

# Pneumatosis Cystoides Intestinalis

## Clinical Experience in a Single Center



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### ABSTRACT

The purpose of the present study was to examine deeply the treatment strategies implemented in four patients diagnosed with pneumatosis cystoides intestinalis and their results. Medical records of four patients who had been treated at the diagnosis of pneumatosis cystoides intestinalis between January 2006 and November 2011 were investigated retrospectively. Three (75%) of the patients were male and 1 (25%) female, with the average age of 47.3±19.3 (21-66) years. Pain in abdomen was the first symptom complained by all four patients on admission. While the findings consistent with the peritonitis were revealed during physical examination in three of the cases, abdominal tenderness was detected in one patient. Of all the cases, 2 were accepted as the primary cases due to unknown etiology, while chronic obstructive pulmonary disease and peptic ulcer were held responsible in the etiology, thus assigning these cases in the secondary pneumatosis cystoides intestinalis group. Plain abdominal x-ray images acquired in all patients revealed subdiaphragmatic free gas collection; moreover, free fluid collection within the abdomen was recognized ultrasonographically in 3 patients. The diagnosis of pneumatosis cystoides intestinalis was established through abdominal computed tomography in the patient followed up under conservative treatment. Mortality and complications occurred in none of the patients. Should the physical examination findings associated with the peritonitis be obscured by subdiaphragmatic free gas, such a patient may be erroneously followed up under conservative treatment. Implementation of a surgical strategy should not be deferred in case of acute abdomen.

**Key words:** Pneumatosis cystoides intestinalis, conservative, surgery

### Pnömozis Sistoides İntestinalis (Tek Merkez Deneyimi)

#### ÖZET

Bu çalışmanın amacı Pnömozis sistoides intestinalis tanılı dört hastada klinik, uygulanan tedavi prosedürleri ve sonuçlarının irdelenmesidir. Ocak 2006- Kasım 2011 tarihleri arasında Pnömozis sistoides intestinalis tanısıyla tedavi edilen dört hasta retrospektif olarak incelendi. Hastaların 3'ü (%75) erkek, 1'i(%25) kadın olup, ortalama yaşı 47.3±19.3(21-66) yıl idi. Hastaların tümünde ilk yakınma şikayeti karın ağrısıydı. Üç hastanın fizik muayenede peritonit bulguları pozitif iken, 1 hastada hassasiyet mevcuttu. Hastaların 2'sinde etiyolojisi bilmediğinden primer olarak kabul edilirken, diğer iki hastada ise etiyolojide kronik obstrüktif akciğer hastalığı ve peptik ülser tespit edildiğinden sekonder pnömozis sistoides intestinalis olarak kabul edildi. Hastaların hepsinde ayakta direkt karın grafisinde subdiaframatik serbest hava izlendi. Hastaların 3'ünde ultrasonografide serbest sıvı tarifleniyordu. Konservatif takip edilen hastada pnömozis sistoides intestinalis tanısı Abdominal tomografi ile konuldu. Hiçbir hastada komplikasyon ve mortalite izlenmedi. Subdiaframatik serbest hava varlığında, fizik muayenede peritonit bulguları saptanmaz ise konservatif takip edilebilir. Akut karın durumunda cerrahi müdahale geciktirilmemelidir.

**Anahtar Kelimeler:** Pnömozis sistoides intestinalis, konservatif, cerrahi

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## INTRODUCTION

First defined by Du Vernoy during dissection of a human cadaver in 1730, pneumatosis cystoides intestinalis (PCI) is a rare disease which is characterized by the presence of gas-filled submucosal and subserosal cysts in the intestinal wall and can involve any part of the gastrointestinal system (1-3). The incidence of PCI was reported to be 0.03% in the general population (4). With the advent and widespread use of computed tomography (CT), the former ratio was reported to have risen to 0.37% (5,6). The general clinical scenario in PCI can vary greatly, ranging from admission with such nonspecific findings as abdominal pain, abdominal distension and rectal bleeding to some life-threatening complications such as intestinal obstruction, ischemia, perforation and intussusceptions, which requires emergency surgery (7,8). The diagnosis can be quite challenging due to non-specific clinical findings and rarity of PCI. Although commonly pursuing a favorable clinical course in most of the cases, the disease is also likely to follow a fulminant course, thus leading to mortality (9). The aim of the present study was to investigate the clinical scene, the treatment procedures implemented and their results in four patients with PCI.

## MATERIALS AND METHODS

The medical files of 4 consecutive patients treated in Dicle University Medical Faculty, General Surgery Clinic for the diagnosis of PCI between January 2006 and November 2011 were evaluated retrospectively, recording the age, gender, symptoms on admission, time that elapsed until hospital admission, accompanying disorders, findings from physical examination, conservative or surgical treatments given, findings of laparotomy in the patients undergoing surgery, length of hospital stay, complications developing postoperatively, and mortality. The indication whether to prefer a conservative or a surgical approach is dictated by the clinical symptoms, and findings from physical examination, laboratory tests and imaging modalities. Decision for surgical operation is given on the basis of peritonitis findings. All the patients undergoing surgical operation received 1 gr/IV second-generation cephalosporin prophylactically under emergency conditions prior to the operation. Conservative treatment comprised nasogastric decompression, bowel rest, total parenteral nutrition and administration of 500 mg metronidazole as antibiotic agent at 8-hour intervals for 7 days (10). Daily physical

examination and white blood cell counting were integral parts of the conservative follow-up.

### *Statistical Analysis*

During the evaluation of the findings obtained from the study, SPSS (Statistical Package for Social Sciences) 13.0 program for Windows was used for the statistical analysis. The quantitative data were presented as mean  $\pm$  standard deviation and median follow up period was calculated.

## RESULTS

### *Demographic and Clinical Findings*

Of all the patients whose demographic and clinical features and implemented treatment procedures are summarized in Table 1, 3 (75%) were male and 1 (25%) was female, with the average age of  $47.3 \pm 19.3$  (21-66) years. The first symptom complained by all patients in common was abdominal pain. 3 patients had the complaints of nausea-vomiting, and 2 suffered from weight loss and constipation. While the findings consistent with the peritonitis were revealed during physical examination in three of the cases, abdominal tenderness was detected in one patient. The biochemical parameters were found to be normal in all patients. Whole blood count disclosed normal number of white blood cells in 3 of the cases, whereas leukocytosis was evident in one case. Of all the cases, 2 were accepted as the primary cases due to unknown etiology, while chronic obstructive pulmonary disease and peptic ulcer were held responsible in the etiology, thus assigning these cases in the secondary pneumatosis cystoides intestinalis group.

### *Radiological Findings*

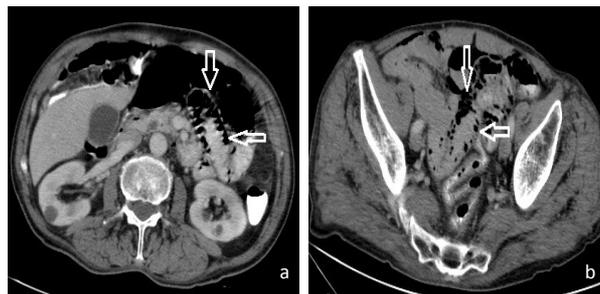
Plain PA chest x-rays and standing abdominal x-rays were obtained in all patients. Subdiaphragmatic free gas view was evident in the plain abdominal x-rays (PAXR) of all the patients, which was also accompanied in the second patient by presence of fluid-gas levels consistent with ileus (Figure 1). Abdominal ultrasonography (USG) recognized intaperitoneal free fluid collection in 3 cases. CT images acquired in the patients followed up under conservative treatment revealed views compatible with gas-filled cysts, other than free fluid collection (Figure 2).



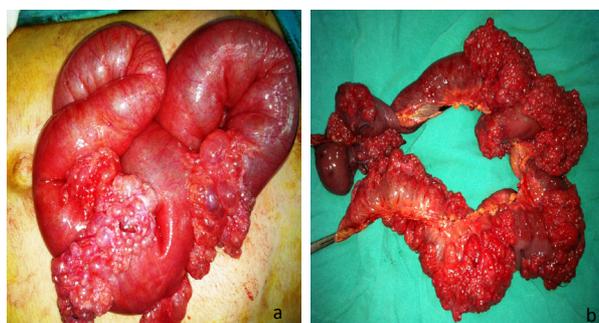
**Figure 1.** Subdiaphragmatic free gas collection and gas-fluid levels related to ileus, revealed by PAXR.

### Surgical Findings

While laparotomy was undertaken in 3 of the patients, 1 was managed conservatively. A perforated peptic ulcer and multiple gas-filled cysts within a 100 cm-long ileal small intestinal segment in addition to ischemia were detected during the laparotomy in the first of those undergoing surgery, followed by implementation of a constellation of procedures, including primary suturing of the peptic ulcer perforation and resection of around 100 cm-long intestinal segment, along with double-barreled enterostomy due to presence of peritonitis and highly edematous intestine. In the second patient, pneumatosis cysts within a 150 cm-long ileal segment in addition to edematous intestines without any ischemia and passage was detected; moreover, peritonitis secondary to intestinal perforation was also evident. As such, a 150 cm-long small intestinal resection and end-to-end anastomosis was undertaken at a distance of 10 cm from the ileocecal valve (Figure 3). During laparotomy of the last patient, multiple gas-filled cysts were observed in the subserosa of the 180 cm ileal small intestinal segment starting 10 cm proximal from the ileocecal valve, with the biggest cyst reaching 3 cm in diameter but with-



**Figure 2.** Gas-filled cystic images related to pneumatosis intestinalis within the small intestinal wall.



**Figure 3.** Views from the 150 cm-long surgical specimen from a patient with pneumatosis intestinalis, (a) during the operation and (b) after the operation.

out any simultaneous intestinal perforation. The patient was suffering from generalized peritonitis. Upon searching for the primary focus, adhesions in thick bands were noted between the liver and the duodenum. After attempts to release these adhesions, a 0.5 cm closed perforation area extending on the same line as a prior pyloroplasty was detected, and then followed by primary repair.

### Developments in the postoperative period

The average length of hospital stay of the patients was  $11.8 \pm 3.8$  (7-16) days. No complication or mortality was observed in the patients. The median follow-up duration was 18 (1-48) months.

**Table 1. Demographic and clinical characteristics of the patients and the treatment procedures.**

NO	Gender	Age(year)	Symtoms	TAH(day)	Physical Examination	Etiology	WBC (/mm3)	Radiological Imaging	Localization	Treatment	Hospital stay( day)
1	M	46	AP+N+V+C	3	Peritonitis (+)	Peptic ulcer perforation	9900	PAXR+USG	Small intestine	Primary suture + ileal resection + double enterostomi	13
2	F	21	AP+N+V+C	4	Peritonitis (+)	Normal	21000	PAXR +USG+	Small intestine	Ileal resection + end-to end anastomosis	16
3	M	56	AP+N+V+C	4	Peritonitis (+)	Peptic ulcer perforation	6500	PAXR	Small intestine	Primary suture	11
4	M	66	AP+C	4	Peritonitis (-)	Normal	7600	PAXR +USG+CT	Small intestine	Conservative	7

M: Male, F: Female, AP: Abdominal pain, N: Nausea, V: Vomiting, C: Constipation, TAH: Time of admission to hospital PAXR: Plain Abdominal X-ray, USG: Abdominal ultrasonografi, CT: Abdominal tomografi, WBC: White blood cell

**DISCUSSION**

PCI is a rare disease with the incidence peaking in the fifties which has a predilection much more toward male subjects and characterized by intramural gas-filled cystic formations involving any part of the gastrointestinal system, along with pneumoperitoneum or pneumoretroperitoneum (7). The average age of the patients included in our study was 47, three of whom were male.

Following three theories have been recognized in the pathogenesis of PCI: Mechanical theory: Increased intraluminal pressure as a result of gas entrapment in the intestinal lumen due to obstruction prompts the gas to dissect through the mucosal layer into the lymphatic canals, therefrom travelling with the help of peristaltic movements distally alongside the submucosal area of the intestine. Pulmonary theory: Movement of the gas of high pressure released as a result of rupture of the gas-filled cysts in a patients with obstructive pulmonary disease through posterior mediastinum into the retroperitoneal space, where it passes into intestinal mesentery, thus eventually reaching the intestinal lymphatics. Bacterial theory: The gas produced by the fermenting organisms such as Escherichia Coli and Clostridium spp. is absorbed and retained by the intestinal submucosa, then moving into the intestinal lymphatics (11,12).

On the basis of the underlying etiology, PCI is divided into two groups as primary and secondary. In the primary pneumatosis (idiopathic), submucosal layer of the colon is the most frequently involved area in 15% of the sufferers, whereas the secondary pneumatosis accompanies other disorders. The disease has a predilection much more towards subserosal layer of the small intestines in 85% of the cases (13,14). Many gastrointestinal system diseases, such as appendicitis, Crohn's disease, pyloric stenosis, ulcerative colitis, diverticular disease, necrotizing enterocolitis, gastroduodenal ulcer and sigmoid volvulus are likely to accompany PCI (10). Some studies reported PCI involving the small intestines as a common accompaniment to peptic ulcer or perforated peptic ulcer in 55% of the cases, which is, however, not documented to be pathognomonic (15,16). Similarly, PCI was also reported to accompany some other diseases which are not associated with the gastrointestinal system such as chronic obstructive pulmonary disease (COPD), collagen tissue disorders, AIDS and glucocorticoid use (17). PCI cases secondary to surgical or endoscopic traumas were also reported (11). Small intestinal

involvement was recognized in all our cases. While 2 patients suffered from the primary disease, the etiology in the patients with the secondary disease was attributed to peptic ulcer or COPD.

Development of pneumoperitoneum secondary to rupture of the cysts in the absence of any findings related to peritoneal irritation is the pathognomonic feature of the disease (18). In PCI, no pronounced correlation has been established among the area of involvement, clinical findings and severity of the disease. PCI may rarely lead to a mortal scenario, including peritonitis or sepsis (19). Such non-specific symptoms as abdominal pain, abdominal distension, nausea-vomiting, diarrhea, constipation, rectal bleeding, tenesmus, and weight loss may be observed in PCI (11). In 3% of the cases, an emergency surgery may be necessitated by such conditions as volvulus, intestinal obstruction, tension pneumoperitoneum, bleeding, intussusceptions, intestinal perforation, peritonitis and abdominal sepsis (20). Three patients underwent laparotomy due to positive peritonitis findings, whereas non-specific clinical findings along with abdominal tenderness in the physical examination were evident in the patient managed conservatively.

To establish a definitive diagnosis of PCI clinically and radiologically is quite challenging. Imaging modalities are of importance in the diagnosis. PAXR, USG, CT, endoscopic imaging modalities and double-contrast colon radiography may prove beneficial in the diagnosis (11). Similarly, PAXR is an easily applicable method which is quite useful in the diagnosis of PCI; moreover, presence of circular gas in the bowel and the mesentery is characteristic of PCI (19). In CT views, presence of gas entrapped within the wall of the bowel is characteristic to PCI; CT can also reveal gas bubbles within the intestinal lumen (4). Over and above, CT can prove beneficial in recognition of the other pathologies associated with intraluminal gas or submucosal fatty cysts (21). However, CT fails to show intestinal ischemia and necrosis (6). In our study, subdiaphragmatic free gas collection was shown through PAXR in all of the cases. Although USG did not provide benefit in the diagnosis of PCI, CT performed in the patient managed conservatively was diagnostic.

The indication whether to prefer a conservative or a surgical approach in the treatment of PCI is dependent on the course of the disease. Most patients are managed conservatively (3, 22). Obviation of the need for surgical intervention and feasibility of conservative treatment

were reported in the patients with benign pneumoperitoneum secondary to rupture of the subserosal gas bubbles (11, 12). The clinical scene of PCI was reported to improve as the factors responsible in the etiology were abolished (23). What has been recommended in the conservative treatment is nasogastric decompression, administration of metronidazole as antibiotherapy and hyperbaric oxygen therapy for absorption of the gas accumulated within the intestinal wall (6, 12, 19). The rate of mortality was reported to range from 22% to 50% in the cases followed up due to the diagnosis of PCI and who did not undergo surgery (24,25). One of our cases was managed conservatively. Metronidazole therapy was given in the postoperative period to the patients undergoing surgery. Hyperbaric oxygen therapy could not be implemented, since our hospital lacked such facility. To us, the fact that our patients benefit from the metronidazole treatment supports the bacterial theory.

Indication for surgical intervention should not be dictated only by the observation of subdiaphragmatic free gas in PAXR. Pneumoperitoneum does not necessarily indicate intestinal perforation, but may also suggest the rupture of gas-filled cysts in PCI (26). Abdominal sepsis, intestinal perforation or clinical scene of peritonitis are the indications for emergency surgery (7,8). Resection was reported not to be necessarily required in the absence of peritonitis, intestinal ischemia and ileus during laparotomy (23). In such cases, mortality may rise to as high as 44 percent, despite surgical intervention (25). While resection was performed in 2 of our patients undergoing surgery due to presence of intestinal ischemia and ileus, 1 patient underwent only a primary repair for the ruptured peptic ulcer without any additional procedure owing to normal appearance of the intestines. No complication or mortality was observed in the postoperative period in our patients.

The general clinical scenario in PCI can vary greatly, ranging from asymptomatic presentation to emergency conditions. The definitive diagnosis can be established on the basis of the clinical and radiological findings, and imaging modalities. When no finding compatible with peritonitis was detected during physical examination in the presence of subdiaphragmatic free gas collection, conservative management is likely to provide favorable results. A close follow-up is mandatory, and conversion to surgical management should not be delayed in case of acute abdomen.

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