



# A Rare Cause of Acute Abdomen: Pneumatosis Cystoides Intestinalis

*Nadir Bir Akut Karın Nedeni: Pnömatozis Sistoides İntestinalis*

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## ABSTRACT

*Pneumatosis cystoides intestinalis (PCI) is a rare disease defined as the presence of gas in the small intestine or colon wall. A 55-year-old female patient was admitted to the emergency service with a complaint of abdominal pain lasting for two days. PCI and intra-peritoneal free air were seen on the abdominopelvic computed tomography scan. The patient underwent an emergency laparotomy. There was no perforation and ischemic bowel segment on exploration. Air bubbles were seen only in the wall of the small bowel loops. No surgical procedure was considered for the patient. The patient was discharged on the seventh day without complication.*

**Key words:** acute abdomen; pneumatosis cystoides intestinalis; laparotomy

## ÖZET

*Pnömatozis sistoides intestinalis (PSİ), ince bağırsakta veya kolon duvarında gaz varlığı olarak tanımlanan nadir bir hastalıktır. 55 yaşında bir kadın hasta iki gündür süren karın ağrısı şikâyeti ile acil servise başvurdu. Abdominopelvik bilgisayarlı tomografi taramasında, PSİ ve periton içi serbest hava görüldü. Hastaya acil laparotomi yapıldı. Eksplorasyonda perforasyon ve iskemik barsak segmenti yoktu. Sadece ince bağırsak kıvrımlarının duvarında hava kabarcıkları görüldü. Hastaya herhangi bir cerrahi işlem düşünülmemedi. Hasta yedinci günde komplikasyonsuz olarak taburcu edildi.*

*Anahtar kelimeler: akut abdomen; pnömatozis sistoides intestinalis; laparotomi*

## Introduction

Pneumatosis cystoides intestinalis (PCI) is a rare disease defined as the presence of gas in the small intestine or colon wall. PCI may also develop spontaneously, although the incidence increases due to pulmonary diseases, systemic diseases, and intestinal diseases. In studies, the prevalence of incidentally detected PCI cases in patients who underwent computed tomography (CT)

was reported as 0.37%<sup>1</sup>. It was first described by the pathologist DuVernoi in 1730<sup>2</sup>. The etiology of PCI has not been fully elucidated. The disease, classified in two forms as idiopathic (15%) and secondary (85%), may present different clinical findings. In the idiopathic or primary form, air cysts are located in the mucosa and submucosa. In addition, no obvious etiological factor was found in the idiopathic form. Air cysts are mostly located in the intestinal wall in the secondary form. In the etiology of secondary form, pulmonary diseases, systemic diseases, intestinal diseases, drugs, and iatrogenic causes can play a role.

The asymptomatic disease can be seen during diagnosis, while life-threatening severe clinical pictures may also occur. If PCI is detected early, it can be treated with a conservative approach. In advanced cases, surgical intervention may be required.

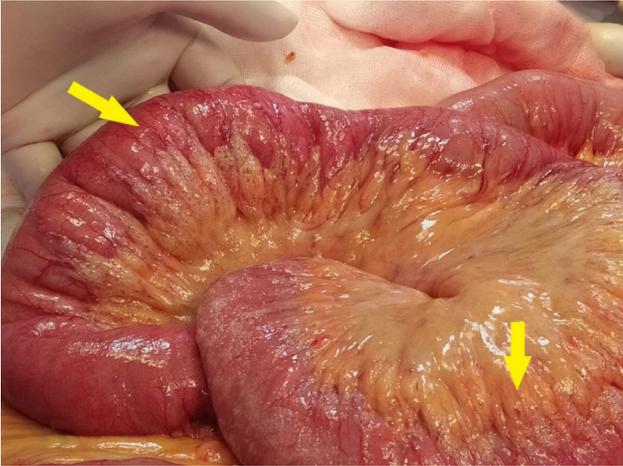
This case report presents a case of PCI who underwent diagnostic laparotomy with intra-abdominal free air on computed tomography.

## Case Report

A 55-year-old female patient applied to the emergency department of an external center with abdominal pain and nausea with vomiting for two days. Upon the presence of right subdiaphragmatic free air in the standing direct abdominal radiograph, the patient was referred to our hospital for further examination. The patient had no history of previous surgery and no comorbid disease. There had never been a similar episode of abdominal pain before.

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**Figure 1.** Intraoperative image of pneumatosis cystoides intestinalis.



**Figure 2.** Air bubbles in the wall of the small intestine.

At the time of admission to the hospital, the patient's arterial blood pressure was 132/86 mmHg, oxygen saturation on room air was 90%, pulse rate was 98 beats/min, and body temperature was 37.6°C. In the abdominal examination of the patient, there was widespread tenderness and defense in all abdominal quadrants, but there was no rebound. Other system examinations were normal. There was normal stool contamination on rectal touch. Laboratory examination revealed leukocytosis (white blood cell count=13200  $\mu$ l/ml), elevated C-reactive protein (51 mg/L), aspartate aminotransferase elevation (78 IU/L), and alanine aminotransferase elevation (63 IU/L). Other parameters were unremarkable.

In the standing direct abdominal X-ray, free air was observed in the right subdiaphragmatic area. Intra-abdominal organ perforation clinic was considered in the patient, and abdominal computed tomography (CT) with contrast was performed to evaluate the perforation localization. There was intra-abdominal free air and PCI on a CT scan at the small intestine segments. The patient underwent a diagnostic laparotomy. At laparotomy, diffuse air bubbles were observed in the small intestine wall from the jejunum to the distal ileum (Fig. 1 and 2). No gastrointestinal organ perforation was observed. No ischemic bowel loop was observed. No additional intra-abdominal pathology was detected during the exploration, and the surgery was terminated by placing a drainage catheter in the pouch of Douglas.

The patient was started on ciprofloxacin 400 mg/200 ml (intravenously every 12 hours) and metronidazole

500 mg/100 ml (intravenously every 8 hours) in line with the postoperative period the recommendation of the infection clinic. Oral feeding of the patient, whose complaints regressed in the postoperative period, was opened on the 2nd postoperative day. The drain of the patient who tolerated oral feeding was removed on the 5th postoperative day, the patient's current antibiotic therapy was completed for seven days, and the patient was discharged on the 7th postoperative day.

## Discussion

More than 90% of pneumoperitoneum occurs as a result of gastrointestinal perforations. Although gastric or duodenal perforation due to peptic ulcer is considered the most common cause of pneumoperitoneum<sup>3</sup>, pneumatosis cystoides intestinalis (PCI) is also a rare cause of pneumoperitoneum.

PCI is defined as the presence of air in the small intestine or colon wall. The diagnosis of PCI can be made preoperatively through radiological imaging methods or intraoperatively. In the etiology of PCI, pulmonary diseases (such as pulmonary fibrosis, asthma), systemic diseases (such as systemic lupus erythematosus, scleroderma), intestinal diseases (such as diverticulitis, intestinal obstruction, enteritis), drugs (such as lactulose, corticosteroids, sorbitol, chemotherapeutic agents), and iatrogenic causes (postoperative anastomosis, barium trauma, endoscopy, jejunostomy tube) can play a role. On the other hand, PCI can rarely be seen spontaneously in rare cases<sup>4</sup>. In the present case, spontaneous PCI was considered because the patient had no known systemic disease, drug use, or history of surgery.

Although there may be symptoms such as vomiting, nausea, abdominal pain, diarrhea, and abdominal bloating before hospital admission, most cases are asymptomatic. Sometimes, life-threatening conditions such as toxic megacolon, intestinal ischemia, or intestinal obstruction may occur<sup>5</sup>. The presented patient had nausea with vomiting and severe abdominal pain that lasted for about two days without relief.

In the radiological diagnosis of PCI, an appearance suggestive of intraperitoneal free air is common. PCI diagnosis is made in the abdominal CT taken for further evaluation after free air is seen on plain X-ray, an easily accessible imaging method in emergency services. This appearance is caused by the perforation of air-filled cysts in the intestinal wall. CT imaging is important in arranging the conservative or surgical treatment of the disease. Intestinal ischemia, total obstruction, or air in the portal vein indicate that the treatment should be planned surgically<sup>6</sup>. While evaluating the etiology of abdominal pain in our case, an abdominal CT was performed, and PCI was diagnosed due to free air under the right diaphragm in the direct X-ray. In our case, diagnostic laparotomy was planned for our patient because the examination findings were positive, and GIS perforation could not be excluded.

PCI management includes emergency diagnostic laparotomy or conventional treatment. Emergency diagnostic laparotomy should be considered in patients with signs of peritonitis, metabolic acidosis, or portal vein gas<sup>7</sup>. Recognition of pneumoperitoneum that does not require surgery is important in preventing unnecessary surgical interventions that cause infections, complications, or prolonged recovery periods in patients<sup>8</sup>. In our case, emergency laparotomy was decided because of peritonitis findings, high inflammatory markers, and radiological intraperitoneal free air. However, no perforation was detected during exploration.

Regardless of the underlying cause of PCI and the presence of symptoms, all patients should be treated medically. Ciprofloxacin and metronidazole therapy should be administered for their activity against aerobic and

anaerobic bacteria to control possible overgrowth of hydrogen-producing bacteria that alter the gut microbiota<sup>9</sup>. Ciprofloxacin and metronidazole were started in the treatment of our case, and the treatment was continued for one week.

In conclusion, PCI may present with an asymptomatic or life-threatening clinic. This situation highlights the importance of carefully evaluating clinical and radiographic findings in PCI's diagnosis and treatment approach in the emergency department. Although PCI clinic may occur due to accompanying comorbid diseases or after drug use, it can also happen spontaneously.

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