

Case Report

Pneumatosis cystoides intestinalis- a morphological curio or a pitfall for surgeons: report of two cases and literature review

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ABSTRACT

Pneumatosis Cystoides Intestinalis (PCI) is an uncommon but well recognised clinical entity in which gas-filled cysts appear in the intestinal wall. PCI can be an incidental finding or it may be detected during radiography or laparotomy. We came across two cases of PCI with characteristic morphological features associated with perforation peritonitis in and sigmoid volvulus respectively. In both cases PCI was not suspected pre-operatively. Both patients underwent urgent surgical exploration for the abdominal emergencies and were discharged in good general condition. It is imperative that the imaging finding of PCI is carefully correlated with the findings of physical examination, clinical history, and laboratory test results to determine which patients can be managed medically by treating the underlying disease and which will require emergency surgery. This decision can be difficult because the origin of the gas is often unclear and the patient's symptoms can be volatile, presenting a major dilemma for the surgeon.

Keywords: Pneumatosis cystoides intestinalis, Pyloric stenosis, Sigmoid volvulus

INTRODUCTION

Pneumatosis Cystoides Intestinalis (PCI) is a rare condition characterised by presence of multiple gas-filled cysts in the intestinal wall. PCI is not a disease but a clinical sign whose pathogenesis is poorly understood.¹

PCI occurs in two forms; primary and secondary to underlying causes. There are benign as well as serious causes of secondary PCI. The imaging appearance of both may look very similar. Correlation with clinical history, physical examination, and laboratory test results is the best indicator of whether PCI is due to a benign or life-threatening cause.

CASE REPORT

Case 1

A 40 year old male was admitted in surgery department with abdominal distension for last 3 days. He complained

of absolute constipation and 3 to 4 episodes of vomiting since 1 day. There was no prior history of similar complaints, surgical intervention or receiving any medication. On clinical examination abdomen was grossly distended with visible gastric peristalsis in lower abdomen and tenderness in epigastric region. Patient did not have sequel of any pulmonary or connective tissue disease. Routine Biochemical and haematological tests were within normal limits. The plain X- ray abdomen showed multiple cysts in a segment of ileum (Fig 1a). USG revealed gastric outlet obstruction with perforation peritonitis. On Gastric Endoscopy hypertrophic pyloric stenosis with foul smelling contents suggesting obstruction were noted.

With diagnosis of gastric outlet obstruction and perforation peritonitis, emergency exploratory laparotomy was performed. Partial gastrectomy was done to relieve obstruction. Perforation was not detected. Resection of the segment of ileum showing multiple cysts of variable sizes was done and ileostomy was performed.

The large intestine and rest of the abdominal organs were normal. Post-operative course was uneventful.

On Gross examination, the specimen of stomach showed remarkable scarring and tightness of the pylorus (Fig 1c). The resected ileal segment measuring about 35 cm, revealed cysts of size ranging from 0.5cm to 1.5cm on serosal aspect (Fig 1b). On histopathological study section from pylorus showed fibrosis. Sections from ileum showed mucosal ulcer and multiple cystic cavities below mucosa and serosa. Cysts were lined by multinucleated foreign body giant cells and inflammatory cells (Fig 1d, e).

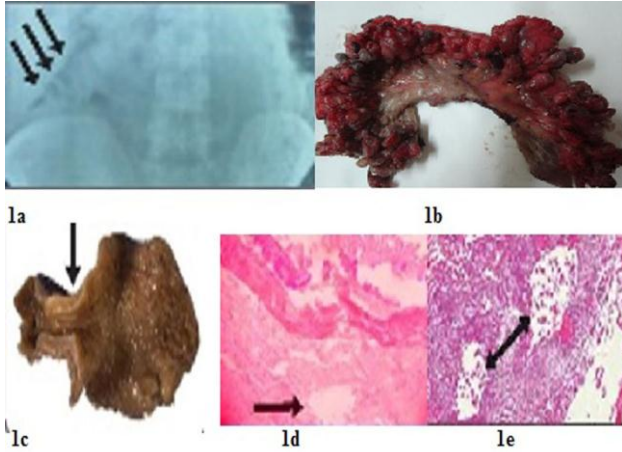


Figure 1a: X ray abdomen shows air filled cyst in ileum.

Figure 1b: Specimen of ileum showing multiple air filled cysts of variable size on serosal aspect.

Figure 1c: Segment of stomach showing hypertrophic pyloric stenosis.

Figure 1d: Microscopic section showing mucosal ulcer and cyst (arrow) HE 40x.

Figure 1e: Microscopic section showing cyst lining consisting of multinucleated giant cells HE 100x.

Case 2

A 80 year male presented with fullness of abdomen and pain for 10 days. He also complained of constipation since last 5 days. There was no history of vomiting. Past history was not indicative of any major illness or operative procedure. Abdomen was distended with generalised tenderness. Abdominal radiograph showed air-fluid level and USG showed over distended bowel loops (Fig 2a, b).

During emergency surgical exploration, volvulus of sigmoid colon was noted with presence of multiple cysts of variable sizes on outer surface of the colonic segment (Fig 2c). Resection of The involved colonic segment with colostomy was performed. Rest of the abdominal organs were normal. The patient was discharged in good general condition.

The surgical Specimen consisted of parts of descending and sigmoid colon measuring about -35 cm. The colon was twisted and dilated with congested external surface. On cut surface there were multiple cysts in muscular wall (Fig 2d).

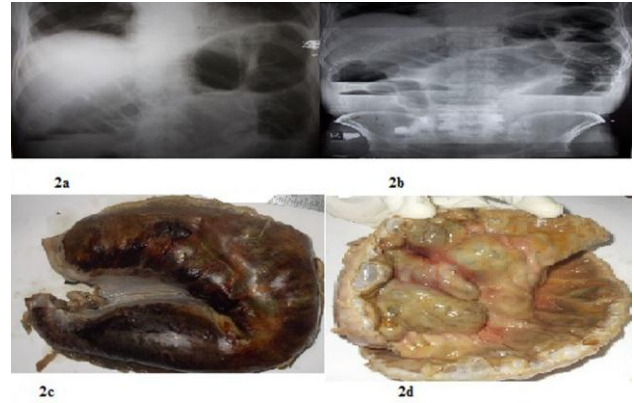


Figure 2a: X-ray abdomen showing gas under diaphragm.

Figure 2b: X-ray lower abdomen showing dilated segment of large intestine.

Figure 2c: Surgical specimen of sigmoid volvulus with congested external surface.

Figure 2d: Cut surface of the gross specimen showing multiple cysts in the wall of intestine.

Microscopically there were multiple intact or partially collapsed cysts lined by giant cells in the muscle layer below an intact mucosal lining.

DISCUSSION

Pneumatosis Cystoides Intestinalis also called intestinal emphysema is a morphologically uncommon condition that is characterised by gas filled cysts in bowel wall. Pneumatosis Intestinalis was first reported by Du Veroni in 1730 as a post-mortem observation.² The overall incidence of PI in general population is quoted to be 0.03% on autopsy series.³ PCI occurs in all age groups and is more common in men.⁴ In infants PCI occurs as a component of necrotizing enterocolitis and often has fatal outcome. PCI can occur anywhere in the gastrointestinal tract, but usually seen in the intestine.

In Adults PCI can be primary (idiopathic) mainly affecting large intestine in about 15% cases or secondary to underlying causes mostly affecting ileum followed by stomach in 85% cases.⁵ Secondary PCI is categorised into three groups⁶ as

Group 1: Patients with PCI as a complication of lung disease and initially normal Intestinal status.

Group 2: Patients who develop Pneumatosis on the grounds of intestinal disease.

Secondary to primary extra intestinal or generalised disease.

Group 3: Patients who develop PCI as a complication of primary gastro-intestinal disease.

The conditions underlying PCI can be differentiated in^{5,3}

- a) Traumatic and mechanical -pyloric stenosis, endoscopy, enteric tube placement volvulus, surgical anastomosis, carcinoma
- b) Inflammatory and autoimmune-Crohns disease, ulcerative colitis, diverticular disease, necrotising enterocolitis, poly-dermatomyositis, scleroderma, multiple sclerosis;
- c) Infectious -Clostridium difficile, HIV and AIDS, Cytomegalovirus, Mycobacterium Species
- d) Pulmonary -chronic obstructive pulmonary disease, Asthma, cystic fibrosis
- e) Drug induced -cytotoxic agents, immunosuppression, corticosteroids,
- f) Other conditions such as transplantation, graft versus host disease, leukaemia, or intestine infarction.
- g) Iatrogenic and
- h) idiopathic categories.³
- i) Pathophysiology of gas formation in PCI is explained by^{2,7}

I) The intraluminal gastrointestinal (GIT) gas that enters the intestinal wall due to increased intraluminal pressure, mucosal injury, or combination of them, steroids and other immunosuppressive agents that cause Payer's patches in the bowel wall to shrink, leading to an alteration of mucosal integrity and also impair tissue repair mechanisms.

II) In bacterial theory, it is stated that gases accumulate after the direct invasion of the bowel wall by bacteria. The intraluminal bacterially produced hydrogen tension exceeds the nitrogen tension in the blood, causing a hydrogen diffusion gradient towards the sub mucosal vessels. This theory explains the pneumatosis observed near blood vessels along the mesenteric border and is supported by the disappearance of the gas after antimicrobial drug therapy.

III) Pulmonary gas- It has been postulated that alveolar rupture can result in the movement of air along vascular channels in the mediastinum, tracking caudally via the retroperitoneum and to the mesentery of the bowel.

Clinical presentation of PCI varies from asymptomatic patients to symptoms and signs associated with life threatening complications such as bowel ischemia, perforation and peritonitis. Generally the symptoms are mild and include abdominal pain, diarrhoea or constipation, flatus distension, or weight loss. Serious symptoms such as bleeding or ileus can also occur.⁹ It is accepted that the clinical manifestations of PCI are not

necessarily associated with the intramural gas or its location in the gastrointestinal tract but rather with the underlying disorder.

While Physical examination findings pertain to the underlying cause, Radiological tools are important for diagnosing PCI .A plain abdominal radiograph usually shows radiolucency within the wall of the GI tract. Gas under diaphragm, e/o pneumoperitoneum

Abdominal radiographic findings are detected in approximately two thirds of the patients with PI.⁷

Abdominal Sonography is commonly applied to the paediatric patient for avoidance of ionizing radiation. Computed tomography is the best imaging modality for establishing diagnosis of PI and can distinguish PI from intraluminal air or sub mucosal fat. It provides the diagnosis of associated pathologic conditions. However pattern on radiology or extent of PCI does not correlate with the severity of symptoms or underlying disease.⁸

Macroscopic elevations of the mucosa or other changes make PCI difficult to distinguish from other lesions evaluated by endoscopy like polyps, carcinoma, lymphoma etc. Colonoscopy has been used to visualise gas cysts in the sub mucosa of the colon.⁶

Although PCI was not diagnosed pre-operatively, after assessing medical history, radiological and surgical findings it can be concluded that our first case belonged to secondary PCI of group 3 category. Mechanical trauma in the form of pyloric stenosis appeared to be the underlying condition. In second case it could not be ascertained whether sigmoid volvulus was the cause or the complication of PCI as same intestinal segment was involved by the two pathological processes and previous clinical record was not contributory. the decision of urgent surgical exploration in both cases was based on the available clinical and radiological findings. Gross and histological exam of the surgical specimen showed distinctive morphological features of PCI.

Morphological features of PCI are quite distinctive. Numerous grapelike small cysts devoid of any contents are seen on the serosa and mesenteric border of the gut on gross examination. The whole length of the involved intestinal segment shows multiple sub-serosal cysts varying in size from a pinhead to about 3.5cm diameter. Crepitus can be elicited while handling the specimen.⁴

On microscopic examination, variable sized cystic spaces lined by macrophages, admixed with an occasional giant cell are seen in the sub mucosa and serosa with pericyclic inflammatory changes.¹

PCI follows a chronic and indolent course when not associated with other abnormalities. Complications including volvulus, intestinal obstruction, tension

pneumoperitoneum, bleeding, intussusception, and intestinal perforation may be seen in 3% of patients.⁹

Specific treatment is not recommended in asymptomatic patients who are detected as having PCI radiologically. Treatment of Pneumatosis cystoides Intestinalis ranges from supportive care including nasogastric decompression, intestinal rest, Broad spectrum antibiotics, metronidazole, steroids, high flow oxygen and hormones to laparotomy. Surgery is indicated in patients with severe pain, rectal bleeding, fever or an evidence of ischemic bowel.^{6,10}

CONCLUSION

Analysis of a detailed history, physical examination and radiological studies are of utmost important in a case of PCI, if pitfall for surgeons is to be avoided.

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