**Pneumatosis cystoides Intestinalis Lipomatosis-A Case Report**

Kafil Akhtar*, Prasenjit S Ray, Anjum Ara, Sadaf Haiyat and Rana K Sherwani

Department of Pathology, Jawaharlal Nehru Medical College, Aligarh Muslim University, Aligarh, India

*Corresponding author

**KEYWORDS**

**ABSTRACT**

*Pneumatosis cystoides* intestinalis is a rare gastrointestinal condition, in the majority of cases an incidental finding which represents a differential diagnostic challenge for endoscopist, radiologist, pathologist and physician. It is usually associated with a wide variety of gastrointestinal or pulmonary diseases. Considering the relative rarity of this condition, it is frequently overlooked. Even if it is correctly diagnosed, its clinical relevance is often misinterpreted. The mechanical theory, which is the most accepted explanation, postulates that gas is forced into the bowel wall by breaks in the mucosa; this is more likely to occur when the intraluminal pressure is higher, as happens in obstructive conditions, during endoscopies, or during infections from gas-forming bacteria.

**Introduction**

*Pneumatosis cystoides* intestinalis with colonic lipomatosis is a rare entity associated with a number of gastrointestinal and systemic disorders. The hallmark of this condition is the presence of multiple gas-filled cysts along the bowel wall (Cyrany et al., 2005). Considering the relative rarity of this condition, it is frequently overlooked. Even if it is correctly diagnosed, its clinical relevance is often misinterpreted. However, keeping in mind of the fulminant diseases that may underlie it, a high index of suspicion is advocated, as an early diagnosis can significantly affect the outcome in the patient (Cyrany et al., 2005; Gagliardi et al., 1996). We report the case of a 45 years old male patient who presented with clinical features of acute intestinal obstruction and was histopathologically confirmed to have PCI with incidental finding of multiple lipomas involving his left colon. A brief review of relevant literature is also undertaken.

**Case report**

A 45 years old Indian male presented with abdominal pain and vomiting for 2 days. He
also had a history of constipation with intermittent passage of mucous in stool over last 6 months. Clinical examination revealed that the patient was dehydrated and his abdomen was distended, mildly tender to palpation and tympanic on percussion. A provisional diagnosis of bowel obstruction was made and the patient was admitted for further evaluation. The CT showed a large number of small gas-filled cysts arising from the wall of the colon. Colonoscopy revealed multiple submucosal sessile polypoid lesions covered with normal mucosa in the colon. CT colonography identified multiple submucosal gas cysts mostly located in the left colon.

Laboratory workup results were within normal limits except a raised total leucocyte count (12,500/mm$^3$) with neutrophilia (differential count N82, L16, M0, B0) and dyselectrolytemia (Na$^+$=130 mEq/L, K$^+$=3.1 mEq/L). Plain abdominal X-rays in erect posture showed multiple foci of localized gas collection in the wall of left hemicolon; no free gas was present under the domes of the diaphragm.

This picture led us to a suspicion of Pneumatosis cystoides intestinalis. The patient was initially managed conservatively with nasogastric suction, intravenous fluids and parenteral antimicrobials. However, since his condition did not show signs of improvement, explorative laparotomy was undertaken and a 30 cm long segment of the distal transverse colon and descending colon was resected.

On gross examination, multiple subserosal and submucosal cystic lesions of varying size (0.5–1.5 cm) were observed in the resected colon specimen. Crepitus could be elicited in the cysts on palpation and they collapsed on incision. The size of the polyps ranged from 0.5–1 cm in diameter and their cut section was solid, homogeneous and yellow in colour.

Microscopic examination revealed cystic spaces devoid of epithelial lining, predominantly in the submucosa and subserosa associated with considerable inflammation of the adjacent gut wall (Figure 1).

On the other hand, the sessile polyps in the submucosa were composed entirely of mature adipose tissue and covered by normal mucosa of the colon (Figure 2).

Thus a diagnosis of Pneumatosis cystoides intestinalis with colonic lipomatosis was made. The post-operative period remained uneventful and the patient is in regular follow up without any signs of recurrence.

**Discussion**

The earliest documentation of Pneumatosis cystoides intestinalis (PCI) dates back to eighteenth century when this entity was described by Du Vernoy from autopsy studies (Cyrany et al., 2005). The condition can affect patients irrespective of age, although there is a bimodal peak in infancy and mid adult age (30–50 years) (Gagliardi et al., 1996; Warner, 2005). Gagliardi et al. (1996) studied 25 cases of PCI in adults and reported a higher incidence in men as compared to women (male: female = 1.5:1).

PCI is characterized by multiple, thin walled, non-communicating, air filled cystic spaces located in the wall of the small or large intestine or both. Other areas may be involved, including the stomach, duodenum, and rarely extra intestinal structures (mesentery, lymph nodes, omentum, and peritoneum) (Cyrany et al., 2005; Gagliardi et al., 1996; Piazza and Stoll, 2007). PCI
involving the colon is also referred to as *Pneumatosis cystoides coli*.

Based on the etiology, PCI has arbitrarily been classified as primary – a benign idiopathic form (15% cases), and secondary – when PCI occurs as a consequence of other underlying diseases (85% cases). Of these, the former mainly involves colon while the latter tend to occur in small intestine (Deshpande et al., 2003). In infants, PCI is almost invariably associated with necrotizing enterocolitis and pursues a fulminant course with a mortality rate of 20–30%. Common predisposing factors include prematurity, low birth weight, aggressive enteral feeding and gut infection by a host of bacterial and viral agents (viz. *Escherichia coli*, *Clostridium perfringens*, *Klebsiella* and *Rotavirus*) (Cyrany et al., 2005; Warner, 2005). In addition to these infectious agents, common underlying conditions leading to PCI in adults are blunt trauma abdomen, ulcerative colitis, systemic lupus erythematosus, dermatomyositis, volvulus, intestinal infarction, amyloidosis, chronic obstructive pulmonary disease, graft-versus-host disease and immunosuppression. Drugs (lactulose, glucocorticoids, chemotherapeutic agents, nonsteroidal anti-inflammatory drugs) and malignancies (cholangiocarcinoma, adenocarcinoma of bowel, Hodgkin’s disease, leukemia) have also been implicated in PCI of adults (Cyrany et al., 2005; Gagliardi et al., 1996; Piazza and Stoll, 2007). Thus the differential diagnosis of PCI include a wide plethora of diseased conditions.

Patients with *Pneumatosis cystoides intestinalis* are frequently asymptomatic or present with diarrhea, constipation, hematochezia, passage of mucus per rectum, vague abdominal discomfort, abdominal pain, distension, excessive flatulence, malabsorption and weight loss. Additional symptoms in infants include lethargy and temperature instability. Depending on the location of the gas filled cysts and the etiology, the range of symptoms in each patient may vary enormously. If the cysts rupture, patients exhibit features of pneumoperitoneum and/or peritonitis (Piazza and Stoll, 2007; Antosz and Zaniewski, 2004; Reynolds et al., 1991).

Pathological findings in PCI are distended bowel loops studded with cysts of varying size (0.5–10 centimeters) and containing air. Subserosal cysts with thinned out walls are indicative of impending perforation. In presence of associated ischemia or infarction, the gut wall may show variable discolouration. Histopathologically, these cystic spaces are devoid of epithelial lining and hence in true sense are pseudocysts. They involve predominantly the submucosa and/or subserosa, and are usually not

---

connected with the gut lumen. Mucosal lining is often distorted with crypt dialatation, cryptitis, crypt abscess and considerable inflammation in the submucosa, muscularis and serosa. Presence of giant cells along the cystic spaces, submucosal microabceseses, granulomas and coagulative necrosis are other notable findings (Cyrany et al., 2005; Piazza and Stoll, 2007; Ellis, 1980).

Colonic lipomatosis is an uncommon benign condition. Submucosal lipomas found in this situation need to be differentiated from colonic pseudolipomatosis. The latter, often associated with PCI, show empty vacuolated spaces in the lamina propria and this feature mimics lipoma. The condition has been attributed to extravasation of lymph or barotrauma, although exact pathogenesis is yet to be explained. The fact that these spaces are not composed of adipose tissue can be confirmed by their negative staining with fat stains (e.g. Sudan black) (Rennenberg et al., 2002).

Radiological evaluation serves as a handy tool in identifying patients with PCI. Plain radiographs of abdomen, barium follow-throughs and ultrasonography have been used to detect this condition with varying sensitivity and specificity. Computed Tomography is the most sensitive investigation to diagnose PCI and its complications. Presence of ‘cushion like’ consistency on endoscopy is highly suggestive of submucosal PCI which often project into the gut lumen as polypoidal lesions (Deshpande et al., 2003; Reynolds Jr et al., 1991). On the other hand, endoscopic appearance of lipomatosis bears close resemblance to familial polyposis due to the presence of multiple sessile polyps (Pieterse et al., 1985).

Management of patients with PCI depends on the clinical scenario and the underlying cause. Asymptomatic patients usually do not require any treatment while those with acute abdomen benefit from immediate surgical intervention coupled with antimicrobial coverage. Thus, management should be tailor made on an individual basis. Overall, adults with PCI have a benign course with a better outcome. Infants, on the contrary, tend to have a rapid downhill course frequently culminating in sepsis, shock and death. Case fatality rate in infants continues to be 20-30% despite early institution of therapy (Warner, 2005; Antosz and Zaniewski, 2004; St. Peter et al., 2003).

**Figure 1** Cystic spaces devoid of epithelial lining, predominantly in the submucosa and subserosa associated with considerable inflammation of the adjacent gut wall
In conclusion, *Pneumatosis cystoides intestinalis* often presents a diagnostic challenge for the clinician, radiologist and pathologist. The fact that needs to be emphasized is PCI is not a disease in itself but a manifestation of a host of underlying disorders. This merits its timely recognition, as that can aid in choosing the appropriate treatment modality and significantly affect the outcome in the patient. Colonic lipomatosis is a benign condition that mimics familial adenomatous polyposis grossly. Polypectomy followed by histopathological examination can distinguish lipomatosis from familial adenomatous polyposis and unnecessary surgery can be avoided.

**References**


