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# Pneumatosis cystoides intestinalis mimicking acute abdomen

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## ABSTRACT

Pneumatosis cystoides intestinalis (PCI) is a rare disease. It was first described by Du Vernoy in 1793 during a cadaver dissection. Air-filled bubble-like lesions are located in the submucosa or the subserosa of the digestive tract. A 55-year-old Turkish male presented to the emergency department with complaints of recurrent abdominal pain and vomiting. Free air was detected in abdominal x-ray and abdominal computed tomography (CT). In exploration, a grape-like lesion consisting of hundreds of cysts was detected on the surface of the small intestine. Bowel resection was performed successfully. The surgical findings and pathological result confirmed the diagnosis of pneumatosis cystoides intestinalis.

**Keywords:** Pneumatosis cystoides intestinalis, abdominal pain, acute abdomen

Case Report

## INTRODUCTION

Pneumatosis cystoides intestinalis (PCI) is a rare clinical pathology characterized by submucosal and/or subserosal collections of free gas, forming cystic lesions within the gastrointestinal tract and related peritoneal attachments (1). Air-filled bubble-like lesions are located in the submucosa or the subserosa of the digestive tract and usually range from 0.5 to 2.0 centimeters in size. Although PCI is most commonly seen in the intestine, it can be detected in all parts of the gastrointestinal tract. The incidence of PCI was reported to be 0.03% in the general population (2). The diagnosis of PCI can be a challenging event. We are presenting a patient who was admitted to the emergency department with abdominal pain for 3 days. He was operated on with a preoperative diagnosis of hollow viscous perforation. The surgical findings and pathological results confirmed the diagnosis of PCI.

## CASE PRESENTATION

A 55-year-old Turkish male presented to our emergency department because of a 7-day history of recurrent abdominal pain and vomiting. His past medical history included 2 laparotomies due to gastric and colonic perforation. He also had a right inguinal hernia that was reduced 3 days ago. On the physical exam, the patient had moderate abdominal distension, tenderness, and suspicious rebound. Laboratory studies demonstrated white blood cell count: 5500 K/UL (normal: 4.8-10.8 K/UL), hemoglobin: 12.7 gr/dL (normal: 12-18 gr/dL), CRP 0.2 mg/dL (normal: 0-0.05 mg/dL), AST: 20 U/L (normal: 10-42 U/L), and ALT: 16 U/L (normal: 10-40 U/L). There was free air under the right diaphragm in the abdominal x-ray (Figure 1). There was free fluid in the right lower quadrant of the abdomen in the abdominal ultrasonography (USG). Computed tomography (CT) of the abdomen and pelvis revealed free air and fluid in the abdominal cavity (Figure 2). Diagnostic laparoscopy was performed. In the exploration, there were free intra-abdominal bubbles in all quadrants. A grape-like lesion consisting of hundreds of cysts was detected on the surface of the small intestine (Figure 3). Free intra-abdominal fluid was found in Douglas pouch. There was partial small intestinal obstruction due to the cystic lesion. Laparotomy was performed. An intestinal segment about 150 cm in length was resected with the cystic lesions. Jejunoileal anastomosis was done. The pathological result of the resected intestinal segment was reported as pneumatosis intestinalis (Figure 4). The postoperative period of patient was uneventful.

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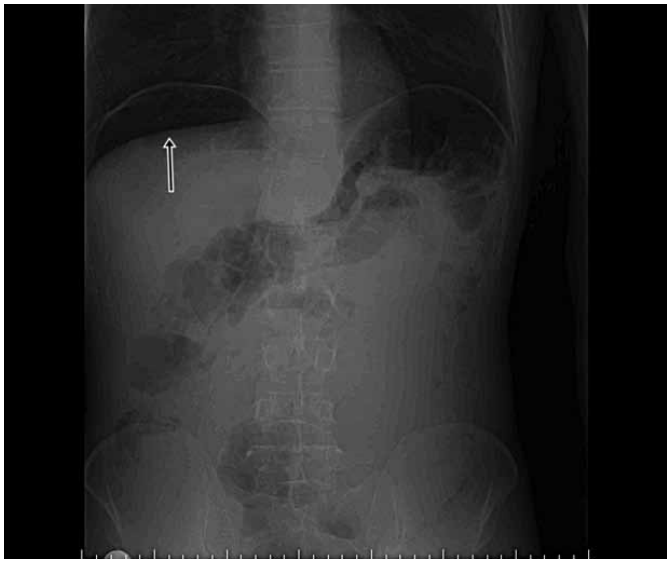

**Figure 1.** Free air under right diaphgram (black arrow) in chest X-ray.

**Figure 2.** Abdominal CT of the patient showing dilated gastric and intestinal segments.

The informed consent was taken from patient for this case presentation.

## DISCUSSION

Pneumatosis cystoides intestinalis is an uncommon disease characterized by gas-filled cysts within the bowel wall, especially in the jejunum, ileum, and colon (3). Its etiology is not fully understood. It was first described by Du Vernoy in 1793 during a cadaver dissection (4). Jamart, in his analysis of 919 patients, found a peak incidence between 41 and 50 years. The man-to-woman ratio was 3:1 (5).

Pneumatosis cystoides intestinalis is classified as primary and secondary PCI. Primary PCI is caused by an autonomous disease. Secondary PCI is more frequent and is associated with a wide variety of underlying conditions, including traumatic and mechanical, inflammatory and autoimmune diseases, infectious and pulmonary diseases, drug-induced, immunosuppression, transplantation, or neoplasm (4,6). Patients with PCI may be asymptomatic or can present with bowel necrosis, perforation, and generalized peritonitis. Development of pneumoperitoneum secondary to rupture of the cysts in the absence of any findings related to peritoneal irritation is a pathognomonic feature of the disease.

**Figure 3.** Free bubble like lesions on the intestinal serosa in laparoscopy.

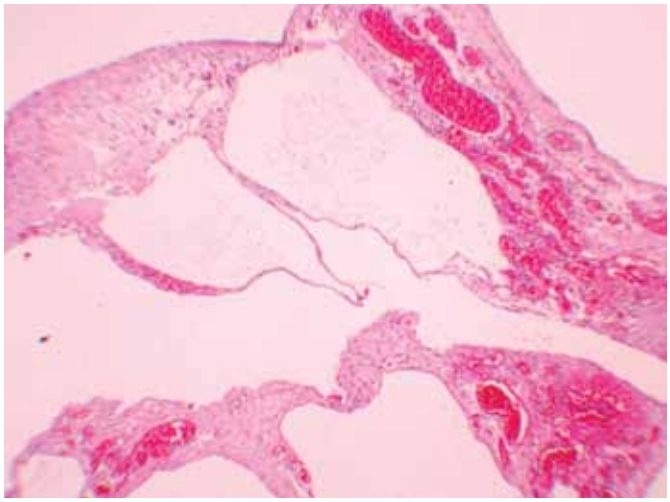

**Figure 4.** Pathological appreance of resected intestinal segment.

Definitive diagnosis of PCI can be difficult. Imaging modalities have importance in the correct diagnosis. Abdominal plain x-ray, USG, CT, colonoscopy, and double-contrast colon radiography may beneficial in the diagnosis. Subdiaphragmatic free gas collection can be detected in abdominal plain films. Bright echoes with acoustic shadowing, representing gas in the bowel wall, can be seen in the USG. CT is the best diagnostic modality, with better sensitivity than plain films or ultrasound (7).

Presence of free gas in the bowel and the mesentery is characteristic of PCI. There are still controversies in the treatment of PCI. Spontaneous resolution of PCI has been reported (6). Conservative treatment, including antibiotics, such as metronidazole in high concentrations, can be effective in the treatment of PCI (7,8). Hyperbaric oxygen was used as a treatment option in PCI (9,10). The decision of surgery is a critical issue in PCI. Free air in the abdominal x-ray or CT is not an absolute indication. Signs of peritoneal irritation or persistent bowel obstruction are the most common reasons for exploratory laparotomy in PCI.

In conclusion, PCI is a rare disease with different clinical presentations. Intra-abdominal cystic lesions with free abdominal air can be detected in radiological examinations. Conservative measures can be successful in some patients. Bowel necrosis, perforation, or persistent obstruction should be treated with surgery.

**Ethics Committee Approval:** N/A.

**Informed Consent:** Written informed consent was obtained from patient who participated in this case.

**Peer-review:** Externally peer-reviewed.

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