

A Case of Pneumoperitoneum Secondary to Pneumatosis Intestinalis: Is Surgery Always Indicated?

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Pneumatosis intestinalis (PI) is a rare condition, which could be associated with a spectrum of disorders that vary from mild to life-threatening.

We report a case of an eighty-five-year-old male who presented with abdominal distension and vomiting. Examination revealed the presence of peritoneal signs, with abdominal and pelvic CT confirming PI of the small bowel, a relatively rare condition. The patient underwent an exploratory laparotomy, during which, a segment of the terminal ileum was resected and an ileostomy was fashioned. The patient, unfortunately, died one month postoperatively due to postoperative complications. This case demonstrates the need for risk stratification guidelines to decide conservative or surgical management in cases of PI in this age group.

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PI is the presence of multiple gas cysts in the walls of the small and large intestines¹. It could affect all ages and the estimated prevalence is 0.02% to 0.003% worldwide². It could be primary or secondary¹. The latter is usually associated with higher mortality rates, whereas the former is mostly benign and often considered an incidental finding. Most patients are asymptomatic. The condition is usually discovered either incidentally during a radiological investigation, during surgery or by the patient presenting with symptoms of the underlying disease³. Due to the rarity of this condition, very few cases have been reported in the literature. In Bahrain, there has been no similar case documented.

The management varies from conservative management to emergency surgery. Many reports have been published of spontaneous recovery from pneumatosis intestinalis²⁻¹⁰. Conservative management includes antibiotics, such as metronidazole and hyperbaric oxygen therapy. Urgent surgical exploration should be reserved for complicated cases with alarming symptoms^{2,3,6,8,11}. Without definitive risk stratification criteria, and with limited evidence regarding the management of these cases, surgeons are faced with a dilemma when to decide for emergency surgery³.

The aim of this report is to present a case of an eighty-five-year-old male who presented with primary PI and to review the management options.

THE CASE

An eighty-five-year-old Bahraini male presented with one-week history of gradually increasing abdominal distension. The distension was associated with a three-day history of vomiting. The vomitus was moderate in amount and contained normal food; he had three episodes per day with no blood or bile staining. The symptoms of abdominal pain, dysphagia, nausea and pruritus could not be assessed due to the unresponsiveness of the patient.

The patient was a known case of hypertension, diabetes mellitus type 2 and schizophrenia. He had been confined to bed for some time. He had no past surgical history and no history of previous hospital admissions.

On examination, the patient was conscious; however, he was disoriented and appeared dehydrated. He had no signs of anemia, jaundice or cyanosis. He did not appear cachectic. His vital signs were within the normal range and his cardiovascular and respiratory examinations were unremarkable. On inspection, the abdomen was massively distended, tender and tense. Bowel sounds were absent. Examination per rectum was unremarkable. The patient had a chronic suprapubic catheter inserted for his benign prostatic enlargement.

The hemoglobin level was 10.1 mg/dL. Complete blood count was normal, as well as liver function tests and electrolytes. Urinalysis and chest X-ray were normal. The lateral decubitus view of his abdominal X-ray revealed large amounts of free intraperitoneal air. Abdominal and pelvic CT revealed dilated small bowel loops with air inside the wall indicating PI. In addition, CT revealed a right inguinal hernia containing air with some air loculi escaping into the subcutaneous tissue of the anterior abdominal wall, see figure 1 (A and B).

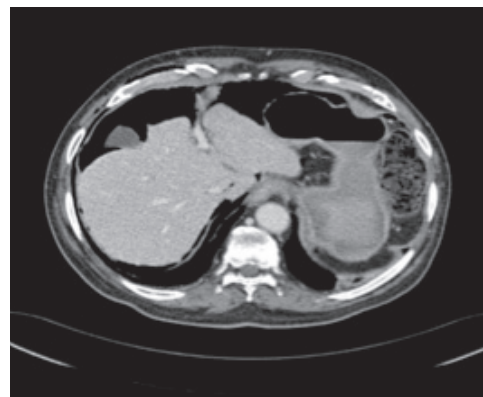


Figure 1A

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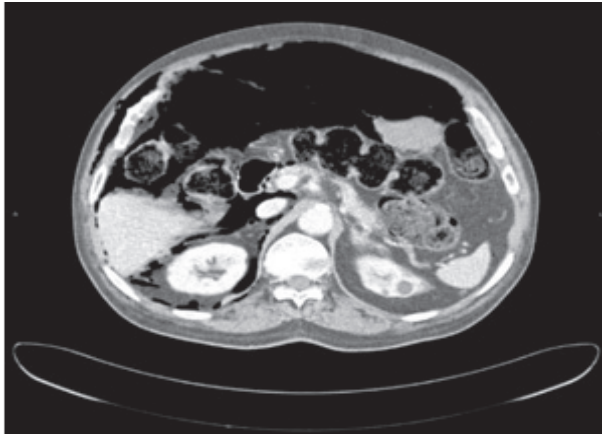


Figure 1B

Figure 1 (A and B): CT Abdomen Revealing Dilated Small Bowel Loops with Pneumatosis Intestinalis and Right Inguinal Hernia

The patient was resuscitated with intravenous fluids and antibiotics (piperacillin-tazobactam and metronidazole). The initial diagnosis was acute abdomen secondary to viscus perforation; therefore, emergency exploratory laparotomy was performed. Intraoperatively, no perforation of bowel or other organs was seen; however, the small bowel loops were dilated and PI was evident in the distal ileum. During the surgery, a segment of approximately 10 to 15 cm was resected from the terminal ileum. An ileostomy with mucous fistula was fashioned. A central line was inserted during surgery to optimize the resuscitation process.

The patient was on ventilatory support for the first two postoperative days and then weaned off. On the 16th postoperative day, the patient had recurrent vomiting with a serum potassium level of 6.8 mmol/l. He was managed according to the standard treatment of hyperkalemia. On the 21st postoperative day, the patient's hemoglobin dropped to 8.7 mg/dL; the ileostomy was closed. Five days after the closure, the patient went into cardiogenic shock; cardiopulmonary resuscitation was administered for 40 minutes; however, the return of spontaneous circulation was not achieved. The patient was declared dead, and the underlying cause was presumed to be a pulmonary embolism.

The histopathological findings of the resected segment of ileum included viable small bowel mucosa with diffuse submucosal, mural and subserosal irregularly sized air spaces. Additionally, there was a foreign body giant cell reaction with mixed inflammatory reaction in the mesenteric fat. Microscopically, flattened macrophages lined the cysts along with multinucleated giant cells.

DISCUSSION

PI is gas cysts within the bowel wall. These air cysts are found in the mucosal and submucosal layers of the bowel wall adjacent to the blood vessels. They are usually on the antimesenteric border of the intestinal wall. Du Veroni had described PI as a disease entity in 1730. Secondary PI is more common than primary PI and is more likely to cause death. Secondary PI could be due to the following: endoscopy, surgical anastomosis,

carcinoma, inflammatory bowel disease, diverticular disease, necrotizing enterocolitis, mixed connective tissue disease, multiple sclerosis, scleroderma, *C. difficile*, HIV, CMV, TB, cytotoxic agents, corticosteroids, COPD, asthma and cystic fibrosis^{8,11}.

The etiology of PI remains unclear; however, multiple hypotheses have been proposed. One mechanism suggests that in the presence of increased intraluminal pressure along with compromised integrity of the mucosa, intraluminal gas dissects along the bowel wall to form cysts. Another theory proposes that gas-producing bacteria gain access to the submucosa through breaks in the mucosa. A third theory suggests alveolar rupture, which leads to alveolar air leaking into the vascular channels of the retroperitoneum, which spread to the mesentery and bowel wall. The content of these cysts are usually hydrogen and nitrogen, and they are most commonly found in the small bowel (42%), large bowel (36%) or both (22%)^{11,12}.

PI could affect all ages; however, it is difficult to determine the incidence or the prevalence, since most of the cases remain asymptomatic and undiagnosed. Symptomatic patients may complain of abdominal pain, vomiting, diarrhea, constipation and weight loss. On examination, there may be abdominal tenderness or generalized distension^{1,2,11}.

Mortality increases in intestinal ischemia, perforation and peritonitis. PI may lead to pneumoperitoneum or ileus formation^{1,2,11}. In particular, pneumoperitoneum is considered diagnostic evidence of a ruptured intra-abdominal viscus. However, in 10% of pneumoperitoneum, the cause is physiological and emergency surgery is not indicated¹³.

Causes of PI: retained air from a laparotomy or laparoscopic procedure, cardiopulmonary resuscitation, adenotonsillectomy, pulmonary tuberculosis, blunt trauma, bronchopulmonary fistula, spontaneous rupture of pulmonary blebs, pneumatosis cystoides intestinalis, endoscopic procedures, postpolypectomy syndrome, peritoneal dialysis, collagen vascular disease, pneumocholecystitis, jejunal and sigmoid, diverticulosis, distended hollow viscus, subclinical perforated viscus, vaginal insufflation, pelvic inflammatory disease, coitus and gynecologic examination procedures¹³.

In cases of ischemia of intestines, there may be acidosis (pH <7.3), hyperamylasemia (>200 IU/L), elevated serum lactate (>2 mmol/L) and/or low serum bicarbonate (<20 mmol/l). Plain X-ray and CT abdomen are the most frequently used to diagnose PI. The presence of linear gas distribution radiologically signifies risk of impending bowel perforation^{11,12}.

The treatment and management of PI, whether conservative or surgical, remains controversial. Khalil et al proposed a checklist for PI, which include critical symptoms, ominous physical examination findings, previous comorbidities, and alternating laboratory or radiological findings that would mandate urgent surgery¹⁴. Another study found that the only statistically significant factors in the differentiation of benign compared to worrisome disease were CT findings of mesenteric stranding, bowel wall thickening and ascites¹⁵. Another study found that the most statistically significant factor for predicting pathological PI is a lactic acid level of more than 2.0; other

significant factors were hypotension or vasopressor need, peritonitis, acute renal failure, need for mechanical ventilation and absent bowel sounds¹⁶.

Retrospectively, in our case, we have come to believe that a more conservative approach in management may have led to better outcome. The decision to perform emergency surgery should be based on high-risk symptomatology, clinical examination and laboratory or radiological findings. Surgical exploration should only be performed when there is suspicion of intestinal obstruction, ischemia, perforation, sepsis, peritonitis or severe lactic acidosis.

The limitations of this case are largely due to its retrospective nature. The information gathered was partially based on subjective observation, which could lead to information bias. It is also difficult to draw comparisons as there was no objective statistical parameter used when evaluating the existing literature on the subject.

CONCLUSION

Urgent laparotomy could be avoided in some cases of PI by using an evidence based criteria for classifying primary compared to secondary. Clinicians must be aware that all necessary investigations must be performed before deciding on the management plan, and that the results must be correlated with the clinical picture of the patient.

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