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A RARE PRESENTATION OF PNEUMOPERITONIUM:PNEUMATOSIS CYSTOIDES INTESTINALIS (PCI)

Surgery

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ABSTRACT

Background: Pneumatosis cystoides intestinalis (PCI) is a rare gastrointestinal complication in the course of connective tissue diseases especially in scleroderma.1 It is a gas-filled cysts in the intestinal submucosa and subserosa. It is a rare disease with reported incidence of 0.03% and can occur in any age group.2 Pneumatosis cystoides intestinalis (PCI) was first reported by Du Vernoi in 1730.3 The pathogenesis of PCI remains unclear; however, six pathophysiologic mechanisms have been proposed including inflammation, physical damage of intestinal mucosa, nutritional imbalance and dysbacteriosis, gastrointestinal dysmotility, and immune dysfunction.4 The clinical signs and imaging features in these situations may mimic true abdominal viscera perforation, thus, it is imperative to diagnose correctly as treatment of PCI is generally conservative. **Objective:** To know about this rare diagnosis so that one can keep this as a differential diagnosis for pneumoperitoneum and thus avoid laparotomy.

KEYWORDS

INTRODUCTION:

Pneumatosis cystoides intestinalis (PCI) is a rare disease characterized by the presence of gaseous cysts containing nitrogen, hydrogen and carbon dioxide in the intestinal wall that may be iatrogenic or associated with a wide variety of conditions.[5] It is associated with various disorders including chronic obstructive pulmonary diseases, autoimmune disorders, and organ transplantation.[6] The exact etiology of the disease is still unknown. It is usually asymptomatic or with nonspecific symptoms such as abdominal pain, diarrhea, rectorrhagia, obstruction, flatulence, and rectal tenesmus, and may be an incidental finding in colonoscopies and radiological studies. Histopathologically, it is characterized by cystic structures lined with histiocytes, giant cells, and granulomatous inflammatory reaction with eosinophils and elastosis.[7] The rupture of cysts can cause pneumoperitoneum, air in the omentum, or pneumoretroperitoneum in less than 3% of cases, conditions that require either conservative or surgical management.[8]

Case Presentation:

A 54-year-old male patient presented to the hospital with complaints of pain in the umbilical region which is radiating towards the right shoulder and abdominal distension from past 3 hours. On abdominal examination there was generalized tenderness. Routine blood investigations and ABG were within normal limits. Chest X-ray was suggestive of gas under diaphragm. USG abdomen was suggestive of excessive air shadow in the abdomen with free fluid. Patient was also a known case of Gastric Outlet Obstruction cause Duodenal Ulcer. Thus, keeping this in view, suspicion of Duodenal Ulcer perforation was kept and patient was planned for laparotomy. On laparotomy, there was a gush of air. Multiple air-filled cysts were noted over the small and large intestine and at the spleen. Diagnosis of PCI was made. Post operative period was uneventful.

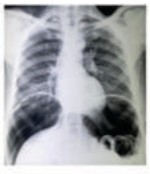
Fig. 1 Gas under diaphragm


Fig. 2 Gas filled cyst in small intestine


Fig. 3 Gas filled cyst in small intestine

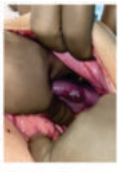
Fig. 3 Gas filled cyst at spleen

DISCUSSION:

Pneumatosis cystoides intestinalis (PCI) is a rare condition characterized by the presence of air-filled cysts present in the bowel wall and mesentery and may occur anywhere in the gastrointestinal tract. The cysts (0.5–10 cm in size) are found most frequently in the terminal ileum and rarely in the proximal small bowel, stomach, and colon.[9] When the air-filled cysts rupture, they cause a pneumoperitoneum, which often is benign in nature. The pathogenesis of PCI is not fully understood, but several mechanisms have been suggested. Mucosal break down from steroids and other immunosuppressive agent cause Peyer's patches in the bowel wall to shrink, leading to an alteration of mucosal integrity and the potential for air dissection. These immunosuppressive agents also impair tissue repair mechanisms, further exacerbating ulceration and bowel necrosis. The bacterial theory suggests that bacterial fermentation of carbohydrates within the gastrointestinal tract leads to excessive gas formation, inducing absorption of this gas into the bowel wall.[10] PCI is a benign lesion and very few patients face cancer. There is no unified standard for treatment of PCI. The most of PCI cases are usually managed conservatively, while exploratory laparotomy is considered if peritoneal irritation or persistent bowel obstruction occurs in PCI.[11]

CONCLUSION:

Pneumatosis cystoides intestinalis (PCI) is a rare condition in which gaseous cysts were formed in the intestinal wall (subserosa and submucosa). Due to the rupture of these cysts, there will be gaseous distension of the abdomen which will cause pneumoperitoneum and mimics like hollow viscus perforation. Thus, one should keep a differential diagnosis of Pneumatosis cystoides intestinalis (PCI) in a case of pneumoperitoneum and should be managed accordingly.

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