CASE REPORT

PNEUMATOSIS CYSTOIDES INTESTINALIS PRESENTING WITH GALL BLADDER DISEASE: AN INCIDENTAL RARE FINDING

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ABSTRACT: Pneumatosis cystoides intestinalis is a rare disease characterized by multiple, multilocular air filled cysts, within submucosa and subserosa of the gastrointestinal tract. PCI is not a disease in itself, but a radiological sign of an underlying problem. The pathogenesis is still unclear. Patients rarely experience symptoms secondary to cysts and often disease is diagnosed incidentally. The treatment ranges from masterly inactivity to immediate surgery. This article presents here, a case of pneumatosis cystoides intestinalis, incidentally detected on laparoscopy, along with symptoms and signs, operative and perioperative management, as well as a literature review of this clinically important entity.

KEYWORDS: Pneumatosis cystoides intestinalis, gall bladder disease, laparoscopy.

INTRODUCTION: Pneumatosis cystoides intestinalis (PCI) is defined as air filled cysts within the wall of the gastrointestinal tract.¹ Duvernoi first documented PCI in 1730 as postmortem finding.² In 1876, Bang described histopathological changes while Hahn in 1899 described PCI in living patients. Baumann-Schenker in 1939 made first radiological preoperative diagnosis of PCI.² The term pneumatosis cystoides intestinalis was coined by Meyer. The incidence of PCI is unknown. Maximum patients are affected in sixth decade. Primary PCI is extremely rare and diagnosed incidentally.

Secondary PCI is due to an underlying disease or condition eg inflammatory and autoimmune diseases, infectious disease, traumatic and mechanical, drug induced, transplantation, immunosuppression or neoplasm.³ Though any part of gastrointestinal tract may be affected; small bowel is involved in 42%, colon in 36% or both in 22% of cases.⁴ Presented here is a case of primary pneumatosis cystoides intestinalis detected in patient of gall bladder diseases who previously had no symptoms related to PCI and was successfully cured after removal of gall bladder pathology.

CASE REPORT: 28 years male, nonsmoker, nonalcoholic presented with complaints of central and right upper abdominal pain, post prandial fullness, nausea, belching and heartburn of 4 month duration. He had total 3 episodes of pain in last 4 months. The pain was insidious onset, off and on, with radiation to back, more on taking meals and was partially relieved by medications (antacids). There was no fever, no bowel/ bladder complaints, no chest complaints, no previous hospitalization and no history of any skin rash.

On examination, the patient had pallor, was anicteric with no lymphadenopathy, skin rash or edema feet. Abdominal examination revealed tenderness in right hypochondrium & epigastrium. Rest of the abdomen was soft

Investigations revealed hemoglobin 10.8 gm%, total leucocyte count 10,800 cells/mm³ and normal ESR. Kidney Function test was within normal limits. Chest X ray was normal. X ray Abdomen showed bowel gas shadow. USG suggested dilated gall bladder with a gall bladder polyp of size 2.0 cm

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with normal gall bladder wall thickness. Considering the size of polyp and previous symptomatic episodes of patients, laparoscopic cholecystectomy was planned.

Intraoperatively, multiple subserosal cysts were seen in small bowel wall (jejunum and ileum) [Fig. 1]. Cyst was also seen in adjacent mesentery [Fig. 2]. Stomach and colonic wall appeared normal. The cyst was left undisturbed and laparoscopic cholecystectomy was performed. The patient had an uneventful recovery. He was started orally next day.

Postoperatively, a search for cause of pneumatosis cystoides intestinalis was initiated. An upper GI endoscopy was performed on 5th day which revealed normal stomach and duodenum. The patient was retrospectively asked for any previous history of skin rash or chest discomfort anytime but had none. The patient was discharged 7th postoperative day. The histopathological report suggested chronic cholecystitis with gall bladder polyp of size 2.1 X 1.5 cm, sessile, adenomatous with no gall stones [Fig. 3]. The final diagnosis made was gall bladder polyp with chronic cholecystitis with primary pneumatosis cystoides intestinalis. The patient had follow up thrice with total relief of symptoms.

CASE DISCUSSION: Pneumatosis cystoides intestinalis is rare radiological sign characterized by multiple, multilocular, cystic or linear, thin walled collections of gas in wall of gastrointestinal tract.

Cysts are usually subcentric, located in subserosa, submucosa & rarely muscularis layer and are lined by mixed inflammatory cells, macrophages, or foreign body giant cells. The subserous cysts are most frequent in small bowel while submucous cysts are predominant in colonic wall. Gastric PI (air within wall of stomach) is uncommon.⁵

The pathogenesis of pneumatosis cystoides intestinalis is unclear, though various theories have attempted to explain exact origin. PCI may be primary (15% cases), with no associated predisposing factors, and is often detected incidentally on radiography, endoscopy or laparoscopy.⁶ It may be secondary (85% cases) to gastrointestinal disorders (obstructive or necrotic), obstructive pulmonary disorders, infectious diseases, drugs and chemotherapeutic agents.⁶

The mechanical theory proposes that intraluminal gas, under increased intra-luminal and intra-interstitial pressure, dissects into bowel-wall through a mucosal defect. This is most accepted explanation. The mucosal injury may be caused by inflammatory process (diverticulitis, necrotizing enterocolitis) or mechanical damage (gastric ulcer, duodenal ulcer, post-surgical anastomosis) or autoimmune and connective tissue diseases and immunosuppression (steroid & cytotoxic medical therapy in post transplantation, HIV & neoplasm). The intraluminal pressure also increases in ileus, upper GI endoscopy & colonoscopy.⁷

The bacterial theory proposes entry of gas forming organism into bowel wall by direct invasion. Disappearance of PCI during antimicrobial treatment supports this view. However, bacteria are not isolated within cysts.⁸

The pulmonary theory is least accepted explanation. The gas travels from ruptured alveoli, through mediastinum, along vessels into retroperitoneal space and then, through mesentery into bowel wall. It explains association of PCI with respiratory conditions eg chronic bronchitis, emphysema, pneumothorax and mechanical ventilation.⁹

The chemical theory suggests that alpha- glucosidase inhibitor therapy for diabetes, inhibits carbohydrate absorption, thus increases intestinal gas production and causes PCI.¹⁰

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When neglected, intramural gas drains into mesenteric veins and subsequently portal venous system. Hepatic portal venous gas (HPVG) indicates grave prognosis, and has mortality rate of 75–85% and is associated with mesenteric ischemia. The pathogenesis of HPVG is not fully understood.¹¹

PCI affects male in sixth decade. Only 8% cases occur below 20 years.¹² Primary pneumatosis is often asymptomatic or has non-specific symptoms eg diarrhea or constipation, bloody stools/mucus discharge, abdominal pain, abdominal distention (meteorism), weight loss and tenesmus. Cyst size has no correlation with clinical manifestations.⁶

Diagnosis of PCI requires high index of suspicion. Plain X ray abdomen shows circular or linear, radiolucent cysts within bowel wall or mesentery in 66% cases (Jamart).¹²

Barium enema shows intraluminal bulging resembling polyps and colonoscopy depicts smooth, submucosal, tumor like protrusions. An attempt to puncture may rupture cyst and cause bowel perforation. CT, being highly sensitive, precisely delineates extent of lesions and differentiates PCI from adenomatous polyp or malignancy.⁹

The treatment of PI ranges from benign disease, (not requiring treatment, 85% cases) to mild disease to life-threatening condition (requiring immediate surgery). Gas cysts persist for long, are gradually absorbed and resolves spontaneously (in 50% patients). Recurrence is common.¹¹

In mild disease, inhalation of 70% humidified oxygen with PO₂ of 250 mmHg for 5 days and hyperbaric oxygen therapy at 2.5 atm (252.5 kPa) for 150 min on three successive days causes resolution of gas collections.¹³ Intestinal rest, total parenteral nutrition, octetide, prokinetic drugs and intravenous antibiotic therapy (Flagyl 600-1600 mg/ day) are other treatment modalities. There is complete recovery in mild disease. Endoscopic puncture and cysts sclerotherapy has also been tried.¹⁴

The indications of surgery are peritonitis, malignancy, signs of ischaemia, severe inflammation/sepsis, metabolic acidosis and portal venous gas. The portal venous gas has high mortality of 37% and is usually associated with mesenteric ischaemia.¹⁵

Complications occur in 3% of cases, are associated with poor prognosis and include obstruction, intussusception, volvulus, hemorrhage, intestinal perforation, pneumoperitoneum and pneumoretroperitoneum.¹³

CONCLUSION: Pneumatosis cystoides intestinalis is a rare disease, mostly asymptomatic and suspicion is based on imaging and clinical finding. It indicates undiagnosed underlying disease. PCI is managed conservatively in most cases. Radiographic pneumoperitoneum may be due to ruptured cysts and not necessarily perforation. Laparoscopy is a useful aid. PCI can be easily missed without proper clinical vigilance and therefore, educating physicians prevents misdiagnosis and unnecessary surgery.

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Figure 1: Showing multiple, subserous, air filled cysts in the small bowel depicting pneumatosis cystoides intestinalis.

Figure 1

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Figure 2: Showing multiple, air filled cysts in the mesentery of the small bowel.

Figure 2

Figure 3: Showing excised gall bladder adenomatous polyp, that was producing symptoms and led to incidental diagnosis of pneumatosis cystoides intestinalis.

Figure 3

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