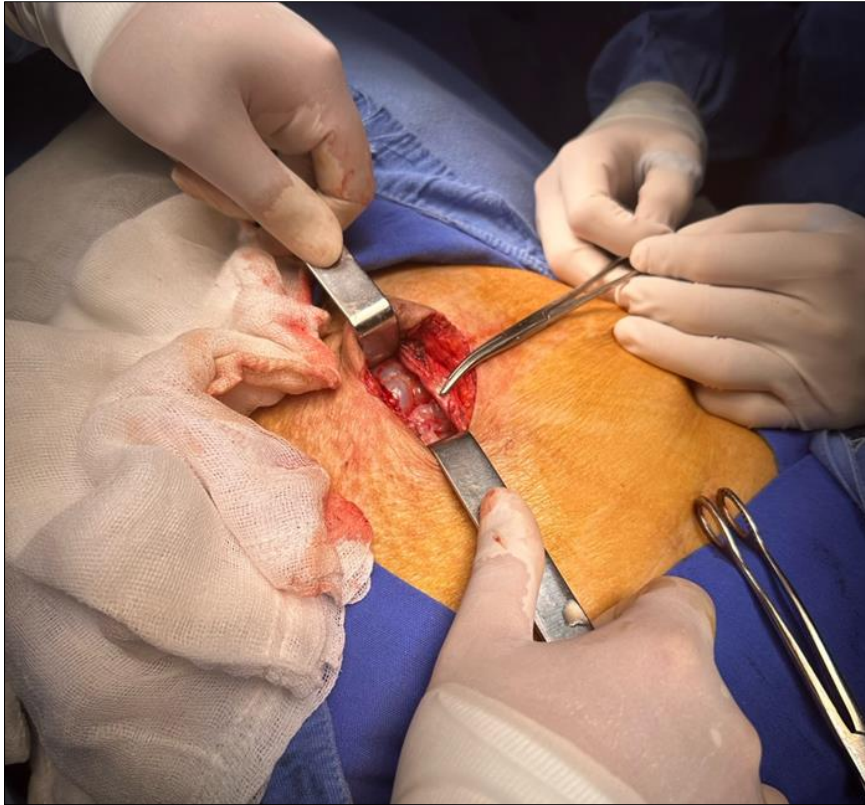


Figure 4: Umbilical defect with the hernia sac opened

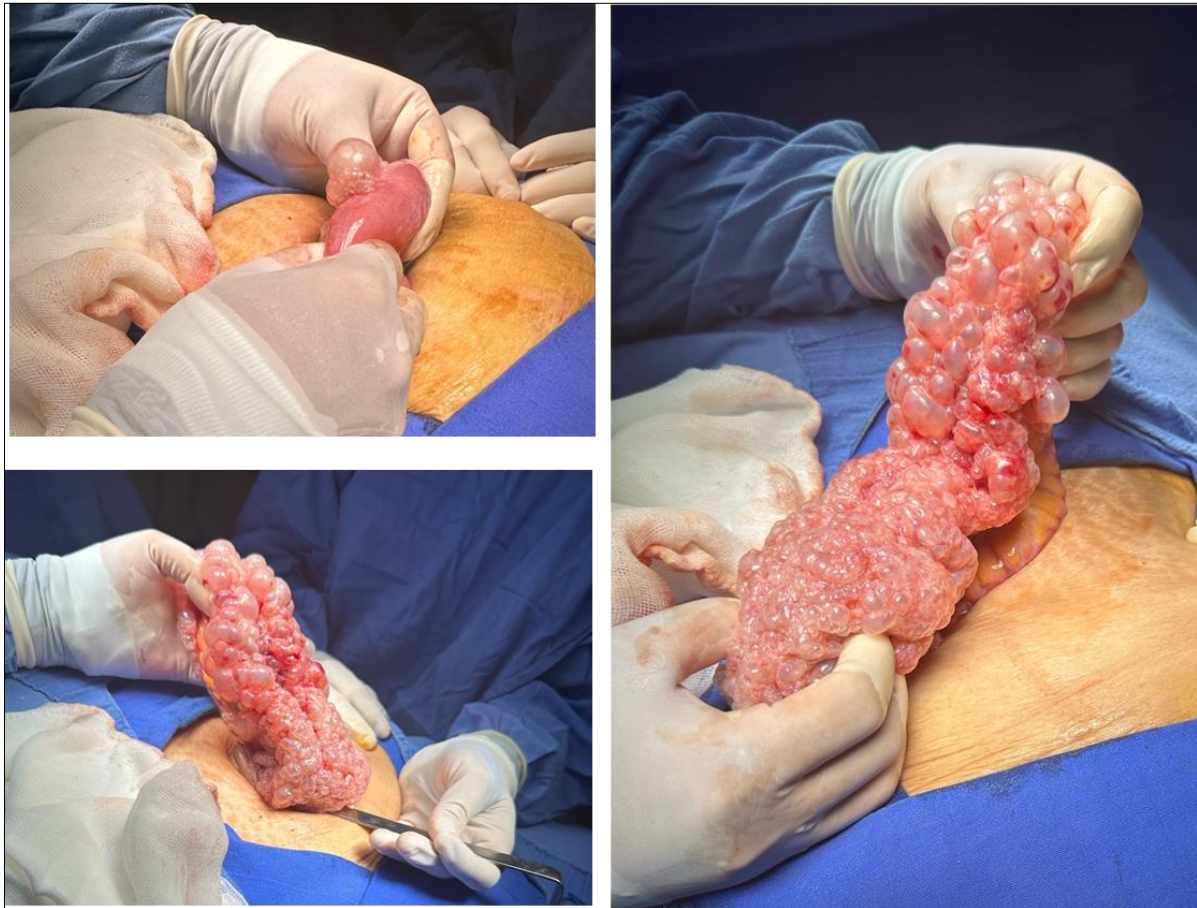


Figure 5: Transoperative images of different segmental of small bowel

DISCUSSION

The etiology of PCI has been associated to different theories: mechanical as intestinal obstruction, inflammatory bowel disease, ischemic bowel disease or colonoscopy resulting in intestinal wall injury; The pulmonary theory: COPD, asthma, interstitial pneumonia; the bacterial theory, based on gas-forming bacteria enter the mucosal barrier through mucosal rents or increased mucosal permeability; the chemical theory associated that malnutrition can prevent carbohydrates digestion and increased bacterial fermentation in the intestine; however none of these theories can explain the presence of cysts in the intestinal wall after the pathologies that could trigger it. The most recent theory reports PCI associated to connective tissue diseases (CTD) [6]. There are some reported cases with RA, ANCA-vasculitis, suggesting vasculopathy in the bowel mucosa. Frequently patients with complicated CTD possess several risk factors for development of more cysts, as bacterial overgrowth due to chronic constipation and the use of corticosteroids and immunosuppressants have also been thought to exacerbate the state of PCI [7, 8].

This diagnosis must keep in mind at Emergency Room, because it could be easy managed as acute abdomen and perform an unnecessary surgical approach. There is not specific treatment for PCI, it will be led by the presentation or biochemical deterioration for each patient. Beyene reported a patient with PCI and bowel obstruction [9], and a laparotomy was needed, any patient with a pH of less than 7.3, bicarbonate level of less than 20 mEq/L, lactate level of more than 2 mmol/L, amylase level of more than 200 U/L, or signs of disseminated intravascular coagulation should undergo emergent surgery [10]. Radiologically, the presence of portal venous gas is highly indicative of severe disease with mortality rates range from 37% to 75% [11]. Surgical intervention could be either the best solution or only an exploratory laparotomy, if there is only an affected segmental, resection and anastomosis must be performed but if the extension involves a long or multiple segments the surgical management must evaluate the risk-benefit of long-segment intestinal resection. Unfortunately, patients with this clinical presentation often have a poor prognosis.

However, for the most PCI cases are usually managed conservatively. Different therapies in literature reported as: alpha-glucosidase inhibitors; antibiotics like metronidazole to harmful intestinal bacteria and diminish the hydrogen production; microbiota regulators; parenteral nutrition for an intestinal “rest” and some authors have based on trials of hyperbaric oxygen therapy use at 2.5 atmospheres for 150 minutes at least 3 sessions, with limited adverse effects [12], but the sustained effect still unclear.

CONCLUSION

PCI is a rare condition of air cysts in the intestinal wall, with an idiopathic etiology, it must be differential diagnosis in asymptomatic patient with radiologic findings of intraabdominal free gas. Nowadays the long-term treatment is unclear but, fortunately, most of the patients are conservatively managed with microbiota regulators, hyperbaric oxygen therapy and adequate control of their CTD, surgical management must be led by biochemical predictors and clinical presentation rather than compatible radiological images with infradiaphragmatic gas. Due to the low worldwide incidence, for the physician it is imperative either know this pathology to keep it in mind and to educate the patients get acquainted with their diagnosis for further follow up and prevent unnecessary surgeries.

Conflicts of Interests: The researchers have disclosed no conflicts of interest.

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