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Case Report / Olgu Sunumu

Pneumatosis Cystoides Intestinalis; A Rare Cause of Ileus Mimicking Intestinal Perforation

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Abstract

Pneumatosis Cystoides Intestinalis (PCI) is a rare condition that many physicians find challenging to diagnose. PCI is characterized by the presence of gas-filled cysts in the submucosa and serosa of the gastrointestinal system. It can develop due to trauma caused by conditions such as pyloric stenosis, peptic ulcers, ileus and endoscopic procedures. The exact etiology of PCI remains unclear. Treatment is directed towards the underlying cause. Surgical intervention should be considered only after the etiology is clarified or when acute surgical pathologies are detected.

In this article, we present a 45-year-old male with a history of peptic ulcer and intermittent abdominal pain for the past 10 years. The patient was referred from an external center with a preliminary diagnosis of hollow organ perforation. During the diagnostic process, PCI and perforation were considered in the differential diagnosis. The patient developed ileus and an acute abdomen during follow-up, which necessitated surgical intervention.

Keywords: Pneumatosis Cystoides Intestinalis, Ileus, Intestinal Perforation

Pnömatozis Sistoides İntestinalis; İntestinal Perforasyonu Taklit Eden Nadir Bir İleus Nedeni

Öz

Pnömatozis sistoides intestinalis (PSİ), birçok hekimin tanı koymakta zorlandığı insidansı düşük bir hastalıktır. Pnömatozis sistoides intestinalis, gastrointestinal sistem submukozasında ve serozasında gazla dolu kistlerin görülmesi olarak tanımlanır. Pilor stenozu, peptik ülser, ileus ve endoskopik işlemlerle ortaya çıkan travmaya bağlı olarak gelişebilmektedir. Etiyoloji tam olarak bilinmemektedir. Tedavi altta yatan etiyolojiye yönelik yapılır. Tedavide cerrahi müdahale, etyoloji aydınlatıldıktan sonra gereklilik halinde veya akut cerrahi patolojiler tespit edildiğinde yapılmalıdır.

Bu yazımızda peptik ülser öyküsü olan ve 10 yıldır ara ara tekrarlayan karın ağrısı olan 45 yaşında erkek hasta sunuldu. Hasta dış merkezden lümenli organ perforasyonu ön tanısı nedeniyle interne edildi. Yapılan tetkiklerde ayırıcı tanıda pnömatozis sistoides intestinalis (PSİ) ve perforasyon düşünüldü. Takiplerinde ileus hali olan ve akut batın tablosu gelişen hastaya cerrahi müdahale edildi.

Anahtar kelimeler: Pnömatozis Sistoides İntestinalis, İleus, İntestinal Perforasyon.

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INTRODUCTION

Pneumatosis cystoides intestinalis (PCI) was first described by Du Vernoi in 1730[1]. PCI is a rare disease characterized by the presence of air-filled cysts in the submucosa and/or subserosa of the gastrointestinal wall, which can affect the entire gastrointestinal system[2]. PCI is mostly a benign condition that is often asymptomatic. It is more commonly seen between the fifth and eighth decades of life and is more prevalent in males[1,3]. The incidence has been reported as 0.03%[4]. It can be idiopathic or more often secondary to various diseases. Diagnosis can be challenging both clinically and radiologically. Computed tomography, ultrasonography, endoscopic imaging methods, and direct abdominal radiographs can aid in diagnosis. The condition responds well to medical treatment. However, surgical treatment should be considered if there are signs of ischemia, perforation, or obstruction in the gastrointestinal system[2].

CASE REPORT

A 45-year-old male with no comorbidities, medication use, or surgical history has been experiencing recurrent abdominal pain, vomiting, and dyspeptic symptoms for 10 years and was diagnosed with gastric ulcer 7 years ago.

The patient reported that his abdominal pain had intensified over the past three days, with increased nausea and vomiting. He had gas and stool discharge. Rectal examination revealed no stool contamination. On abdominal examination, tenderness was present in the epigastric region, with no defense or rebound. There was abdominal distension. Laboratory parameters showed WBC: 7.9 and CRP: 101 mg/L. A direct radiograph showed free air under the diaphragm and air-fluid levels in the small intestine (Figure 1). Abdominal CT revealed air-filled cysts in the submucosa and subserosa of the small intestine, resembling a cluster of grapes (Figure 2).

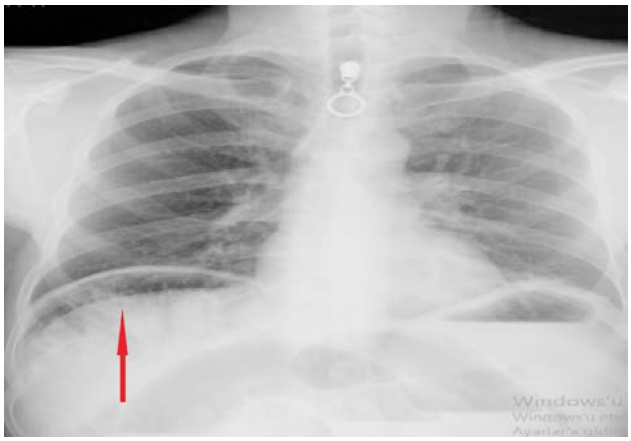

Figure 1. Pneumoperitoneum on Abdominal X-ray (Red arrow: Sub-diaphragmatic free air)

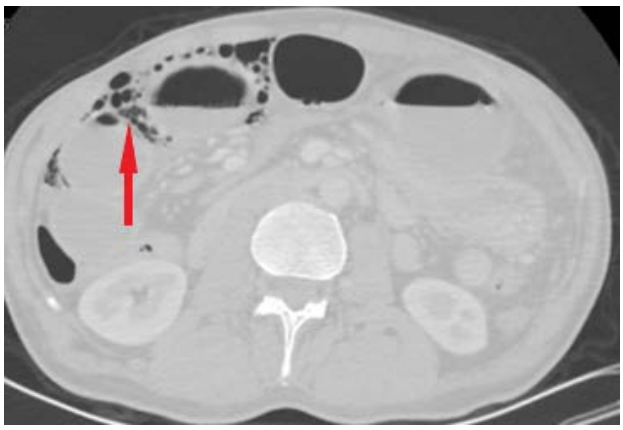

Figure 2. PCI Abdomen BT Image (Red arrow: PCI)

The patient was clinically monitored with a preliminary diagnosis of closed lumen organ perforation and PCI. Oral intake was stopped, and appropriate hydration and antibiotic therapy were administered. Endoscopy revealed edema in the gastric mucosa, with blunted folds and increased vascularity. An ulcer deforming the pylorus and minimally extending into the duodenal area was observed. Four-quadrant biopsies were taken from the lesion, and additional sampling was done from the corpus. The postbulbar lumen was dilated, and the papilla was observed to be normal.

Due to increasing abdominal distension and signs of acute abdomen on examination, an

emergency surgery was decided on the third day of medical follow-up. Intraoperative exploration revealed that the bowel loops were excessively dilated and the pyloric region of the stomach was inflamed. A narrowing that allowed passage through the pylorus and bulb was detected, but no intervention was performed. In the small intestine, there were extensive subserosal air bubbles starting 300 cm from the Treitz ligament and continuing to 20 cm proximal to the ileocecal valve (Figure 3). The small bowel loops were observed to form a "gato" appearance. No perforation area was seen. PCI was present. The patient, who had no history of abdominal surgery, had brids extending from the falciform ligament to the small bowel mesentery, thought to be secondary to chronic inflammation. These brids were dissected using sharp and blunt dissection. Approximately 150 cm of the small intestine, starting 300 cm from the Treitz ligament and extending to 20 cm proximal to the ileocecal valve, was resected (Figure 4). A double-barrel ileostomy was created. A nasogastric tube was placed in the patient and advanced to the fourth part of the duodenum. Retrograde decompression was performed. A Jackson Pratt drain was placed to visualize the ileostomy site and the rectovesical area.

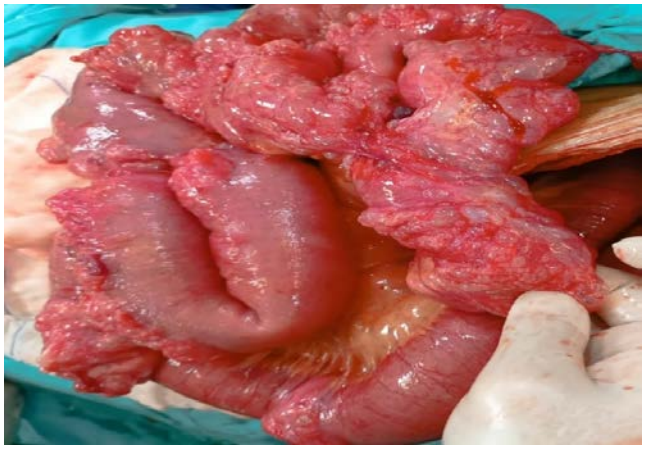

Figure 3. PCI Intraoperative Image

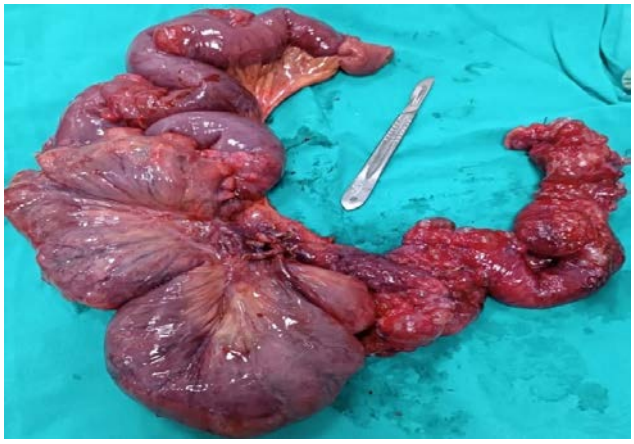

Figure 4. Specimen Image

On the first postoperative day, the drain had a serous appearance, and the ostomy was functioning. Vital signs were stable, there was no abdominal pain, and the abdominal examination was normal. The drain was removed on the 5th postoperative day, and the patient was discharged on the 7th day with recommendations.

DISCUSSION

PCI is divided into two groups: primary (idiopathic) and secondary[2]. Primary pneumatosis occurs in 15% of cases and often involves the colon. The secondary type is associated with conditions such as digestive system stenosis, obstructive lung disease, pyloric stenosis, peptic ulcer, gynecological cancers, amyloidosis, systemic lupus erythematosus, gastrointestinal vasculitis, Sjögren's syndrome, mixed connective tissue disease, abdominal trauma, medication side effects, surgery, and malnutrition, and it frequently involves the small intestine[1,6].

There are five hypotheses regarding the pathogenesis of PCI[1,6-8]:

1. Mechanical theory: If there is an increase in pressure within the intestinal lumen for any reason, the gas within the lumen diffuses into the intestinal wall due to damage to the intestinal mucosa. It can also develop as a result of gastrointestinal wall damage occurring after

gastrointestinal ulcers and perforations or endoscopic procedures and surgical operations.

2. Pulmonary theory: In patients with chronic lung disease that may cause alveolar rupture, an emphysematous condition first occurs in the mediastinum, and later gas is formed in the intestinal wall via the aorta and mesenteric vessels.

3. Bacterial theory: Bacteria colonizing the intestinal mucosa cause gas formation in the intestinal wall through fermentation.

4. Nutritional and Chemical Theory: Due to insufficient nutrition, bacterial fermentation increases, and the resulting intense gas causes submucosal damage by leading to distension and ischemia in the intestinal wall. Recently, PCI has been reported to develop during treatment with α-glucosidase inhibitors (α-GI), oral antidiabetic drugs.

5. PCI related to chemotherapy, hormone therapy, and connective tissue disease.

PCI lesions are mainly found in the colon (46%), small intestine (27%), and stomach (5%)[7,9]. PCI typically presents with non-specific symptoms such as abdominal pain, abdominal distension, diarrhea, nausea-vomiting, hematochezia, and weight loss.

PCI is generally diagnosed using endoscopy, endoscopic ultrasonography (EUS), direct radiography, and abdominal CT[6]. The appearance on abdominal CT is submucosal/subserosal cystic areas resembling a cluster of grapes[10,11]. In PCI, portal venous gas values can also be observed; this association strongly suggests bowel necrosis due to thrombosis of the superior mesenteric artery or non-obstructive mesenteric ischemia[8,11]. Lassandro et al. found portal venous gas in approximately 25.5% of PCI patients in their study and noted that the incidence of bowel obstruction and mortality was increased in these patients[11].

PCI is often incidentally detected on abdominal CT performed for another reason, and these patients are usually asymptomatic. Sometimes complications such as bowel obstruction, perforation, intussusception, or volvulus may be seen due to PCI[8,11]. Endoscopically, grape-like or cystic circular forms, and linear gas-containing cysts can be detected. It should be differentiated from polyps. Subepithelial cystic structures should be distinguished from abdominal tuberculosis. On endoscopy, irregular shapes must be distinguished from tumors and Crohn's disease. In EUS, gas findings in the mucosal layer beneath the cyst can guide the diagnosis of PCI[7,12].

Surgical intervention is not absolutely necessary in the treatment of PCI. The decision for surgical intervention should be considered if there are obstructive pathologies, perforation, or clinical and laboratory deterioration. In the absence of acute surgical pathologies, a conservative approach is the preferred treatment method: Bowel rest, appropriate hydration, antibiotics, and hyperbaric oxygen therapy can be applied. Hyperbaric oxygen therapy is an effective treatment option if there are no signs of bowel wall damage[3-5].

CONCLUSION

PCI is generally asymptomatic. In 85% of patients with asymptomatic PCI, treatment is not necessary. It has been reported that surgery is not required in patients with pneumoperitoneum due to the rupture of subserosal air sacs, and they can be treated medically. Medical treatment should be the first-line approach. Acute surgical pathologies (perforation, abdominal sepsis, peritonitis, gastrointestinal ischemia) must be ruled out. In cases where peritonitis signs and ileus are present, as in our patient, surgical intervention should be considered. A comprehensive evaluation of the clinical history of the patient with PCI and a proper understanding of the

differential diagnosis of PCI are essential to avoid unnecessary surgery.

Patient Consent: Consent was obtained from the patient for the use of patient data, radiological and intraoperative images.

Declaration of ConflictingInterests: No conflict of interest was declared by the authors.

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