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Pneumatosis Cystoides Intestinalis of Small Bowel: Rare Disease Mimicking Acute Abdomen

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Abstract: ***Pneumatosis cystoides intestinalis (PCI) is a low incidence disease characterized by the presence of multiple gas-filled cysts within the submucosa or subserosa of the small intestinal wall. It occurs in approximately 0.03% of population. Its pathogenesis is uncertain and several pathogenic mechanisms have been proposed to explain its origin such as Crohn's disease, intestinal stenosis, ulcerative colitis, drug use, extra-gastrointestinal diseases and chronic obstructive pulmonary disease. Most consider its pathogenesis interrelated to an increase in intra-intestinal pressure and the accumulation of gas produced by aerogenic bacteria. In this study, we present the case of acute abdomen who underwent exploratory laparotomy and who had pneumatosis cystoides intestinalis determined histologically.***

Keywords: Pneumatosis cystoides intestinalis (PCI), small bowel, hollow viscus perforation, submucosal empty cysts

1.Introduction

Pneumatosis cystoides intestinalis (PCI) also termed pneumatosis intestinalis, pneumatosis coli (when occurring in colon only) is an uncommon condition characterized by gas in the bowel wall" (1)". Air-filled bubble-like lesions are located in the submucosa or the subserosa of the gastrointestinal tract. Although the large bowel may also be involved" (2)". Exact prevalence is not known as asymptomatic course in majority of cases may lead to under estimation" (3)". It can affect any age group, although it is more prevalent in older population than in young adults and infants. Infantile PCI has higher mortality than adult forms, and it is usually associated with acute necrotizing enterocolitis.

The first pathologic description of PCI has been attributed to Du Vernoy, a French pathologist who described it during an autopsy dissection in 1730. Subsequently it was named by Mayer as Cystoides intestinal pneumatosis in 1825 and Gazin et al. described the first clinical report in a patient who underwent surgery in 1946" (4)".

It has been divided into two forms: Primary PCI is a benign idiopathic condition, while secondary PCI can be associated with various underlying conditions, including necrotising enterocolitis, obstructive lung disease, inflammatory bowel disease, and connective tissue diseases" (5)". In adults, approximately 15% of cases are primary, and 85% are secondary.

Morphologically PCI may occur in two different types: bubble-like (cysts in intestinal wall), typical of the primary type, and band-like (continuous lines), correlated with secondary PCI" (6)".

2.Case Report

An 82 year old man was admitted to the emergency department presented with a 4 day history of abdominal pain, nausea and constipation. The pain was insidious in onset, intermittent and non-radiating. His past history was significant for asthma and under medical control.

During clinical examination, patient was found to have a distended abdomen and generalized tenderness. No organomegaly or masses were detected. Bowel sounds were present and active. On investigations, Chest X-ray revealed gas under diaphragm. A computed tomography (CT) scan showed hollow viscus perforation with pneumoperitoneum, suspicious for posterior wall of the stomach with mild abdomino-pelvic ascitis. Considering the patient's physical examination and radiological findings, the provisional diagnosis of hollow viscus perforation perforation was kept and surgical exploration was performed.
Patient underwent exploratory laparotomy with resection of the ileal segment and end to end anastomosis and specimen was sent to the department of Pathology.

We received a part of small intestine measures 58cm in length. Outer surface (serosal aspect) showed grape like vesicles ranging in size from 0.5 to 1cm in diameter. On cut opening, grapes like vesicles were empty and had crackling sound. (Figure 1).

Microscopic examination of the sections from small intestine with grape like vesiclesrevealed submucosal and subserosal cysts lined by multinucleated giant cells and dense acute and chronic inflammatory cell infiltrate. Ill defined granulomas were also seen scattered in the subserosal region (Figure2a, 2b). Keeping in view these findings diagnosis of pneumatosis cystoides intestinalis was made.

3.Discussion

PCI, refers to gas within the bowel wall, also known as pseudopneumatosis, intestinal emphysema, and bullous emphysema of intestine. It is an uncommon condition. PCI has recently come to increased clinical attention due to improved radiographic identification. Number of males affected with PCI is 3.5-fold higher than that of females" (7)".

PCI can be divided into two types: primary and secondary. The primary or idiopathic type, accounts for about 15% of cases. The secondary type, comprises the remaining 85% of cases. The secondary type is associated to a wide spectrum of predisposing factors. like gastrointestinal diseases, including gastroduodenal ulcers, pyloric stenosis, small bowel obstruction, appendicitis, ischemic or inflammatory intestinal diseases; lung diseases, including asthma or chronic obstructive pulmonary disease (COPD); and autoimmune disease" (5, 6)". Though PCI is distributed throughout the digestive tract, the most common site is the colon (47%); (distal stump of the transverse splenic flexure colon, particularly the descending and sigmoid colon, is most commonly affected), followed by small bowel (27%), stomach (5%), and both colon and small bowel (7%).

The exact etiology remains obscure and PCI is considered to be caused by a myriad of underlying pathophysiological processes that range from benign to life-threatening conditions" (8)". There are three hypotheses of PCI pathogenesis: (1) mechanical theory: involving an increase in intraluminal pressure that causes mechanical damage and mucosal rupture of the intestinal wall, leading to the migration of gas from the gastrointestinal cavity to the intestinal wall; (2) pulmonary theory: chronic lung diseases such as chronic obstructive pulmonary disease, asthma, and interstitial pneumonia lead to alveolar rupture, causing mediastinal emphysema and release of gas along the aorta and mesenteric blood vessels into the intestinal wall; and (3) bacterial theory: aerogenic bacteria penetrate the intestinal mucosal barrier, ferment in the intestinal wall, and produce gas" (9)".

As it has been observed that the cause of pneumatosis intestinalis is not exclusively limited to GI tract pathology. While debatable, it has been suggested that cystic fibrosis, asthma, and other obstructive bronchopathologies such as COPD can cause PCI. This may be due to these entities resulting in chronic cough which increase transabdominal pressure and could thereby cause transmucosal air dissection" (10)". Our patient was a known case of asthma and as per pulmonary theory it may be reasonable to hypothesize a subclinical respiratory disorder that may have contributed to the development of PCI.

In addition, a suggestion that the cystic spaces in PCI are dilated lymphatic vessels has been lingering in literature. In the early days, this condition was also termed lymphopneumatosis cystoides intestinalis or cystic lymphopneumatosis based purely on assumption. In an experiment conducted half a century ago, Staudacher and Bencini injected a dyed fluid into the lymphatics of hogs and demonstrated that the distended lymphatics formed cystic spaces. Some investigators also noticed a spatial relationship of pneumatosis to lymphoid tissue, which is indeed a frequent histologic finding. By light and electron microscopic study in the case of PCI, Haboubi et al claimed to identify some evidence of the cystic spaces being dilated lymphatic vessels in nature" (11)".

CT is the most sensitive tool to localize the PCI and to make differential diagnosis between primary and secondary PCI. Currently there is no consensus on the appropriate management of PCI. Around 50% can be successfully managed conservatively. Majority of idiopathic cases do not require surgical intervention and disappear spontaneously.

PCI is an uncommon entity which can represent a wide spectrum of diseases and a variety of underlying diagnoses. It can be easily confused with intestinal polyp, tumors, inflammatory bowel disease or others even if colonoscopy and radiological investigations are performed due to a lack of awareness and rarity of presentation of PCI. It is not a common finding with little research on this topic.

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Figures

Figure 1: Macroscopic appearance of resected part of small intestine revealing multiple gas filled cysts.

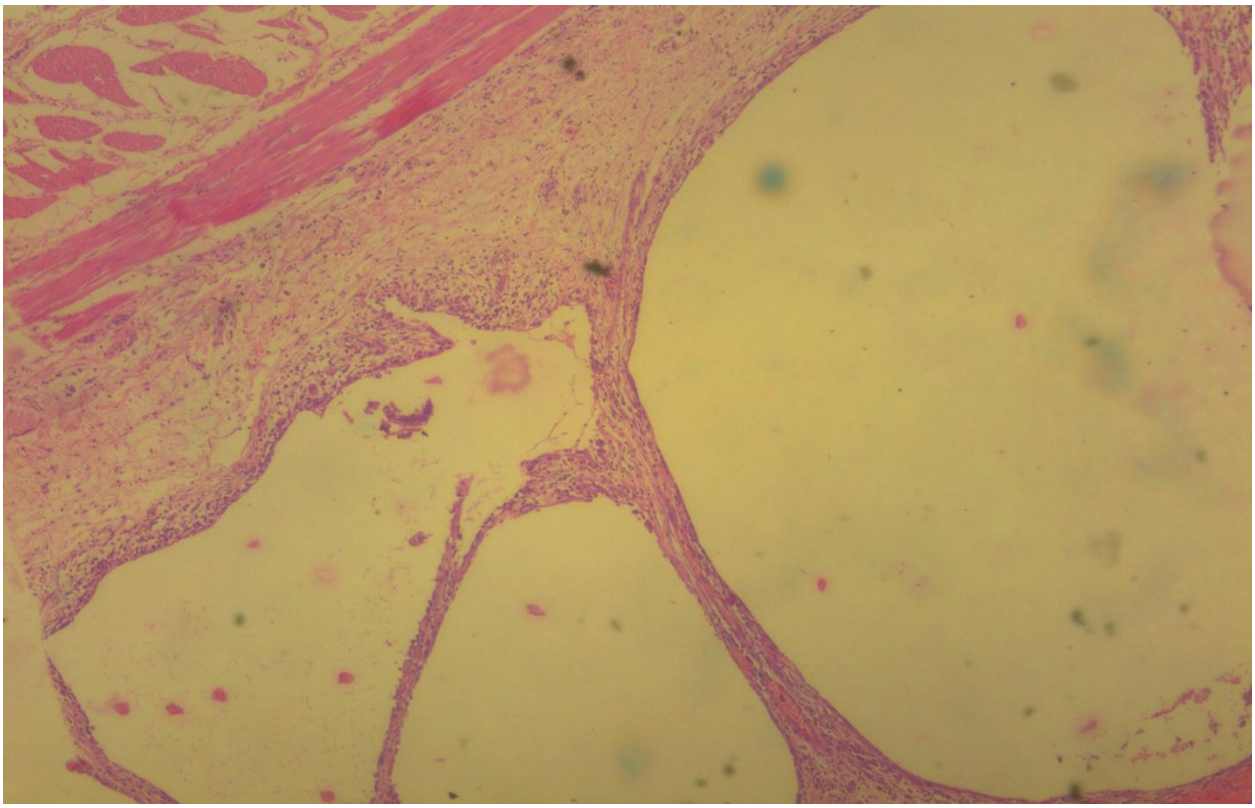

Figure 2a: Photomicrograph showingvariable sized empty submucosal (Haematoxylin and Eosin x10).

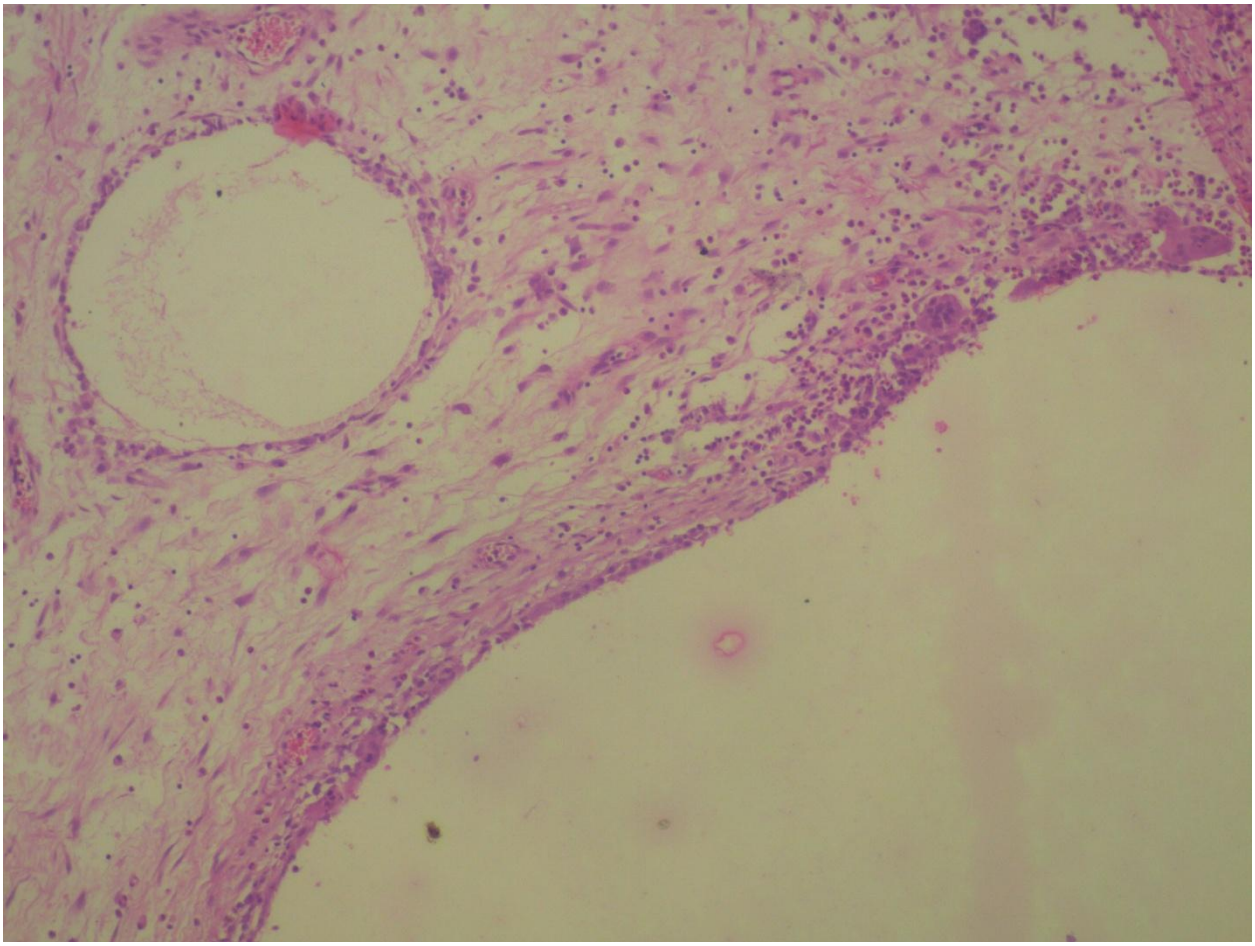

Figure 2b: Photomicrograph showing submucosal cysts lined by multinucleated giant cells (Haematoxylin and Eosin x40)