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PNEUMATOSIS CYSTOIDES INTESTINALIS- DISTINCT AND RARE ENTITY- A CASE REPORT.

Pathology

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ABSTRACT

Pneumatosis cystoides intestinalis (PCI) is a distinct entity occurring in the gastrointestinal wall having multiple submucosal and subserosal pneumocysts. A middle-aged 55 years male, who was a labourer presented with intestinal obstruction. Resected specimen after laparotomy showed multiple submucosal, gas-filled cysts on the entire resected specimen. Microscopy showed a characteristic picture of PCI composed of multiple, submucosal cysts lined by multinucleated giant cells.

KEYWORDS

Pneumatosis cystoides intestinalis, PCI, gas-filled spaces

INTRODUCTION:

Pneumatosis cystoides intestinalis (PCI) is a distinct entity occurring in the gastrointestinal wall having the unique appearance of multiple submucosal and subserosal pneumocysts of varying dimensions.[1,2,3,4,5,6,7,8,9,10] PCI is an extremely rare entity with an incidence of 0.03% till 2018. It was first described by Du Vernoy in autopsy specimens[2,4,8]. It is widely reported in publications. However, we didn't find clinical trials and demographic studies. Mostly the publications consist of case reports, few case series, and very scant literature on systematic review.

Case Report:

The present case was a middle-aged man of 54 years, daily-wage labourer on a causal and informal basis. He threw himself into this tough job for 25 years. He belongs to poverty-ridden strata. He had no significant past medical history.

He was admitted to the hospital with a history of pain in the abdomen since 8 days and abdominal distension for 2 days. The patient didn't have rectal bleeding, vomiting, constipation, fever, or respiratory diseases. Physical examination showed tenderness and distension of the abdomen. Rests of the systems were within normal limits. With these clinical features, the diagnosis of intestinal obstruction was made. Laboratory investigations viz CBC, LFT, KFT, and serum electrolytes were within normal limits. Total serum proteins were low. USG showed dilated bowel loop of 7cm length and was filled with gas. Minimal free fluid around it was noted. CECT confirmed the dilation of 7cm of the large bowel loop. Few of them were showing air fluid levels. The diagnosis of sigmoid volvulus causing obstruction was given. Laparotomy was done and the specimen was sent for histopathological examination.

The specimen consisted of a 32 cm long colonic segment which was dilated in its entirety. On opening the lumen, multiple polypoidal grape-like lesions of variable dimensions were seen diffusely spread all over the segment (figure 01). Multiple bits were taken for microscopic examination. Microscopy (figure 02) showed numerous submucosal (gas-filled) spaces lined by flat and multinucleated giant cells. The overlying mucosa showed minimal inflammation. There was no evidence of chron's disease. The diagnosis of pneumatosis cystoides intestinalis was formulated.

DISCUSSION:

PCI is an uncommon condition characterized by numerous gas-filled cysts in the submucosa and subserosa[1-8]. Male to female ratio is equal with no sex predilection. It can occur at any age[1]. It is seen in the benign and fulminant forms. Fulminant is common in pediatrics and complicates ischemic bowel disease[1].

It is associated with a wide etiological spectrum which causes abnormal accumulation of gas[1-10]. Various hypotheses have been put forth in the causation of PCI.

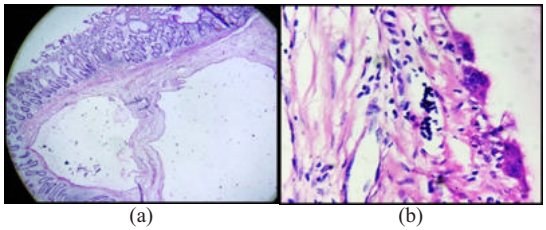
1. Mechanical theory- It is associated with many diseases like inflammatory bowel disease, intestinal obstruction, ischemic bowel disease, gastro-intestinal tumor, ano-rectal surgeries and bowel preparation for surgery. These diseases increase the intraluminal pressure causing mucosal damage and escape of intestinal gas into the damaged or ruptured mucosa[3,4,8].
2. Pulmonary theory: PCI is seen with many diseases like chronic obstructive pulmonary diseases, asthma, and interstitial pneumonia which cause alveolar rupture. Gas from alveoli enters into the mediastinum and travels along the aorta, and mesenteric vessels to the intestine[3,4,8,10].
3. Bacterial theory: Gas producing bacteria crosses the mucosal barrier and produces gas in the bowel wall[3,4,8,10].
4. Chemical and malnutrition theory: It is hypothesized that malnutrition prevents the degradation of carbohydrates causing increased bacterial fermentation leading to large quantities of gas production in the lumen which causes ischemia of surface epithelium. Gas within the lumen enters into the mucosa and submucosa[8]. The role of α glucosidase inhibitors has also been hypothesized. It is thought to cause suppression of carbohydrate degradation and absorption. This generates a large volume of gas breaking through the mucosa[3,8].
5. PCI associated with chemotherapy and hormone therapy has also been reported[8].

The present case is an unskilled daily-wage labourer who waits at the market place to be hired for any type of chore-like lifting or carrying the loads. The duration and intensity of the task are variable but usually long and moderate to severe so that he could fetch the optimal livelihood for the family. These labourers are vulnerable to many health problems like musculoskeletal injuries and gastro-intestinal diseases. Amongst the gastrointestinal problems, it may range from abdominal pain, diarrhea, hemorrhoids, stress on abdominal muscles, gastro-intestinal bleeding, and ischemic injury to the bowel[9]. The authors believe that labourers and persons doing strenuous exercise have a similar negative impact on the gastro-intestinal tract. It depends on the type and intensity of task or exercise, age, physique, nutritional status, diet, accostimization to work, endurance, and tolerance. In the present case, his physically demanding manual work might have shunted the blood from the gastro-intestinal tract to musculo-skeletal system. This might have caused hypoperfusion[11,12]. Costa RJS et al[12] quoted that hypoxia causes injury to the surface of lining epithelium which ends up in dysfunction and necrosis of epithelial cells. The area of necrosis releases cytokines adding to dysfunction. This also causes increased permeability. He further stated that the increased sympathetic activity causes changes in peristalsis. The cumulative effects of all these mechanisms cause gas produced by aerobic bacteria

to enter into submucosa through mucosal rent. The repeated task coupled with other factors had a cumulative effect on the evolution of multiple gas-filled spaces resulting in PCI.

The present case underwent laparotomy for intestinal obstruction and the specimen was sent for a histopathology examination. Gross examination of PCI shows polypoidal grape-like masses protruding through the mucosa. Histopathological examination shows multiple submucosal or subserosal cysts lined by multinucleated giant cells. Mucosa may demonstrate features of cryptitis or crypt abscesses[1,5]. In our case, similar gross and microscopic findings were observed. Hence we faced no difficulty in diagnosis as gross morphology and microscopy were in conformity with PCI. However, we did consider lymphagiomatosis, pseudolipomatosis, diverticulosis, and colitis cystis profunda in differential diagnoses. Lymphagiomatosis show thin vascular channels containing lymph and lined by flattened cells[1,5]. Mucosal pseudolipomatosis characterized by gas filled spaces in lamina propria secondary to gas infusion during endoscopy. Colitis cystitis profunda typically shows intramural mucus containing cysts[1,5]. The microscopy of the present case was non-congruent with the above differential diagnoses. Hence diagnosis of PCI was formulated.

Figure 01- Gross photograph of the colon showing multiple polypoidal grape-like lesions and submucosal cysts of variable sizes.



(a) (b)

Figure 02- a) Microphotograph of colon showing submucosal cysts (40X) b) Cyst is lined by multinucleated giant cells (400X).

Clinical features in PCI have a varied range from abdominal pain, diarrhea, per-rectal bleeding, signs and symptoms of intestinal obstruction. The present case had signs and symptoms of intestinal obstruction. This can be attributed to mechanical obstruction by grape-like lesions. Authors believe that these lesions might be present since a long time. But tolerance and endurance of the patient did not cause symptoms. The repeated insults caused sizable growth of lesions. The chronic process of development is manifested acutely by intestinal obstruction.

CONCLUSION:

Clinical manifestations of PCI are varied and pose the surgeons for misdiagnosis. Imaging studies coupled with endoscopy are essential. Many cases can be treated conservatively. In resected specimens, histopathology remains the gold standard for an exact diagnosis. PCI is a fairly common condition with an optimistic prognosis. The underlying mechanism of development of PCI in the present case might be similar to gastro-intestinal syndrome seen in individuals with strenuous exercise.

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