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The appearance of free-air in the abdomen with related pneumatosis cystoides intestinalis: Three case reports and review of the literature



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ABSTRACT

INTRODUCTION: Pneumatosis sistoides intestinalis (PSI) is a rare condition with unknown origin, defined as the appearance of gas-filled cysts in the intestinal wall. It usually occurs due to respiratory infections, tumor or collagen disease, traumas, immunosuppression.

PRESENTATION OF CASE: Three patients with PSI were examined that followed up and treated in our clinic. The first patient was hospitalized for emergency treatment of previously diagnosed free-air under the diaphragm. He had a defense on physical examination and free-air was detected in X-ray and abdomen CT. We decided to laparotomy and peroperatively, stenotic pylorus with an abnormally increased stomach and gas-filled cysts were seen in the terminal ileum. Antrectomy and gastrojejunostomy with partial ileum and cecum resection and end ileostomy were performed. The second patient underwent laparotomy because of intraperitoneal free-air and acute abdomen. Partial ileum and cecum resection and ileotransversostomy were performed. The third patient with intraperitoneal free-air was treated with antibiotics, oxygen treatment and bowel rest.

DISCUSSION: PSI is usually asymptomatic. Plain radiographs, USG, CT, upper gastrointestinal endoscopy, colonoscopy can use for diagnosis. Treatment of PSI depends on the underlying cause; include elemental diet, antibiotics, steroids, hyperbaric oxygen therapy and surgery.

CONCLUSION: In patients with asymptomatic and symptomatic PSI are different treat. Symptomatic PSI can be safely treated antrectomy and gastrojejunostomy with partial ileum and cecum resection.

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1. Introduction

Pneumatosis sistoides intestinalis (PSI) a rare condition that is characterized by gas-filled cysts in the intestinal submucosa and subserosa.^{1–3} Its etiology is unclear and several hypotheses have been proposed.² PSI may occur as primary or secondary and often seen terminal ileum in the gastrointestinal tract.^{3–5} Clinics of PSI is broad, ranging to asymptomatic from diffuse peritoneal signs. Respiratory infections, tumor or collagen disease, traumas, immunosuppression and the use of steroids are associated with PSI.⁶ Imaging tools, endoscopy, surgery and sometimes pathological examinations are used in the diagnosis.^{7,8} Although PSI is usually benign disease, sometimes high mortality is observed. In asymptomatic patients are used hyperbaric oxygen treatment, antibiotics and rest to bowel; in patients with signs of peritonitis

are recommended to surgical treatment.^{3,6–9,16–21} We want to discuss 3 case of with PSI that followed-up and treated in our clinic.

1.1. Case report 1

A male patient aged 62 was hospitalized for emergency treatment of previously diagnosed free air under the diaphragm. At the time of arrival the patient reported supraumbilical pain, nausea and vomiting in the past two weeks. The problems of abdominal discomfort and pain connected with taking food had started 21 years earlier, with intermittent remissions and there was a history of stomach bled before 15 years. A few months before, the intensity of the symptoms had increased and continued with nausea, vomiting and weight loss (15 kg in a year) and his body mass index (BMI) was 17 kg/m². He often received proton pump inhibitors and there was not history of gastroscopy. On physical examination the abdomen was tenderness and defense at palpation in the epigastrium. In laboratory analysis hemoglobin was 10.3 g/l and white blood cell was 11.6 × 10³/μL. X-ray showed free-air under the

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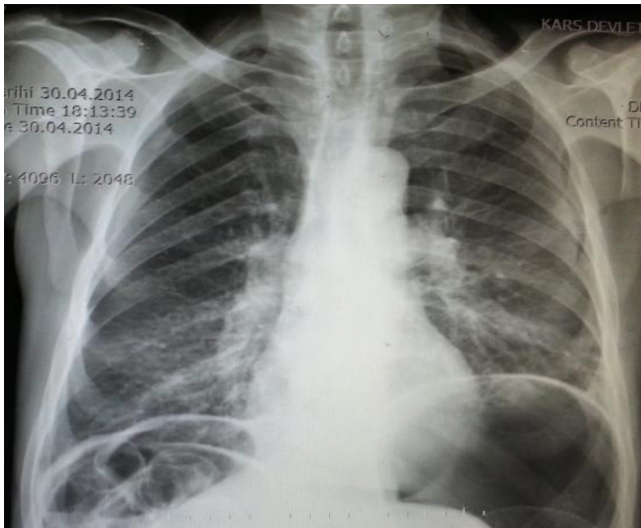


Fig. 1. X-ray free air under the diaphragm.

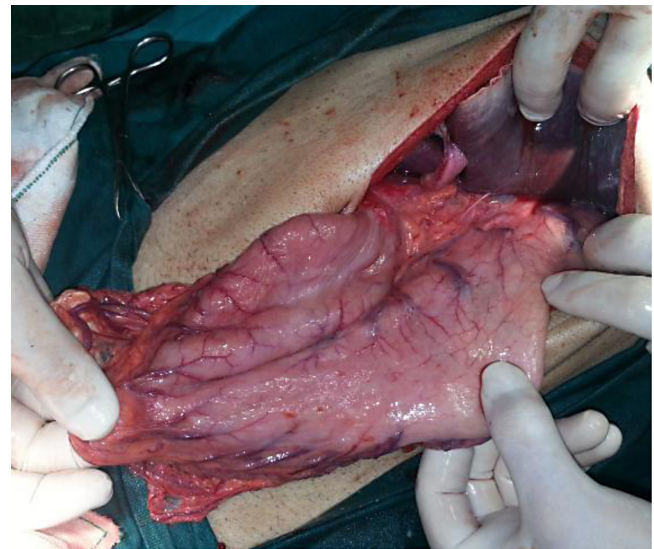


Fig. 3. Stenotic pylorus and dilated stomach.

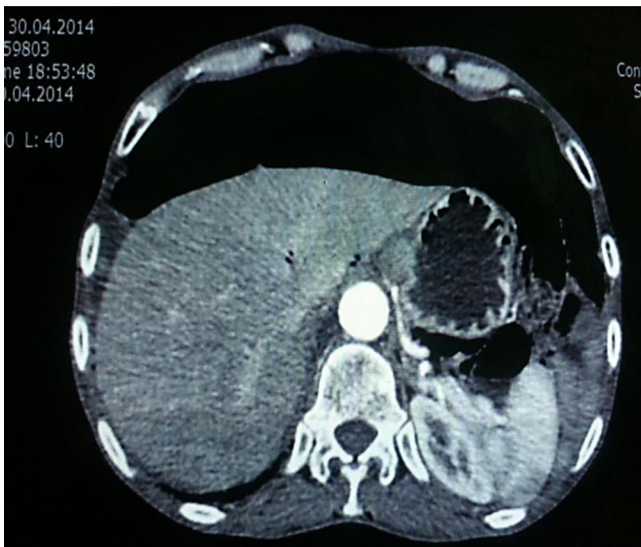


Fig. 2. Abdominal CT showed intraperitoneal free air.



Fig. 4. Gas-filled cysts in the terminal ileum.

diaphragm (Fig. 1). Contrast-enhanced abdominal CT was detected free-air in the abdomen (Fig. 2). The patient was admitted and prepared for the operating theatre where the operation was started. A supraumbilical median laparotomy was performed. After opening the abdominal cavity and exploration, the following finding was present: stenotic pylorus with an abnormally increased stomach and gas-filled cysts were seen in the terminal ileum (Figs. 3–5). After that we decided to make the first resection of that antrectomy and were performed retrocolic, isoperistaltic gastrojejunostomy. Partial ileum and caecum were resected, after then was performed end ileostomy. The operative wound was closed after haemostatic control and drainage set. Specimens were sent to the pathology department.

At first, when the specimen sections from the ileum and proximal colon; macroscopic findings were noted that cystic lesions were filled with air and in greatest diameter 2 cm. Under the microscope, cystic lesions were swollen mucosa, with air filled in submucosa and subserosa. Additionally, the cysts were not communicated with each other and in different sizes which means that the finding at the terminal ileum was pneumatosis cystoides intestinalis (Figs. 4–6).



Fig. 5. Gas-filled cysts at mucosa in the terminal ileum.

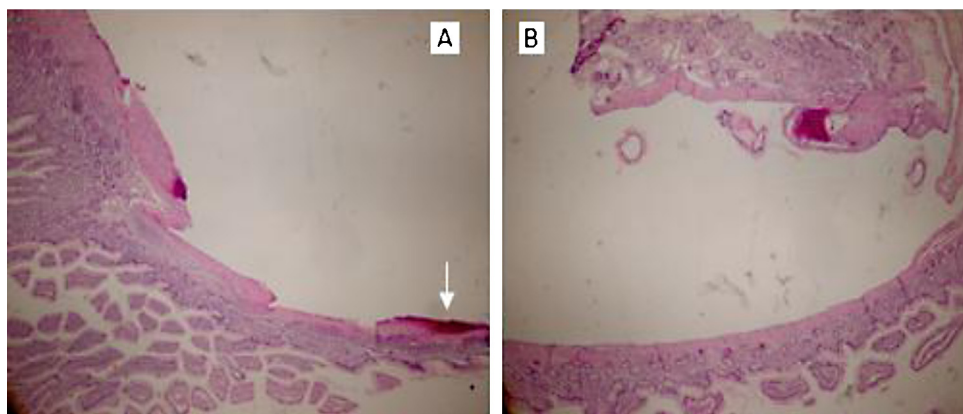


Fig. 6. Histopathological appearances of gas-filled cyst. A) The Low-cubic cystic epithelium in serosa of ileum (white arrow, H&E $\times 100$). (B) Gas-filled cysts in serosa cecum.

At second, operative material of stomach with increased dimensions, decreased elasticity, tall and prominent mucosal folds without macroscopic erosions and ulcerations, astenotic place on the pylorus with the lumen passable for 0.6 cm (Fig. 3). Microscopic finding, there was seen chronic inflammatory infiltration with lymphocytes and intestinal metaplasia into the antrum with bleeding erosions focus and an oedematic edge of the pylorus. In addition to, *Helicobacter pylori* was viewed intensively.

The postoperative follow-up was occurred duodenal stump leakage and underwent relaparotomy. The periton was debrided, did tube duodenostomy and placed drains. The patient discharged from the hospital on the 35th postoperative day. The ileostomy was closed after 1 month later. He resumed normal activity in the follow-up visit 1 month after the second surgery.

1.2. Case report 2

A male patient aged 70 was admitted with abdominal pain and vomiting in emergency room. He has received proton pump inhibitor therapy for long years and weight loss (BMI = 17 kg/m²). He had not undergone gastroscopy and often received conservative treatment. On physical examination the abdomen was tenderness and defense at palpation in the epigastrium and upper right quadrant. X-ray showed to free-air under diaphragm. In the laboratory white blood cell was $9 \times 10^3/\mu\text{L}$ and amylase was 220 U/L. Intraperitoneal free air and millimetric contrast agent leakage in curvature minor of the stomach has been detected with contrast-enhanced abdominal CT. Therefore, we were decided laparotomy. In exploration were seen gas-filled cysts in the terminal ileum and pyloric stenosis that 2 \times 1 cm diameter and permitting passage. Partial ileum and cecum resected and were performed site to site ileo-transversostomy anastomosis. He was discharged on postoperative day 8 and performed upper gastrointestinal endoscopy a month later. We detected pyloric stenoses that did not allow of endoscopy passage in to the duodenum. Multiple biopsies were taken and intestinal metaplasias were reported by a pathologist. Electif antrektomy and gastroenterostomy were performed after a month. Pathology reports were benign and determined diffuse intestinal metaplasia and widespread involvement of *Helicobacter pylori*. He was discharged from hospital on eighth postoperative day. The patient resumed normal activity in the follow-up visit 20 day after the surgery.

1.3. Case report 3

A male patient aged 81 was admitted with abdominal pain from internal medicine of clinics. The patient had been operated before 4 years and had congestive heart failure, chronic lung disease

and prostate disease. Because of upper gastrointestinal bleeding were given two units of erythrocyte suspension. His BMI was 21 kg/m². On physical examination was detected median incision scar and only tenderness at palpation in the epigastrium and there was no acute abdomen. The patients had gas or stool output and white blood cell was $5.6 \times 10^3/\mu\text{L}$. Free-air was determined under diaphragm and intraperitoneal in X-ray and abdomen CT, respectively. Upper gastrointestinal endoscopy and colonoscopy were performed and detected hiatus hernia, erythematous pangastritis and grade 2 internal hemorrhoids. The wide involvement of *H. pylori* was reported after antrum biopsies. The 2–4 lt/dk oxygen and antibiotic therapy were given to patients. The patient subsequently relieved and was discharged from the hospital after on fourteen day. His general condition is good and follows up with outpatient.

2. Discussion

Pneumatosis sistoides intestinalis (PSI) a rare condition that is characterized by gas-filled cysts in the intestinal submucosa and subserosa, is most frequently observed in the terminal ileum of gastrointestinal tract.^{1,2} PSI is more frequently seen in the first days of life and 60 years of age and its estimated prevalence is 0.03%.^{2,3} Its etiology is unclear and there are plenty of theories about the pathogenesis of pnemautosis intestinalis, including mechanical, bacterial, pulmonary, and caused by the increase in the mucosa permeability. The most accepted theories are the bacterial and the decrease in the mucosa permeability.^{4,11} There are three pathologic forms; microvesicular or pseudolipomatosis, cystic and diffuse.⁴ Our 3 patients were over the age of 60 and gas-filled cystic lesions were localized in the ileum. *H. pylori* can be considered in etiology of PSI that identified in the biopsy of 3 patients (Figs. 4–5).

Pneumatosis cystoides intestinalis is classified as primary disease (15%) of unknown cause and secondary (85%) associated to gastrointestinal diseases, as intestinal obstruction⁵, cystic fibrosis, peptic ulcer, diverticula, inflammatory bowel disease,²³ mesenteric infarction, chronic intestinal pseudo-obstruction,⁷ respiratory infections, tumor²⁴ or collagen disease, traumas, immunosuppression and the use of steroids.^{3,8} We believe that PSI develops in our three patients which secondary to gastrointestinal obstruction or pyloric stenosis.

The history of patient, symptoms and physical examination are important for diagnosis of PSI. Patients are usually asymptomatic. Diarrhea, constipation, rectal bleeding, tenesmus, nonspecific symptoms such as weight loss and abdominal pain may occur in symptomatic patients.³ Volvulus, intestinal obstruction, blood pressure pneumoperitoneum, rectal bleeding, intussusception and intestinal perforation can be seen by 3%.^{4,7,8} There was rectal

bleeding in a patient and in three of our patients had abdominal pain and weight loss (Table 1).

The imaging methods are important in the diagnosis of PSI. These include plain radiographs, USG, barium series, CT, CT-colonoscopy, magnetic resonance imaging and MRI-colonography, endoscopy, colonoscopy.^{3,10–12} X-ray can show intraperitoneal free-air and abdominal CT can detect pneumoperitoneum, portomesenteric venous air, bowel wall thickening, bowel dilatation.^{12,13} Lee et al.¹³ reported in their work of eighty-four patients that importance of CT in the diagnosis of PSI.

A definitive diagnosis is difficult with the upper gastrointestinal endoscopy and colonoscopy, because of submucosal cysts can be mixed with polyps.¹⁴ A laparoscopic exploration is quite useful to confirm a PCI diagnosis, if the physical examination findings are suspicious, and particularly in cases that are not preoperatively diagnosed clearly using the above-mentioned radiological methods. Diagnostic laparoscopy provides the convenience of converting to open surgery as well as confirming the diagnosis.^{3,12,13}

Treatment of PSI depends on the underlying cause, so that include elemental diet, antibiotics, steroids, hyperbaric oxygen therapy and surgery.^{3,7,8,11,12,15–17} In asymptomatic patients with free-air at X-ray and abdominal CT reporting gastrointestinal perforation direction, is a great handicap for the surgeon. So we should be careful in the diagnosis of disease.

Because of spontaneous remission can see rate of 50% in treating the disease and may occur gas-filled cysts again after surgery. There is not a specific treatment in asymptomatic patients. Conservative approaches, including nasogastric decompression, intestinal rest, antibiotic therapy and oxygen, are recommended for patients with positive examination findings and normal biochemical parameters who are confirmed radiologically to have no intestinal ischemia or perforation.^{2–4,15} In asymptomatic patients can be given hyperbaric oxygen (HBO) or absolute atmospheres (non-HBO) therapy may be given. Oxygen therapy has long been recognized as an effective therapy for PI, leading to cyst regression on imaging and symptom resolution.¹⁸ Applying 250 mmHg PO₂ pressure or 70% oxygen inhalation for 5 days or 2.5 atmospheres of hyperbaric oxygen pressure for 150 min/day for 3 consecutive days can lead to resolution of gas collection within a cyst.^{10,13,24}

Shimada et al.¹⁹ in their series of 15 cases showed that were successfully treated with hyperbaric oxygen. Togawa et al.²⁰ gave HBO therapy to seven patients and atmospheres absolute (ATA, non-HBO) to 20 patients in asymptomatic patients with PSI. HBO therapy was given mean of 4.7 days, ATA treatment was given mean of 14.6 days and have found lower recurrence rates.

In asymptomatic patients with PSI is not necessary surgery treatment.^{3,7,10,13} Surgical treatment should be considered in patients who do not respond to medical therapy. Early surgical intervention should be considered in any patient with signs of perforation, peritonitis, sepsis or predictors of poor outcomes.^{4,5,9,10} Emergency surgery should be considered in PSI patients with the pH is less than 7.3, actual bicarbonate level is less than 20, amylase is higher than 200 U/L or signs of disseminated coagulation intravascular. Similarly, patients who are immunosuppressed with underlying liver disease, sepsis, and hypotension are all at a higher risk of mortality from PI.²²

Radiologically, the presence of portal venous gas is highly indicative of severe disease, and when present, mortality rates range from 37% to 75% (7, 22).

In patients with symptomatic PSI, laparotomy and laparoscopic exploration should be considered. Arıkanoğlu et al.³ in a series of seven cases showed that safely performed ileal resection and anastomosis in 3 patients, underwent laparotomy in 2 patients, choledocotomy, drainage and cholecystectomy in a patient, and laparoscopic exploration in a patient. We performed to 2 patients ileal and cecal resection and anastomosis with antrectomy and

Table 1
Demographic and clinical characteristics of the three patients with PSI.

No.	Age	BMI (kg/m ²)	Sex	Complaints	Medical history	WBC (10 ³ /μL)	Signs peritonitis	Free air in X-ray and CT	H. pylori	Location	Etiology	Surgical treatment	Length of hospital stay (d)	Postoperative complication	Follow-up (months)
1	62	17	♂	AP, N, V	Upper GI bleeding	11.1	T and D	Yes	Yes	Terminal ileum	Secondary	Ileum + cecum resection and anastomosis with antrectomy and GE	45	Duodenal stump leakage	2
2	70	18	♂	AP, V	Upper GI bleeding	9	T and D	Yes	Yes	Terminal ileum	Secondary	Ileum + cecum resection and elective antrectomy and GE	8	No	3
3	81	21	♂	AP	Bowel obstruction and Upper GI bleeding	5.6	T	Yes	Yes	Terminal ileum	Secondary	Non-surgical	7	No	3

WBC: white blood cell; AP: abdominal pain; N: nausea; V: vomiting; BMI: body mass index; H. pylori: *Helicobacter pylori*; GI: gastrointestinal; T: tenderness; D: defans; GE: gastroenterostomy; CT: computed tomography.

gastroenterostomy. There are not recurrences any patients. One of our patients was treated with non-surgical (Table 1).

3. Conclusion

Pneumatosis intestinalis is a rare condition that may be occur a sign of an underlying systemic disease. In PSI patients with peritonitis can be safely treated ileal and cecal resection and anastomosis with antrectomy and gastroenterostomy.

Conflict of interest

There are no conflicts of interest.

Funding

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Ethical approval

Consent was obtained from the patient for publication.

Author contributions

Mehmet Aziret contributed in study concept, design, data collection and writing the paper. Hasan Erdem contributed in design and interpretation. Yiğit Ülgen and Şahin Kahramanca helped in data collection. Süleyman Çetinkünar contributed towards data analysis. Hilmi Bozkurt contributed in study concept. İlhan Bali helped in interpretation. Oktay İrkörücü contributed towards writing the paper and study concept.

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