

Case report

Pneumatosis Cystoides Intestinalis complicating Ulcerative Colitis

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Abstract:

Pneumatosis cystoides intestinalis (PCI) is a disease in which small gas-filled cysts appear in the intestinal wall. Its pathophysiology and clinical relevance are poorly understood. A 60 year old lady presented with acute abdomen with features suggestive of intestinal obstruction. The intra operative impression was Intestinal Lymphangioma. The histological diagnosis was Pneumatosis Cystoides Intestinalis complicating Ulcerative Colitis.

Key words: Pneumatosis cystoides intestinalis (PCI), ulcerative colitis.

Introduction:

Pneumatosis cystoides intestinalis is an uncommon condition characterized by gas filled cysts within the bowel wall [1]. In the literature there is little information about it's incidence [2]. The first description of pneumatosis intestinalis was by DuVeroni in 1730; it was first defined as the presence of gas in an abnormal location in the body [3]. PCI is characterized by multiple, thin walled, non-communicating, 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 extra intestinal structures (mesentery, lymph nodes, omentum, and peritoneum) but this is less common [4]. PCI is considered a finding of equivocal

importance, diagnosed through radiography, endoscopy or pathological examination [5].

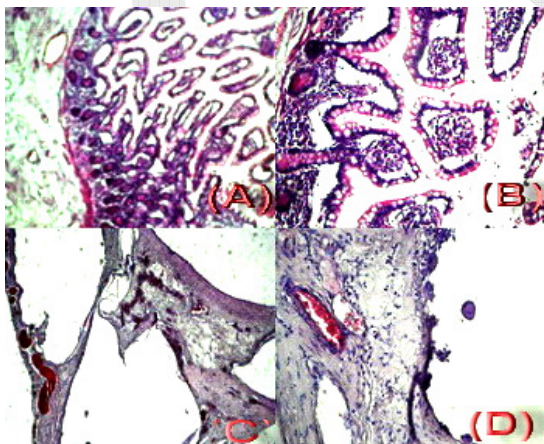
Case report:

We present a case of a 60-year old female who was admitted to Emergency Department, at Damanhour national Medical Institute. The patient presented with severe abdominal pain of short onset. On clinical examination, the abdomen was grossly distended and markedly tender on deep palpation. There was history of repeated diarrhea and bloody stools and the patient was on treatment for inflammatory bowel disease. The routine laboratory tests, including serum blood count, electrolytes and liver biochemistry were all within normal limits. The physical and the laboratory findings didn't help to reach diagnosis. The plain X-ray showed massive peumo-peritoneium as well as large amount of intramural gas within small and large bowel. A provisional diagnosis of bowel obstruction was made. The operative decision was right hemicolectomy resection and anastomosis. The intra operative impression was intestinal lymphangioma. Grossly, the resected specimen measured 50 cm in length. Along the entire length of the specimen, on the outer surface of the intestinal loop, multiple cysts formations of various sizes could be seen; The cysts contained air. The intestinal lumen was seen apparently normal [Figure:1].



Figure: 1) Intestinal loop presented with multiple cysts formations

Histopathologic study of the resected specimen revealed edematous mucosa showing prominent changes including diffuse mononuclear inflammatory infiltrate in lamina propria as well as crypt abscesses formations; plasma cells were common at the base of crypts; suggesting ulcerative colitis. The histology of the cysts showed flattened macrophages lining the cysts and multinucleated giant cells were commonly found, all suggestive of an inflammatory process [Figure: 2]. The histological diagnosis was Pneumatosis Cystoides Intestinalis (PCI) complicating ulcerative colitis.



(Figure: 2) Intestinal mucosal edema and hyperplasia (A) with marked crypt abscesses formations (B) multiple cysts formation (C) the macrophages are seen lining the cysts as well as multinucleated giant cells (D)

Discussion:

PCI occurs in two forms; primary pneumatosis intestinalis (15% of cases) is a benign idiopathic condition in which multiple thin-walled cysts form in the submucosa or subserosa of the colon. This primary form is often termed pneumatosis cystoides intestinalis. The secondary form (85% of cases) is associated with obstructive pulmonary disease, as well as obstructive and necrotic gastrointestinal disease [6].

The exact origin of PCI is debated. Many theories have been advocated. First, there is the mechanical theory. It suggests that gas under pressure is forced into the bowel wall through a mucosal defect. It is probably involved in PCI associated with trauma, surgery, endoscopy and in cases that involve bowel obstruction. Second, there is the bacterial theory [2]. Mucosal disruption may be caused by trauma, obstruction, ischemia, or inflammation to the bowel wall, which allows bacteria or gas to invade the bowel wall. A bacterial origin of the gas is suggested by the high concentration of hydrogen within the cysts. Steroids, immunosuppressants, and chemotherapeutic agents are also thought to lead to pneumatosis through increased mucosal permeability; the mechanical and bacterial theories of pathogenesis have come to be most widely accepted. The mechanical theory postulates that compressed air within the intestinal lumen penetrates the mucosa through microscopic or macroscopic breaks, dissects down along the mesentery, and reenters the intestinal lumen distally along small vascular and lymphatic channels [7].

The bacterial theory proposes that gas-forming bacilli enter the submucosa through breaks in the mucosa and produce gas in the intestinal wall. However, repeated attempts to culture organisms from the cysts have been unsuccessful [6 & 7].

Conclusion:

Pneumatosis cystoides intestinalis is a rare clinical entity, understanding of the causes and

associations of this finding will help physicians make appropriate decisions about surgical, medical or conservative management.

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