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Pneumatosis Cystoides Intestinalis: Uncommon Cause of Emergency Laparoscopic Surgery

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Abstract

Introducton: Pneumatosis cystoides intestinalis is a rare entity of unclear etiology. It is defined as the presence of gas filled cysts within the wall of the gastrointestinal tract. Pneumatosis cystoides is found accidentally during endoscopy or radiographic examination. Asymptomatic pneumoperitoneum can pose a differential diagnostic challenge especially versus perforation of hollow viscus. This report describes a case of pneumatosis cystoides intestinalis discovered by a pneumoperitoneum.

Case presentation: A 57-year-old man admitted to the emergency department for acute onset of abdominal pain. On examination, we found mild to moderately tender over the epigastric and umbilical regions without any presence of rigidity or rebound tenderness. Laboratory tests revealed leukocytosis and high C reactive protein level. A plain abdominal radiograph showed the presence of pneumoperitoneum. The diagnosis of perforation of an intraperitoneal hollow organ was suspected. The patient underwent emergency laparoscopic surgery. Intraoperative exploration found the presence of several gas filled cysts within the bowel wall. No additional surgical procedure was performed.

Conclusion: Pneumatosis cystoides intestinalis is often an incidental finding during endoscopy or X-ray imaging. Rupture of subserosal pseudocysts can lead, in about 30% of cases to pneumoperitoneum without signs of peritonitis. So, the surgical decision should not be taken only on the radiologic image but should be coupled to the clinical symptoms in order to avoid unnecessary surgery.

Keywords

Pneumatosis cystoides intestinalis; Leukocytosis; Abdominal pain

Introduction

Pneumatosis cystoides intestinalis (PCI) also known as “gas cysts of the intestine” is a rare condition. It is defined as the presence of gas filled cysts within the wall of the gastrointestinal tract. The incidence is reportedly 0.3% based on computed tomography results [1]. Pneumatosis cystoides intestinalis is classified as idiopathic or primary (15%), when no clear underlying cause is founding, and secondary (85%) associated with numerous of diseases [2,3]. Often, it is asymptomatic or pauci symptomatic, and discovered during a radiological or endoscopic examination. Rarely, the diagnosis is established following acute abdominal pain. This article describes a case of primary PCI presenting as acute abdominal pain simulating perforation of a hollow organ.

Case Presentation

A 57-year-old man who had no medical or surgical history admitted to the emergency department for acute onset of abdominal pain. On examination, the patient was afebrile and had a heart rate of 87 beats/min, a respiratory rate of 13 breaths/min, blood pressure of 135/87 mmHg. The abdomen was non-distended, we found mild to moderately tender over the epigastric and umbilical regions without any presence of rigidity or rebound tenderness. Bowel sounds were present in all four quadrants. Laboratory tests revealed inflammation (white blood cell count 11800/mm^2; C-reactive protein 45 mg/dL). The rest of the biological assessment was without abnormalities including lipase. A plain abdominal radiograph showed the presence of pneumoperitoneum (Figure 1). In the presence of acute abdominal pain, inflammatory syndrome and pneumoperitoneum, the diagnosis of perforation of an intraperitoneal hollow organ was suspected. The patient underwent emergency laparoscopic surgery. Intraoperative exploration found the presence of several gas filled cysts within the bowel wall (Figures 2 and 3). There was no effusion or digestive perforation. The diagnosis of pneumatosis cystoides intestinalis was retained. No additional surgical procedure was performed. The postoperative course was uneventful and proved to be without any complications.

Discussion

Pneumatosis cystoides intestinalis is not a disease but a rare phenomenon that affects the gastrointestinal tract. It is characterised by the presence of gas-filled pseudocysts in the submucosa or subserosa of the gastrointestinal tube. Any part of the gastrointestinal tract can be involved. Most frequently, PCI is found in the small intestine, followed by the colon [4]. The extension of these gas filled cysts may be mesenteric, retroperitoneal and may cause pneumoperitoneum or retro pneumoperitoneum. In our case, only the small bowel was involved. PCI can appear at any age, with peak occurrence between the fifth and eight decade, its incidence is reportedly 0.3% based on computed tomography results [1]. We can distinguish two groups of PCI: primary and secondary. The primary or idiopathic type is not associated with other coexisting diseases, accounts for about 15% of cases, and does not need any therapy in general. The secondary type comprises 85% of cases and accompanies numerous diseases [5]. The etiopathology of PCI is multifactorial. Several theories exist to explain the pathogenesis of PCI [6]. In the mechanical theory, predisposing factors such as bowel obstruction, gastroenteric tumor, colonoscopy may increase intra-luminal pressure that causing mechanical injury to the intestinal wall

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and a break in the mucosa, which allows gas migration. In chronic pulmonary disease, there is direct diffusion of air from ruptured alveoli through the mediastinum, retroperitoneum and mesenterium [7]. The biochemical theory suggests a disorder between excessive hydrogen produced by bacteria and its consumption in the lumen of the large bowel leads to high luminal pressure. It then rapidly diffuses into the wall, forming pseudocysts. Finally, PCI is also associated with connective tissue disease, hormonal therapy and chemotherapy [7]. In our case we could not identify any specific predisposing factor, so we considered the patient as developing primary PCI. Clinically, the majority of patients are asymptomatic or pauci-symptomatic and pneumatosis cystoides is found accidentally during endoscopy or radiographic studies. Moreover, it can be revealed by non-specific signs. The most common symptoms are abdominal pain and diarrhea (53%), followed by abdominal distention, vomiting, bloody stool, and constipation. Nearly 16% of patients have complications such as intestinal obstruction and perforation [7]. Physical exam is often poor; occasionally we can palpate abdominal masses in the case of big cysts. The diagnosis is essentially based on the complementary exams. A plain abdominal radiograph can show localized gas collection, pneumoperitoneum or retropneumoperitoneum. Asymptomatic pneumoperitoneum complicating PCI results from the rupture of the cyst and can pose a differential diagnostic challenge especially versus perforation of hollow viscous and leads to unnecessary surgery as the case of our patient. So, the surgical decision should not be taken only on the radiologic picture of pneumoperitoneum, but should be coupled to the clinical symptoms. On endoscopy, sessile polyps are seen isolated or clustered, and covered with normal mucosa. The removal of polyps is contraindicated because of the risk of perforation. Abdominal computerized tomography is the most sensitive method for detection of PCI and it has an additional value in detection of complications or underlying disease. The treatment is discussed according to the clinical presentation. The majority of patients have no complaints and PCI can regress spontaneously [8], no treatment is necessary. Some forms may have a chronic evolution with periods of remission [9]. If the symptoms persist, a conservative approach of therapy is allowed. It combines oral antibiotic therapy against anaerobes (metronidazole is most often prescribed), hyperbaric oxygen therapy and a diet without residues low in carbohydrates [10]. The modalities and duration of this treatment are still discussed. Endoscopic treatment (polypectomy) should generally be avoided. In the case of the secondary form, the underlying disease should be treated first. Surgical therapy is indicated in case of complications such as intestinal obstruction or perforation. It consists of resecting the pathological intestinal segment. Laparoscopic surgery is preferred.

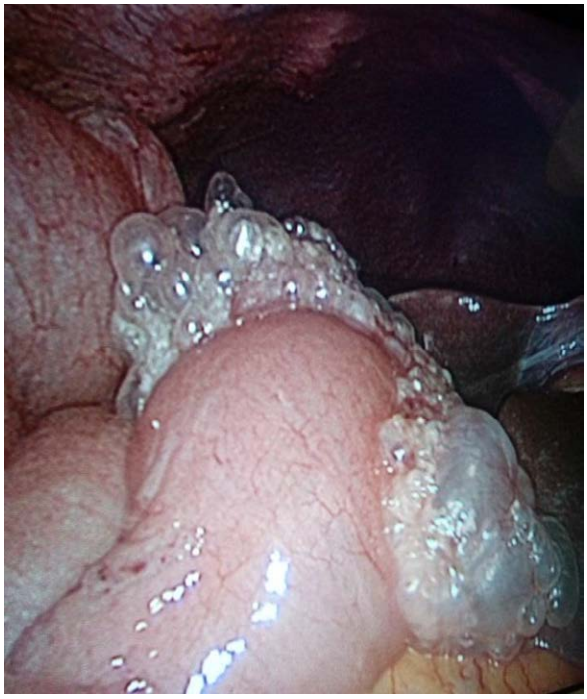

Figure 1: A plain abdominal radiograph showed the presence of pneumoperitoneum.

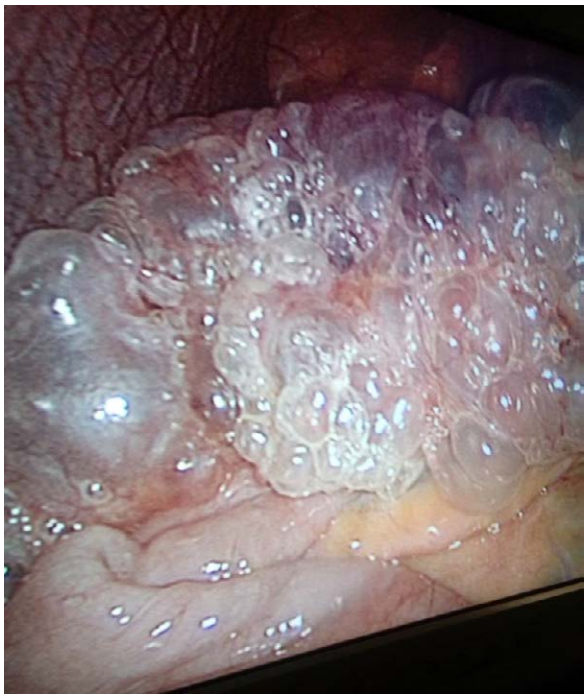

Figure 2: Intraoperative exploration found the presence of several gas filled cysts within the bowel wall.

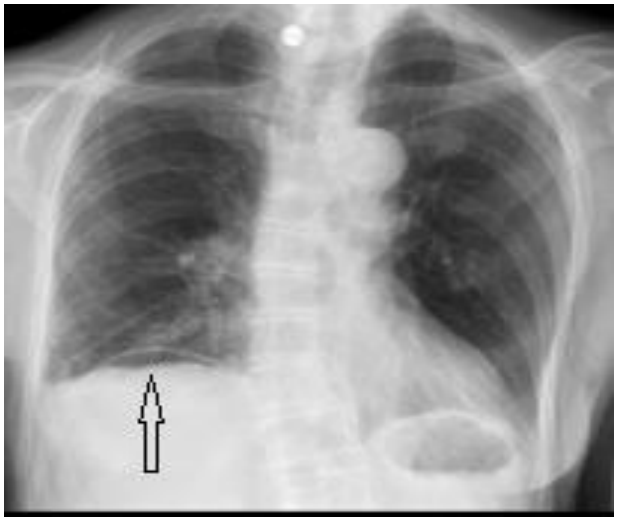

Figure 3: Pneumoperitoneum.

Conclusion

Pneumatosis cystoides intestinalis is a rare entity of unclear etiology. It is often an incidental finding during endoscopy or X-ray imaging. Rupture of subserosal pseudocysts can lead, in about 30% of cases to pneumoperitoneum without signs of peritonitis. So, the surgical decision should not be taken only on the radiologic image but should be coupled to the clinical symptoms in order to avoid

unnecessary surgery. Primary pneumatosis cystoides intestinalis is often asymptomatic and it may regress spontaneously. In case of persistent symptoms, a conservative approach of therapy is allowed.

Conflict of Interest

The authors report no conflict of interest.

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