Case Report

Chronic Spontaneous Pneumoperitoneum with Pneumatosis Cystoides Intestinalis of the Small and Large Intestine

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ABSTRACT: Pneumoperitoneum can be an alarming radiological finding and a manifestation of a surgical emergency that warrant urgent intervention, or it can be a manifestation of chronic benign condition that can be managed conservatively. The sequela of misdiagnosing pneumoperitoneum due to surgical abdomen as a chronic benign pneumoperitoneum can be life-threatening and misdiagnosing chronic spontaneous pneumoperitoneum due to chronic condition as surgical emergency will lead to unnecessary surgical interventions. Diagnosis of chronic spontaneous pneumoperitoneum can be challenging to the unwary healthcare-providers. We present a case of chronic pneumoperitoneum secondary to pneumatosis cystoides intestinalis that has been managed conservatively.

KEYWORDS: Small Bowel Lymphoma, Pneumatosis Intestinalis, Pneumoperitoneum, Spontaneous Pneumoperitoneum and Non-Surgical Pneumoperitoneum.

Introduction

Pneumoperitoneum (PP) is defined as the presence of free air in the peritoneal cavity. Pneumoperitoneum is a radiological diagnosis. It can be detected on an upright chest or plain abdominal radiography or abdominal computed tomography (CT), usually as pockets of free air under the diaphragm. However, pneumoperitoneum sometimes can be missed on non-erect plain radiography. Pneumoperitoneum can be an alarming sign for surgical abdomen, or it can be chronic manifestation of certain medical conditions. Managing PP is usually challenging and can be a diagnostic dilemma that may lead to unnecessary surgeries to the incautious healthcare providers and at the same time, the consequences of misdiagnosing surgical PP as a chronic non-surgical condition can be serious and life threatening [1].

Case presentation

A 77-year-old male with history of small bowel lymphoma in remission for 5 years who presented with complaints of intermittent nonspecific diffuse abdominal pain and bloating for more than 5 years. The patient reported improvement of abdominal pain with deep palpation of his belly or with abdominal massage.

When he develops worsening abdominal pain and bloating, he would take a 7 to 10-day course of oral metronidazole, with significant improvement in his symptoms and he remains asymptomatic for few months and then his symptoms recur again.

Physical examination was devoid of abdominal tenderness or any signs that may suggest peritonitis. The patient remains afebrile with no symptoms of sepsis even when he has abdominal pain.

He always has normal vital signs on examination and normal white blood cell count, basic metabolic panel, liver function tests and lactic acid on routine blood tests on multiple occasions.

Computed tomography of the abdomen and pelvis with contrast revealed a 5cm pocket of free air under the diaphragm and air within the jejunal and cecal walls (Figure 1,2,3) consistent with PP and pneumatosis intestinalis (PI). Small bowel enteroscopy examination was normal except for multiple jejunal diverticula (Figure 4).

Colonoscopy was unremarkable except for non-bleeding pancelonic diverticulosis. Hydrogen breath test for small intestinal bacterial overgrowth (SIBO) was positive.

The patient was initially treated with a course of metronidazole 250mg three times daily for the SIBO and his symptoms improved.
Initially he was requiring intermittent courses of metronidazole 250mg three times a day for 7 days every few months for his intermittent symptoms over the last three years.

However, over the last year he started having more frequent symptoms needing metronidazole 250mg three times a day for the first 7-10 days of every month.

Recently this regimen has still been causing frequent exacerbation of his symptoms and now we are treating him with alternating doses of metronidazole 250mg three times daily for the first 10 days of one month and then doxycycline 100mg twice daily for the first 10 days the next month with acceptable control of his symptoms.

He also follows a low carbohydrate diet as much as possible.

The alternating antibiotics are intended to decrease antibiotic resistance.

The patient gave a written informed consent for this case report to be published.

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**Figure 1.** Axial view of the CT of the abdomen and pelvis with contrast showing pockets of free air in the peritoneal cavity. One of them measuring 5cm (bottom arrows) and other smaller pockets of free air in the peritoneal cavity (upper yellow arrows) consistent with pneumoperitoneum.

**Figure 2.** Axial view of the CT of the abdomen and pelvis with contrast showing cecal wall pneumatosis (yellow arrows).

**Figure 3.** Coronal view of the CT of the abdomen and pelvis with contrast showing pockets of intraperitoneal free air consistent with pneumoperitoneum (yellow arrows).
Discussion

Pneumoperitoneum is divided into spontaneous PP in 10% of the cases and secondary PP in the remaining 90% of the cases.

The latter occurs secondary to inflammatory conditions and/or conditions that need surgical intervention such as perforated viscus, appendicitis, diverticulitis or intrabdominal inflammation.

However, pneumoperitoneum can be iatrogenic such as post operatively following abdominal surgeries including open laparotomy or laparoscopy, or gastrostomy tube placement, or rarely after endoscopic procedures such as mucosal resection or submucosal dissection without a frank perforation.

Spontaneous PP can be seen also in generalized sepsis, pneumatosus intestinalis, peritoneal dialysis, or conditions associated with increased intrathoracic pressure such as pneumomediastinum, recent cardiopulmonary resuscitation, positive pressure ventilation or obstructive lung diseases.

Also, PP was reported to occur in women after sexual intercourse or in pelvic inflammatory diseases [2,3,4].

Pneumatosis intestinalis is a condition characterized by cyst formation in the submucosal and subserosal layers that can affect any segment of the bowel.

Jamart and colleagues showed that the most frequent affected segment is the small bowel followed by large bowel and both, small and large bowels together being affected simultaneously the least [5].

In 85% of cases, PI is secondary to other medical conditions such as bowel obstruction including pseudo-obstruction, sepsis, enterocolitis, bowel ischemia, connective tissue diseases, Crohn’s disease, and alpha-1-glucosidase inhibitor use [5].

Pneumatosis intestinalis is spontaneous in 15% of the time.

The “benign spontaneous” PI is generally pneumatosus cystoides intestinalis (PCI).

Pneumatosis cystoides intestinalis is the most common abdominal cause of spontaneous PP [6,7].

As in our case, the patient has recurrent abdominal discomfort likely from the large cysts in small and large bowel due to his PCI and then he feels clinically better after these cysts rupture. The air from the ruptured cysts then leaks into the peritoneal cavity and trickles under diaphragm leading to pockets of free air in the abdominal cavity and subphrenic area resulting in spontaneous PP.

There are three postulated theories as the underlying mechanism for PCI:

1) Mechanical theory: a significant increase in intraluminal pressure leading to gas
penetration through the mucosa and dissection through bowel wall layers.

This theory is supported by the fact that PI can be seen after colonoscopy or endoscopy without transmural perforation.

Also, it was reported that PI can be reproduced experimentally by performing mucosal incisions in an excised colonic segment followed by air insufflation [8].

2) Bacterial theory: bacteria traverse bowel wall and produce gas in the intramural compartments.

This theory is supported by the clinical response and gas disappearance with antibiotics administration, as it was demonstrated in our case.

3) Pulmonary theory: alveolar rupture results in air in mediastinum, which can pass through pleuroperitoneal openings and track through the peritoneum producing PP and through the bowel wall producing PI [7,9].

In our case, the patient has multiple small bowel diverticula, which likely led to SIBO, as evidenced by the positive breath test.

Therefore, the excessive gas production and increased intraluminal pressure resulted in gas penetration through the mucosa and bacterial translocation through the mucosa to different bowel wall layers producing multiple cysts.

Thus, the mechanical and bacterial theories appear to be the pathophysiological explanation of PCI in our case.

This is supported by the fact that his symptoms resolve with metronidazole treatment.

The clinical manifestations of primary PCI are nonspecific such as abdominal pain, bloating, diarrhea, nausea, and vomiting.

Diagnosis can be made based on radiological findings and/or endoscopic findings.

PCI rarely can lead to complications that may require surgical intervention.

Rarely, it may lead to bowel obstruction due to extremely large cysts that may obstruct the lumen, or it can precipitate intussusception or volvulus.

Cyst rupture sometimes can lead to hematochezia [6].

Chronic asymptomatic spontaneous PCI leading to spontaneous pneumoperitoneum can be managed conservatively by monitoring the patients without interventions.

Chronic symptomatic spontaneous PCI presenting with abdominal discomfort or bloating is usually managed with antibiotics, hyperbaric oxygen, or elemental diet.

The postulated mechanism of action of the aforementioned measures is by altering the gut microbiome.

In rare cases, surgery may be indicated when patients are having persistent significant symptoms from chronic PCI or manifesting with obstructive or ischemic symptoms.

Case series and case reports reported the use of endoscopic sclerotherapy or cyst puncture in PCI-induced bowel obstruction [10,11,12,13].

Worrisome symptoms or signs that may accompany pneumoperitoneum and may warrant surgical emergency are:

1) the presence of abdominal and/or pelvic fluid collection along with PI and PP,

2) symptoms or signs of peritonitis,

3) bowel obstruction,

4) metabolic acidosis and/or lactic acidosis due to the possibility of necrotic bowel or underlying infection,

5) hemodynamic instability,

6) portal venous gas [14].

Conclusion

Patients presenting with PCI with or without spontaneous PP and without worrisome features can be managed conservatively without surgical intervention.

However, each patient presenting with spontaneous PP should be evaluated on a case-by-case basis prior to deciding to proceed with conservative management, taking into consideration the clinical setting, patient presentation, risk factors, physical examination, laboratory results and imaging findings.

The consequences of misdiagnosing surgical PI or PP as a benign nonsurgical condition can be dangerous and life threatening.

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Conflict of interests

None to all authors.

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