

# An Unusual Case of Small Bowel Volvulus Associated with Pneumatosis Intestinalis: A Case Report

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## Research Article

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# Abstract

**INTRODUCTION:** Pneumatosis cystoid intestinalis (PCI) is a rare disease reported in the literature affecting 0.03% of the population. It has a variety of causes and its manifestation may change widely. In the acute complicated form of pneumatosis intestinalis, management is challenging for physicians and surgeons.

**CASE PRESENTATION:** We present a case of a 60-year-old patient who was admitted to our surgical department with a symptomatology suggestive of small bowel occlusion. Computed tomography demonstrated ileal volvulus associated with parietal signs suffering and pneumoperitoneum. An emergent exploratory laparoscopy followed by conversion was performed demonstrating segmental ileal pneumatosis intestinalis secondary to a small bowel volvulus due to an inflammatory appendix wrapping around the distal ileum. Detorsion, retrograde draining, and appendectomy were performed.

**DISCUSSION:** PCI is an uncommon disease, affecting 0.03% of the population. It is usually presenting as a marginal finding resulting from various gastrointestinal pathologies. This case is exceedingly rare in the literature, featured by the ileal volvulus due to appendicitis.

**CONCLUSION:** This work emphasizes the importance of surgical procedures in the management of symptomatic pneumatosis intestinalis.

## Introduction:

Pneumatosis cystoid intestinalis (PCI) is a low-incidence pathology defined by the existence of air in the small intestine or colon's wall (1). PCI can affect any portion of the gastrointestinal tract and could be present in any layer such as the mucosa, submucosa, or subserosa (1–3). It can either be presented as a secondary form in 85% of cases or an idiopathic form in 15% of cases (1, 2). The secondary pattern occurs more frequently in gastrointestinal causes such as bowel obstruction (3, 4). The management of PCI is challenging to surgeons especially in symptomatic cases (5). We report a rare case of ileal pneumatosis cystoides associated with small bowel volvulus, presenting with acute abdominal pain.

## Case Report:

A 60-year-old male patient without any medical or surgical history consulted the emergency department for 24 hours of abdominal pain, distension and vomiting. The patient had presented this pain a year earlier which faded away spontaneously. On physical examination, tachycardia and distended abdomen with mild tenderness were noted. White blood count was 8840 E/mm<sup>3</sup> and C reactive protein was 36 mg/l. X-ray of thorax and abdomen showed dilated small bowel, multiple fluid levels and pneumoperitoneum (Figure A).

An abdominal CT scan was performed, revealing distended small bowel loops upstreaming transitional levels like a 'whirl sign' (Figure B1), a bubbly pattern across the length of the small bowel associated with parietal suffering signs (Figure B2), abundant pneumoperitoneum (Figure B3), and a pathological meso-cealic appendix (Figure B4). The CT scan has suggested a diagnosis of ileal volvulus due to the meso-cealic appendix.

We initially decided to perform laparoscopy. Intraoperatively, small bowel loops were much dilated not allowing intraperitoneal exploration. Gas-filled cystic lesions on small bowel serosa were identified. There was no evidence of perforation. We did choose to convert for better and prudent exploration. A volvulus was found, involving a two-and-a-half clockwise turn around a long, pendulous small bowel mesentery, the strangled bowel was greatly congested

(Figure C1). At the base of the volvulus, an inflammatory appendix was wrapped around the last loop of the ileum (Figure C2). Also, multiple gas-filled subserosal cysts, differently sized, on the wall of the ileum were encountered (Figure C3). When the ileum was re-rotated, Small bowel loops had preserved vitality. The entire colon was normal. Detorsion, retrograde draining, and appendectomy were performed.

Postoperatively, the patient completed a five-day course of intravenous metronidazole. There were no postoperative complications. Anatomopathological examination revealed an inflammatory appendix without malignancy. A lower endoscopy was completed after surgery. It showed the presence of two polyps on the rectum and the transverse colon in low-grade dysplasia with no other lesions.

## Discussion:

PCI is an uncommon disease (0.03% of adults) (6). Its pathogenesis is still not clear (7). According to its etiology, literature classifies this entity as a primary or secondary type (8). The idiopathic type usually affected the left colon and is rarely reported in the literature. We found thirteen cases of primary PCI described in the international literature. We summarized the cases in Table 1. However, the secondary type affected frequently the small intestine and the right colon (9). Its pathogenesis is multifactorial, can be explained by 3 theories: mucosal disruption, bacterial theory, and pulmonary disease (1, 7, 10, 11). The mucosal disruption is due to the dissemination of bowel gas through a mucosal defect into lymphatic channels (1, 10). Wu et al. (12) found that high altitude is a new theory explaining PCI's pathogenesis: Highland areas induce passage of intraluminal gas into the submucosa damaging the mucosa. Mucosal damage can result from bowel occlusion, inflammatory process, and cytotoxic medical treatment (11). The pulmonary cause is confirmed in patients with asthma and chronic bronchitis: in these cases, the rupture of alveoli causes the migration of air bubbles from interstitial spaces through the mediastinum and from the retroperitoneum to the blood vessel of the intestinal wall (1, 8, 10). However, the bacterial theory is explained by bacterial gas entry due to a defect on the bowel wall lymphoid tissue (8, 10). This mechanism can justify the use of antibiotics (1). Chemotherapy or hormonal therapy, and systemic sclerosis were reported in the literature as a cause of PCI (1). Finally, while keeping in mind these theories, their pathogenesis has not been yet fully clarified (1).

Besides, the disease's location on the digestive tract may be helpful to guide the etiology. So pyloric stenosis or gastric cancer can lead to a proximal pathology; however distant one might be due to mesenteric ischemia or diverticulitis (9).

PCI is a rare entity reported in the literature, but nowadays PCI reports' number has been increasing because of the widespread use of CT scan and colonoscopy (9). This pathology is more frequently asymptomatic (1). Whereas in some cases, they may present with symptoms such as abdominal pain, constipation, distension, diarrhea, or bleeding (8, 9). Incidentally PCI can induce surgical complications such as bowel obstruction, intestinal perforation, volvulus, intussusception, and bleeding, which require surgical intervention (13).

Intestinal obstruction can be a rare complication of PCI. This event depends on the size and number of the cysts which lead in certain cases to a reduction of the intestinal lumen, volvulus, perforation, and hemorrhage (7, 11). In the literature, PCI associated with volvulus is much more uncommon. Besides this association, one of the highlights of our case is the long and hypermobile small bowel mesentery. Moreover, PCI is discussed to be a mechanical factor leading to irreversible volvulus, also it is disputed that volvulus contributes to ischemia which is an etiological factor leading to PCI (3, 14).

Imaging findings may be helpful to confirm PCI's diagnosis, especially on CT scans (1, 6). Computed tomography can show a grape cluster aspect within the wall of the intestine (1). Three patterns of pneumatosis have been reported in

the literature using CT scan imaging: bubble cystoid, a linear pattern, and a circular pattern (1).

Pneumoperitoneum can be explained by the rupture of the cyst on the wall intestine, without any evidence of peritoneal irritation or digestive perforation, like in our case. So that we should be wise to correlate clinical and radiographic findings, when free air is present below the diaphragm in chest X-ray (8, 9). Pneumatosis intestinalis and portomesenteric venous gas (PVG) are generally debated independently in the literature. This association of radiological findings usually concludes to the presence of mesenteric infarction, but they may indicate occasionally nonischemic conditions. So that their presence should not be always regarded as signs of severity (13, 15, 16). Moreover, according to literature, the rare findings of PCI and PVG can be present in asymptomatic patients without ominous signs, as described in the series of Sooby et al. (17), including 88 patients with PCI/PVG of which 19 with benign PCI, and of these 19, 6 patients had both PCI and PVG. These patients were put under surveillance, and they had no uneventful recovery.

The management of PCI is not well established, there are no standard therapeutic rules (10). However, the mandatory in its management is to judge whenever it is benign or life-threatening (11). So that it is established that if a CT scan shows intestinal infarction, urgent surgery is mandated. If no signs of intestinal damage is found, a conservative treatment is regarded to be ideal (13). The common conservative procedure is to use metronidazole Antibiotics, which affects intestinal bacteria by the suppression of hydrogen production, and hyperbaric oxygen therapy (2, 6, 8, 10). Nevertheless, a surgical procedure is indicated in complications such as peritoneal irritation or intestinal obstruction (2, 10).

The second particularity in our case is that the volvulus of the small bowel is due to acute appendicitis. This entity is explained by the wrapping of the appendix, due to its particular length, around the ileum occurring volvulus and strangulation (18, 19). According to the literature, this mechanism resulted from adhesion of the inflamed appendix to the posterior peritoneum forming a turn of the spire of the ileum last loop resulting in volvulus (18, 19). In summary, our case is exceedingly rare in the literature, featured by the ileal volvulus due to appendicitis.

Table: Review of the literature (2008–2021) illustrating cases of Idiopathic PCI, and PCI secondary to surgical etiologies.

Author	Age	Physical examination	Abdominal radiography	CT scan	Treatment	Etiology of PCI
Year	M/F					
<b>González et al. (20)</b> <b>2021</b>	66 Y M	Normal		Intestinal pneumatosis	Conservative: surveillance	Idiopathic
<b>Moyon et al. (6)</b> <b>2020</b>	79 Y M	Tachycardia  Abdominal distension  Mild pain on the lower abdomen without tenderness	–	The bubbly pattern across the length of the small bowel, Multiple cystic round shapes in the wall of the jejunum, and its mesentery, Large pneumoperitoneum	1/Conservative: surveillance + oxygen + broad-spectrum antibiotics  2/ Worsening pain laparoscopy:  No evidence of perforation / gas-filled cystic lesions	Idiopathic
<b>Takahashi et al. (21)</b> <b>2019</b>	17 Y F	Mobile mass in the right lower quadrant		Colocolic intussusception of the ascending colon with air in the bowel wall	Endoscopy: fine-needle aspiration	Idiopathic
<b>Suda et al. (22)</b> <b>2018</b>	80 Y M	Slight abdominal distention	Dilatation of the small intestine + retention of gas in the one long segment of the small intestinal wall	Massive gas-filled cysts within the wall and mesentery of the small intestine  No portal venous gas  No intestinal ischemia	Conservative: antibiotics	Idiopathic
<b>Wang et al. (23)</b> <b>2018</b>	56 Y M	Normal	Normal	–	High-frequency electrosurgical resection of the gas cysts + ATB Bifidobacterium	Idiopathic
<b>Romano-Munive et al. (24)</b> <b>2017</b>	54 Y F	Normal		Pneumatosis cystoides intestinalis	Conservative	Idiopathic
<b>Furihata et al. (15)</b> <b>2016</b>	81 Y M	Severe epigastric tenderness + distension No peritoneal signs	Massive free gas bilaterally in the subdiaphragmatic spaces	Massive free gas bilaterally under the diaphragm	Conservative: intravenous infusion of antibiotics, and nasogastric intubation	Secondary to chronic obstipation
<b>Fraga et</b>	66 Y	Abdominal	–	Gas in the	Conservative	Idiopathic

<b>al. (25)</b> <b>2016</b>	F	distention No signs of peritonitis		abdominal wall, at the level of the transverse and rectosigmoid colon		
<b>Tseng et al. (26)</b> <b>2014</b>	50 Y F	Abdominal distension without tenderness	Gas in the bowel wall	Pneumatosis intestinalis of the right side of the colon	Conservative	Idiopathic
<b>Slesser et al. (5)</b> <b>2011</b>	74 Y M	Abdominal distention Soft and non-tender	Bilateral free subdiaphragmatic air	Extensive pneumatosis intestinalis involving the small bowel with free intraperitoneal air secondary to malrotation of the proximal small bowel	Laparotomy: Extensive and bulky pneumatosis intestinalis extending from the duodenal-jejunal flexure to the terminal ileum	Idiopathic
<b>Nagata et al. (27)</b> <b>2010</b>	23 Y M	Tenderness in the right iliac fossa	--	Intussusception of the ascending colon Ovoid radiolucencies with smooth margins	Intussusception easily resolved by colonoscopy Laparoscopy-assisted partial ascending colectomy was performed 1 month after his initial presentation	Idiopathic
<b>Arora et al. (4)</b> <b>2009</b>	27 Y M	Signs of acute abdomen	Gas under the diaphragm	--	Laparotomy: Perforated duodenal ulcer + The terminal 180 cm of the small bowel to the ileocecal junction showed multiple thin-walled, tense, air-filled cysts on the serosal surface Limited right colectomy and resection of the involved small bowel	Secondary to a surgical cause
<b>Liau et al. (3)</b> <b>2008</b>	76 Y	Signs of generalized peritonitis	Massive subdiaphragmatic air and multiple dilated loops of small bowel with well-demarcated wall	Pneumoperitoneum gas-filled cysts on the wall of small bowel loops	Laparotomy: extensive small bowel infarction due to volvulus of most of the small bowels, multiple gas-filled subserosal	Secondary to a congenitally long mesentery

## Conclusion:

PCI is a rare disease whose diagnosis is offering a challenge for surgeons. This rare condition can often be associated with benign diseases or it can be proof of intestinal necrosis. Although surgery is mandatory in the complicated pattern, the treatment of asymptomatic forms is more likely conservative. Besides, both surgical and medical approaches can efficiently compete with these challenging diagnoses.

## Declarations

### Conflict of interest for all authors:

The authors declare no competing interest.

## Funding:

None.

## Compliance with Ethical Standards:

The patient has provided both verbal and written consent for the publication of this article. It was made sure that his identity will be kept a secret at all levels.

## Consent:

Written informed consent was taken from the patient regarding the publication of this case report. It was made sure that his identity will be kept a secret at all levels. A copy of a written request is available for review if requested.

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## Author contributions:

All authors were involved in the researching, writing, and editing of the manuscript.

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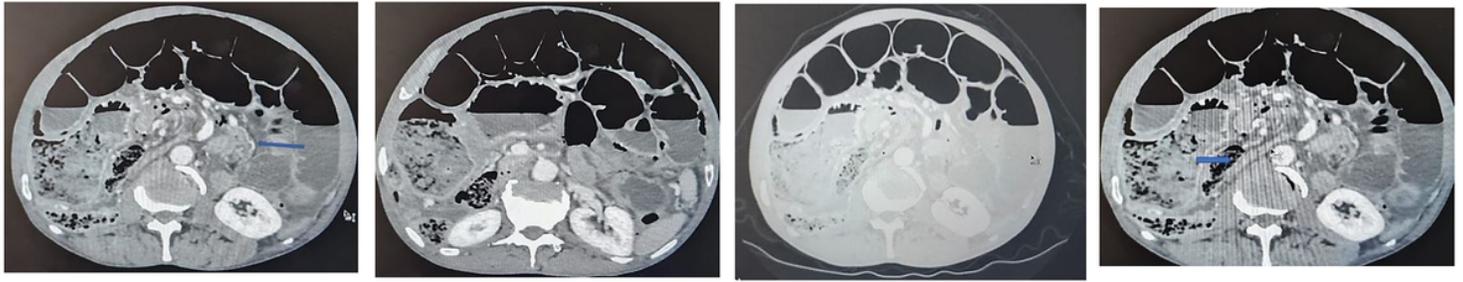
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## Figures



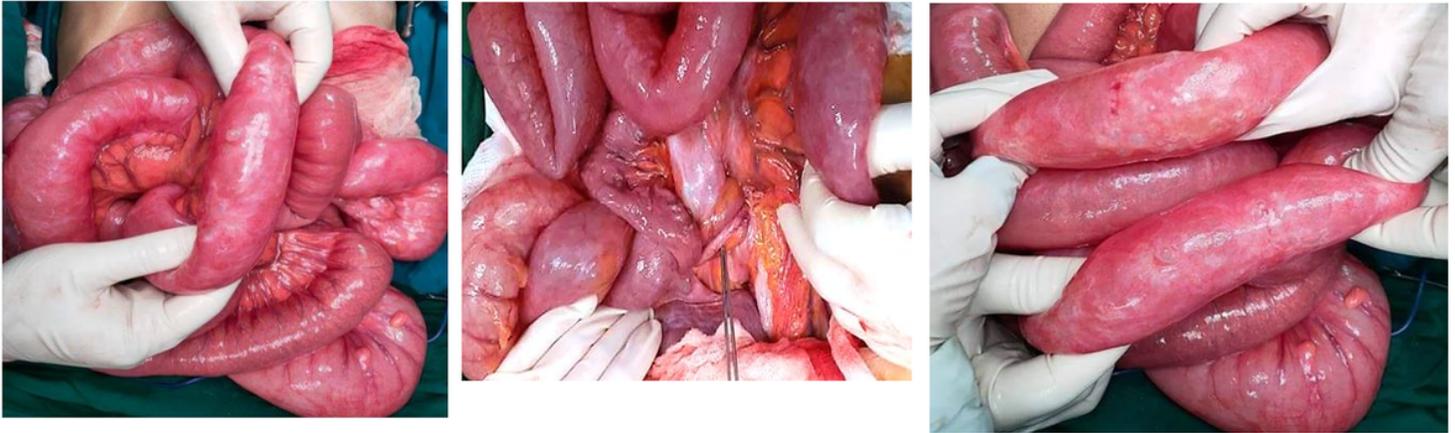
**Figure 1**

Dilated small bowel, multiple fluid levels and pneumoperitoneum on X-ray



**Figure 2**

B1: Ileal volvulus: dilated bowel segments associated with whirl sign B2: Multiple bubble lesions among ileal loops on Abdominal CT B3: Abundant pneumoperitoneum B4: The inflammatory appendix



**Figure 3**

C1: Ileal loop volvulus, involving a two-and-a-half clockwise turn around a long, pendulous small bowel mesentery C2: At the base of the volvulus, the inflammatory appendix wrapped around the last loop of the ileum C3: Segmental pneumatosis intestinalis involving the ileal loops