

A Patient Suffering from Pneumatosis Cystoid Intestinalis

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Article information	Abstract
<p>Article history: Received: 17 Sep 2010 Accepted: 18 Oct 2010 Available online: 20 June 2011</p> <p>Keywords: Pneumatosis Cystoid Intestinalis Pneumoperitoneum Pelvic Trauma</p> <p>*Corresponding author at: Kerman University of Medical Sciences, Kerman, Iran. E-mail: aghaei_afshar@yahoo.com</p>	<p>Pneumatosis Cystoid Intestinalis (PCI) is a relatively rare disorder that occurs in different parts of the stomach and intestines as gas-filled cysts. It is predicted to be prevalent among 0/2- 0/03% of the population. In 85% of cases, Pneumatosis Cystoid Intestinalis is caused by a specific disease. In this article, a patient will be introduced who was admitted 3 days after the pelvic trauma presented with symptoms of intestinal obstruction. The conducted examinations showed generalized abdominal tenderness and in CXR plenty of free air was observed under the diaphragm. After the primary diagnosis of intestinal obstruction (ileus), the patient was put under laparotomy. During laparotomy, PCI was seen under the intestinal serosa and a significant amount of air was released and many parts of contusion were observed in the small intestine along with fibrin formation without pus which was restored and the patient was discharged in a well general condition.</p> <p>Copyright© 2012 Zahedan University of Medical Sciences. All rights reserved.</p>

Introduction

Pneumatosis Cystoid Intestinalis (PCI) is an apparently rare disorder, a few cases of which has been reported so far, and during the searches conducted throughout the country, no case was found similar to ours, and trauma has not been identified as the underlying cause of disease. This disease occurs in the small intestine, colon and stomach as multiple non-correlated gas-filled cysts with thin wall with no epithelial, and it can occur as submucosa or subserosa.

It is predicted to be prevalent among 0.02-0.003% of the population [1]. In 85% of cases, Pneumatosis Cystoid Intestinalis is caused by a specific disease, which is called the secondary PCI [2]. Also in 15% of cases, no underlying cause can be found for it which is called primary or idiopathic PCI. The secondary type is observed at conditions such as inflammatory bowel disease, diverticular, pseudomembranous colitis, sigmoid volvulus, pyloric stenosis, collagen vascular diseases, obstructive pulmonary disease and asthma (Table 1) [2].

The secondary type will cause a high rate of mortality when it is caused by necrotizing or intestinal puncture diseases. Idiopathic PCI is always a benign disorder. It is often asymptomatic and is incidentally diagnosed in a patient with mild symptoms or no symptoms [1,2]. The patients are occasionally admitted with several clinical symptoms such as abdominal pain, abdominal distension, bloody stools, constipation and diarrhea [3]. The disease is often diagnosed accidentally during imaging, endoscopy or laparotomy. It is observed as linear or cystic lucencies in a plain abdominal radiography [4,5]. If the patient has no symptoms and no underlying disease, there is no need to treat. To treat the underlying disease, oxygen

therapy and antibiotics are recommended for symptomatic cases. If symptoms are severe, surgery is recommended [6].

Table 1. Diseases Associated with (PCI)

A- Abdominal emergencies: intestinal ischemia, intestinal infarction, intestinal obstruction, necrotizing enterocolitis
B- Mucosal damage: peptic ulcer, Crohn's disease, ulcerative colitis, eating hot food stuff, diverticular perforation
C- Infections: AIDS, clostridium difficile, tuberculosis, Whipple disease, cryptosporidium, mycobacterium avium-intracellulare (MAI), CMV
D- Pulmonary diseases: Artificial respiration device, CF, asthma, COPD
E - Endoscopic procedures: Gastroscopy, colonoscopy, sclerotherapy, biliary stent placement
F - Gastrointestinal motility disorders: diabetes, scleroderma, HD, intestinal pseudo-obstruction, illusion bypass, pyloric stenosis or obstruction
G- Immunological diseases: Steroids, chemotherapy, PTLD disorders, bone marrow transplantation, AIDS, amyloidosis, GVHD, other transplantations, collagen vascular diseases

Case presentation

The case is a 40-year old patient who was admitted to Afzalipour Hospital in Kerman for generalized abdominal pain and recurrent vomiting 1-2 hours after eating. The patient developed pelvic trauma in a car accident approximately 10 days ago and three days after the accident suffered from generalized abdominal pain which is associated with abdominal distension and inability to pass gas or defecate. The patient also notes that during that period he has been admitted to hospital for several times, but he was given no certain treatment and he was discharged after supportive therapies. Nevertheless, the pain continued and he referred to this institute to continue

treatment. The performed examinations showed that abdomen was soft and slightly distended and the patient had generalized abdominal tenderness with no ascites and organomegaly. Other examinations were normal. Gas passing or defecation was occasionally noted. A significant amount of free air was observed under the diaphragm during the performed CXR (Fig. 1).

Air bubbles were observed throughout the intestine in the upright abdominal radiography (Fig. 2). Endoscopy was performed for him, which was normal. The patient was put under laparotomy and as his abdomen opened, a significant amount of free air was released, and many parts of contusion were observed in the small intestine along with fibrin formation with no pus. Other parts were normal. Those parts with suspicious perforation were restored and the patient was discharged in good condition.

Figure 1. Observation of free air under the diaphragm in patients with PCI



Figure 2. Observation of air bubbles throughout the intestine



Discussion

PCI is a rare disorder and a few cases of which have been so far reported and occurs in the small intestine, colon and stomach as multiple non-correlated gas-filled cysts with non-epithelial thin wall [1,2]. Other areas such as duodenum and extra-intestinal tissues, such as mesenteric, lymph nodes, omentum, falciform ligament and peritoneum are rarely involved [5]. More severe presentation of the disease such as intestinal infarction, pseudomembranous enterocolitis or necrotizing enterocolitis is rare especially in babies [6].

PCI is divided into two categories of primary and idiopathic. Secondary PCI often involves the small intestine and ascending colon, while the idiopathic type has a tendency to involve the left colon [1,2]. Few cases have been reported with simultaneity of small bowel obstruction and PCI. Small bowel obstruction is presented as one of underlying causes of PCI. PCI may be observed at any age, but it is most common between 30 and 50 years of old [7]. Pathogenesis and natural development of PCI have not been quite identified; various hypotheses are presented to justify PCI development [2].

The Mechanical Theory assumes that tearing into the gastrointestinal wall, gas seeps through the wall. In fact, the content gas of these cysts contains 70% nitrogen and 20% oxygen (like gas composition of air). However, in some studies, the hydrogen concentration has been reported above 50% [8]. On the other hand, oxygen inhalation treats these cysts and the proponents of this assumption rely on these reasons [8,9]. Bacterial theory regards gas creation by bacteria, such as *Clostridium perfringens*, as the cause of PCI and knows the treatment of disorder with antibiotics, such as metronidazole, as a reason to support its claim. Pulmonary theory claims that air released from ruptured alveolar gets into mediastinum and retroperitoneum and it considers gas composition of cysts and prevalence of disease in patients with COPD and asthma as reason to support the claim. Anyway, pathogenesis of PCI remains open to debate [3-5]. According to the conducted studies, the most common etiology which causes this disease is gastrointestinal disorders. In a study conducted in 2005 in Turkey, the cause of this disease is attributed to the ruptured duodenal ulcer and Meckel's diverticulum [9].

In another study conducted in 2008 in Japan, the cause was also attributed to consumption of alpha-glucosidase inhibitor in a diabetic person and the motility disorder followed by it. However, in all the conducted searches, no case was found similar to our report and trauma is not mentioned as the underlying cause of disease [10].

PCI can affect all three layers of gastrointestinal tract. Submucosal cysts are polyp-like and bluish. Subserosal cysts are close to vessels and are seen as bright blue multiple bulges. The cysts have no real wall and in 50% of cases they automatically disappear [9]. PCI can be symptomatic according to the involvement area. For example, it causes vomiting, flatulence or weight loss in small intestine and diarrhea, hematochezia or constipation in colon as symptoms [9]. Physical examination is usually normal and sometimes abdominal distention is seen in these patients. The abdomen may be tender and sometimes a mass may be seen, but fever and symptoms of peritoneal irritation are rarely observed [7,9]. 3% of patients might have complications which are as intestinal obstruction (ileus), volvulus pneumoperitoneum (sometimes recurrent), intussusception, intestinal perforation and bleeding [8]. The disease is often diagnosed incidentally during imaging or endoscopy or laparotomy. It is observed as linear or cystic lucency in plain abdominal radiography. In addition, air may be

observed in peritoneum or retro peritoneum. Radiography with barium is useful to determine the lining of the intestine or stomach. Submucosa type in barium enema is confused with polyposis pseudopolyposis or intraparietal hematoma [4]. Abdominal CT scan is more accurate than plain radiography in the diagnosis of intestinal wall gas. CT scans show mesenteric venous gases or port system in cases when PCI is associated with intestinal infarction. PCI endoscopy shows colonic submucosa as multiple soft, smooth, round and blue polypoid masses within lumen [4,5]. Since cysts contain compressed air, they will shrink if they are ruptured during biopsy.

Plain abdominal radiography and endoscopy were also used on our patient for diagnosis, which were consistent with other studies [4]. Definitive diagnosis of disease in our patients was done during laparotomy, which was also consistent with conducted studies [10]. If the patient has

no symptoms and no underlying disease, there is no need to treat.

To treat the underlying disease, oxygen therapy and antibiotics are recommended for symptomatic cases. If symptoms are severe, surgery is recommended [6]. It should be noted that observing free air within peritoneum of any patient who has not been recently under laparotomy or laparoscopy, indicates rupture of one of hollow viscera and suggests the need for emergency surgery. The only exception is PCI which in most cases does not require surgery, despite the air under the diaphragm [6-8].

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