Summary

Background: We report a rare condition of Pneumatosis cystoides intestinalis with hepatic portal venous gas in a 63-year-old man with good prognosis.

Case report: A 63-year-old man presented with fever, shaking chills, headache and malaise from three days before admission. The blood pressure was 130/80 mmHg; body temperature was 39.2° centigrade; respiratory rate was 18 per minute, and pulse rate was 100 per minute. Physical examination revealed a mildly icteric sclera, Leukocyte count was 3800 per µl, hematocrit was 38%, and in urinalysis Bilirubin was +3 positive total Bilirubin was 5.7 mg/dL, Bilirubin direct: 3.5 mg. Blood culture was positive for E. coli. The patient was given high-pressure oxygen, and antibiotic therapy. Fever was diminished and clinical status was better. The patient was discharged one week after therapy, while CT-scan showed no air in the portal venous system and intestinal gas-filled cysts was decreased.

Conclusions: Many studies showed that Pneumatosis cystoides intestinalis alone can be seen in benign conditions. But Pneumatosis cystoides intestinalis with hepatic portal venous gas are signs of severe problems in abdomen with poor prognosis. We report a rare case of this condition with good outcome.

Key words: Pneumatosis cystoides intestinalis • hepatic portal venous gas

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**BACKGROUND**

Pneumatosis cystoides intestinalis (PCI) is a rare condition characterized by multiple, thin walled, noncommunicating, air filled cysts. They have no epithelial lining and are located in the wall of the small or large intestine or both. Other areas may be involved, including the stomach, duodenum, and extraintestinal structures (mesentery, lymph nodes, omentum, and peritoneum) but this is less common [1]. It was first described by Du Vernoi (1730), and the term was coined by Mayer in 1925. Usually PCI will be discovered incidentally in asymptomatic or mildly symptomatic patient and follows a benign course. Sometimes it is associated with fulminant illness including bowel infarction, pseudomembranous enterocolitis or necrotizing enterocolitis. PCI can be classified as either primary (idiopathic) or secondary. The secondary form can be associated with conditions including chronic obstructive pulmonary disease and intestinal obstruction. It has also been reported in patients with Crohn’s disease, especially in those who receive steroids [2]. In Koss’s report (1952), only 15% of cases were idiopathic with a male predominance of 3.5:1 [1]. However, more recent reports state an equal male to female ratio. The most frequent symptoms are diarrhea, vague abdominal discomfort, and abdominal distension and occasionally, hematochezia, mucus per rectum, and weight loss may occur. Idiopathic type of PCI tends to involve the left colon whereas secondary cases mostly involve the small bowel and ascending colon. Complications, including: volvulus, pneumoperitoneum, intestinal obstruction, intussusceptions, hemorrhage, and intestinal perforation, occur in 3% of cases. PCI has been considered as one of the cause of prolonged recurrent asymptomatic pneumoperitoneum [3]. Diagnosis is mostly made on plain abdominal radiograph by presence of linear, curvilinear, or cystic luencies in the bowel wall. Pneumoperitoneum or retroperitoneal air may also be seen. Barium studies may help to confirm the site of bowel wall involvement. Abdominal CT scan is more sensitive to detect intramural gas [4]. CT scan is also capable of detecting portal or mesenteric venous gas, which when associated with pneumatosis, indicates bowel infarction. On endoscopy, PCI appears in the form of multiple, round, soft, pale to bluish polypoid masses in the bowel lumen.

**CASE REPORT**

A 63-year-old man presented with fever, shaking chills, headache and malaise from three days before admission. Nausea, vomiting, abdominal pain, urinary symptoms and difficulty on defecation were absent. No urinary, respiratory and cardiac symptoms were observed. Family history was negative, and there was no history of alcohol consumption, smoking, abdominal surgery or sphinctrotomy. The blood pressure was 130/80 mmHg; body temperature was 39.2° centigrade; respiratory rate was 18 per minute, and pulse rate was 100 per minute. Physical examination revealed a mildly icteric sclera and normal respiratory and cardiac finding. The abdomen was soft and no mass or tenderness was detected. The rectal examination was normal.

Laboratory findings were as follows: blood urea nitrogen, creatinine, arterial blood gas, and alkaline phosphatase level were all in normal limits. Leukocyte count was 3800 per µl, hematocrit was 38%, and in urinalysis Bilirubin was +3 positive.

Total Bilirubin was 5.7 mg/dL, (Bilirubin direct: 3.5 mg), ALT: 89(NL: 10-40 IU), and AST: 95(NL: 10-40).

Plain abdominal radiography showed elevation of the right hemi diaphragm. Echocardiography was normal, and liver ultrasound showed echogenic regions with
gas-filled areas. Abdominal CT scan showed large amounts of hepatic portal venous gas (HPVG) (figure 1), a small amount of fluid in the right pleural space and air was present in the small bowel and ileum wall (figure 2). Because of toxic appearance and fever, three times blood culture was taken and antibiotic treatment (gentamicin, metronidazole and ceftriaxone) was started. Surgical consultation was performed for roll out of acute abdomen, and intestinal infarction in first day. No surgical problem was detected. By these imaging findings and roll out of surgical problem, the diagnosis of PCI and HPVG was established. The patient was given high-pressure oxygen, and antibiotic therapy was continued. The patient was discharged one week after therapy, while CT-scan showed no air in the portal venous system and intestinal gas-filled cysts had decreased by 80%. After 3 months follow up, the patient was in satisfactory condition and had normal hematological and liver function tests.

**DISCUSSION**

The patient is a 63 years old man without any previous medical problem and gastrointestinal surgery, which admitted with diagnosis of PCI and HPVG. No any surgical or non-surgical causes were detected. The patient improved with medical treatment.

The secondary form of PCI is seen in COPD, Crohn’s disease, scleroderma, lupus, lymphoma Whipple’s disease, tuberculosis, and AIDS [2,5]. It mainly occurs in the 30-50 years age group [1,2,6]. PCI and HPVG strongly suggest intestinal necrosis or severe systemic disease, which, of course, was not seen in the above patient. However, according to the study performed by Chavarri, 0.3% of patients had history of abdominal surgery [7] and the patient presented by Ibarra, this condition was diagnosed after glairo-sanguinolent diarrhea [8]. In a 60-year-old dialysis patient, pneumothorax and HPVG was reported and laparotomy showed no signs in favor of bowel ischemia [7]. According to Monnæuse’s study, HPVG has poor prognosis (mortality rate 75%-90%), which was associated with intestinal necrosis [9]. An 82-year-old female, who although had HPVG and PCI, had no signs in favor of bowel ischemia HPVG was seen in a 63-year-old patient who had cholangitis after pancreaticoduodenectomy [10]. Similar conditions were seen following CPR [11]. Wiesner present 8 patients with different non-ischemic etiologies for portal-venous gas.

The etiologies for portal-venous gas included: abdominal trauma; large gastric cancer; prior gastroscopic biopsy; prior hemicolecotomy; graft-vs-host reaction.

large paracolic abscess; mesenteric recurrence of ovarian cancer [12]. Gilbeau reported three cases presented with mesenteric and portal venous gas secondary to mesenteric infarction [13]. An 82-year-old female, who although had HPVG and PCI, had no signs in favor of bowel ischemia [14]. HPVG tends to disseminate peripherally in the liver until it reaches a distance 2cm from the capsule and is usually accompanied by bowel infarction [15]. This condition is accompanied by fluid in the abdomen, bowel pneumatosis, paralytic ileus, diverticulitis, cholangitis, and thrombophlebitis of portal vein, hepatic artery embolization, after colonoscopy and even barium enema. As a whole, it seems that PCI and HPVG is secondary to air embolization and increased intra-intestinal pressure and gas formation by anaerobic bacteria. In addition, it has been reported in asthma [16] and polymyositis [17]. All of the above studies showed that PCI alone can were occurred in benign conditions. But PCI with HPVG is in favor of severe problems in abdomen with poor prognosis. It is unusual for a patient with acute abdomen, HPVG and PCI to survive. Our case with diagnosis of HPVG with PCI who responded very well to medical therapy is a rare condition with good outcome.

Sonography and CT scan can be used for diagnosis of PCI+HPVG . As previously mentioned, primary diagnosis is based on plain abdominal radiography and CT-scan. In this way, CT-scan have capability to detect even small amounts of air and distinguishing air present in the portal vein from pneumoboli [18]. However, some trials have been made to use ultrasoundography in diagnosis of HPVG are being conducted [19-21]. The treatment of PCI includes high-pressure oxygen and broad-spectrum antibiotics therapy. In patients with asymptomatic PCI a specific therapy has not been established. Symptomatic patients should receive 55%-75% oxygen for 4 to 10 days until the Po2 concentration reaches 200 to 300 mmHg [22].

**CONCLUSIONS**

PCI alone may be seen in benign conditions. But PCI with HPVG are signs of severe problems in abdomen with poor prognosis. We report a rare case of this condition without any primary abdominal problem but with E.coli septicemia, with good outcome. For this reason PCI with HPVG are not seen always with poor prognosis.
Case Report

REFERENCES