

Pneumatosis Cystoides Intestinalis: A Benign Cause of Pneumoperitoneum

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Abstract

Pneumatosis cystoides intestinalis (PCI) is an uncommon condition where cyst-like locules of gas are present in the submucosa or subserosa of the gastrointestinal tract. The majority of cases are benign and are associated with drugs such as chemotherapy agents or conditions including chronic obstructive airways disease. We present the case of PCI resulting in pneumoperitoneum in a 72-year-old male patient on chemotherapy for esophageal adenocarcinoma. While he was noted to have an extensive pneumoperitoneum and mesenteric emphysema on imaging, he remained clinically stable with a benign abdominal examination and was managed conservatively with intravenous antibiotics and fluids. This case highlights the importance of benign PCI as a differential to bowel ischemia in patients with pneumatosis intestinalis and pneumoperitoneum, particularly given the possibility of avoiding operative management and the resultant morbidity.

Keywords: Pneumatosis cystoides intestinalis; Chemotherapy; Pneumoperitoneum; Pneumatosis intestinalis

Introduction

Pneumatosis cystoides intestinalis (PCI) is an uncommon condition where cyst-like locules of gas are present in the submucosa or subserosa of the gastrointestinal tract. The majority of cases are benign and are associated with drugs such as chemotherapy agents or conditions including chronic obstructive airways disease [1, 2]. As in fulminant cases such as those secondary to bowel ischemia, benign PCI can result in pneumoperitoneum, mesenteric or retroperitoneal free gas and is an important differential in clinically well patients given the possibility of non-operative management [2]. We present a case of PCI resulting

in pneumoperitoneum in a 72-year-old male patient on chemotherapy for esophageal adenocarcinoma. Despite his radiological findings, he remained clinically well with normal hemodynamics and a benign abdominal examination which prompted a conservative approach to management that was successful.

Case Report

Investigations

A 72-year-old male patient currently undergoing neoadjuvant chemo-radiotherapy (CROSS protocol) for distal esophageal adenocarcinoma was referred to hospital after being incidentally noted to have a pneumoperitoneum on an outpatient computed tomography (CT) of chest, abdomen and pelvis. He had completed his final round of chemotherapy 1 week prior and had undergone the CT for staging prior to a multidisciplinary team meeting at which stage he was to be considered for an esophagectomy. Relevant past medical history included a previous smoking history with emphysematous changes on CT; however, he was not on any treatment and had a reasonable exercise tolerance being able to walk up two flights of stairs. He had also been treated for lung adenocarcinoma with a left upper lobectomy a decade prior. He was not on any regular medications. Confounding his presentation was his history of having a jejunostomy inserted 3 months previously for feeding due to dysphagia from his partially obstructing distal esophageal malignancy.

On presentation, he reported increasing shoulder tip pain over the past 48 h and generalized lethargy which was not dissimilar to following his previous rounds of chemotherapy. He denied worsening abdominal pain and fevers, however did report the development of non-blood containing diarrhea. He was afebrile and hemodynamically stable with a heart rate of 94 bpm, blood pressure of 129/86 mm Hg and oxygen saturation of 96% on room air. He was comfortable and mobilizing independently around the emergency department. On examination, he was clinically dehydrated and was noted to have a mildly distended abdomen which was tender diffusely across the upper abdomen without peritonism.

Diagnosis

He was found to be leukopenic in the setting of being 1 week post treatment with carboplatin and paclitaxel with a white cell

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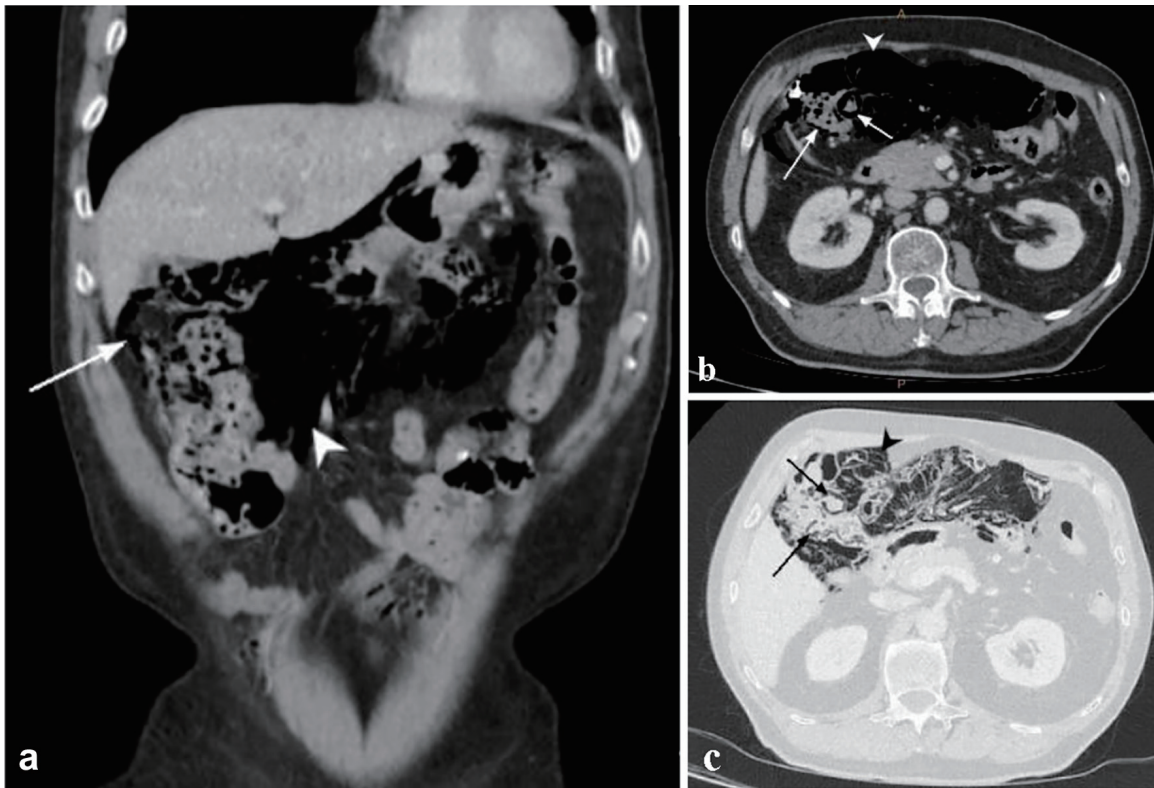


Figure 1. Contrast-enhanced CT of abdomen and pelvis: (a) coronal view in soft tissue window, (b) axial view in soft tissue window, (c) axial view in lung window, showing intramural locules of gas within the right sided colon (arrow) and extensive gas within the mesocolon and mesentery (arrowhead) and pneumoperitoneum outlining the liver margin. CT: computed tomography.

count of $1.5 \times 10^9/L$. He had a normal acid-base balance on venous blood gas, no electrolyte abnormalities and slightly elevated C-reactive protein of 16 mg/L. His intravenous contrast CT showed a pneumoperitoneum predominantly in the right subhepatic space with extensive mesenteric gas and intramural gas locules predominantly within the hepatic flexure (Fig. 1). There was no evidence of free fluid, bowel wall thickening or hypoenhancement to suggest ischemia.

Treatment

While his immunosuppression was potentially masking clinical and biochemical signs of bowel ischemia, his examination and history were reassuring and the decision was made to proceed with non-operative treatment. He was kept fasting with his jejunostomy feeds suspended, commenced on nasal prong oxygen, intravenous fluids and intravenous piperacillin/tazobactam given his neutropenia. Given he remained clinically well overnight with a soft abdomen on examination the next morning, he was recommenced on his jejunostomy feeds. Over the next 72 h his abdominal tenderness resolved and his inflammatory markers remained stable and on day 3 his antibiotics were de-escalated to amoxicillin/clavulanic acid administered via his jejunostomy. His diarrhea however was ongoing and after his stool bacterial and viral polymerase chain reaction (PCR) and *Clostridium difficile* toxin re-

turned as negative, he was commenced on loperamide with good effect.

Follow-up and outcomes

He was discharged home on day 6 of admission on a further 5 days of antibiotics with follow-up arranged with his upper gastrointestinal surgeon and medical oncologist. He underwent a repeat CT of abdomen and pelvis 2 months post admission which noted resolution of PCI, pneumoperitoneum and mesenteric gas.

Discussion

PCI refers to the radiological findings of cyst-like intramural gas locules within the intestinal tract, commonly within the small intestine and colon but can occur from esophagus to rectum [1]. Approximately 15% are idiopathic whilst secondary PCI is associated with a variety of etiologies as outlined in Table 1, including gastrointestinal causes such as enterocolitis, ischemia and gastrointestinal surgery. Pharmacological causes include anti-neoplastic agents as in this case, steroids or immunosuppressants and alpha-glucosidase inhibitors. Non-gastrointestinal causes include obstructive airways disease, hematological malignancies, acquired immune deficiency syn-

Table 1. Causes of Secondary PCI

Gastrointestinal causes	Non-gastrointestinal causes
Gastrointestinal ischemia	Obstructive airways disease
Bowel obstruction	Asthma
Volvulus	Chronic obstructive airways disease
Intussusception	Cystic fibrosis
Pyloric stenosis	Connective tissue diseases
Necrotizing enterocolitis	Drugs
Inflammatory	Anti-neoplastic agents
Enteritis	Steroids and other immunosuppressants
Appendicitis	Alpha-glucosidase inhibitors
Diverticulitis	Lactulose
Inflammatory bowel disease	Malignancy
Peptic ulcer disease	Lymphoma
Abdominal trauma or mucosal injury	Leukemia
Gastrointestinal surgery or endoscopy	Infective
	<i>Clostridium difficile</i>
	Acquired immune deficiency syndrome
	Cytomegalovirus
	Graft-versus-host disease

PCI: pneumatosis cystoides intestinalis.

drome and autoimmune conditions perhaps due to the use of glucocorticoids [2, 3].

While the exact pathophysiology remains unclear, it is believed to relate to one or more of the following: mucosal injury, gas producing bacteria and increased intestinal pressure such as from intestinal obstruction or airway obstruction [1, 2]. In the setting of chemotherapy, the development of mucositis with reduced mucosal integrity and altered intestinal flora coupled with underlying immunosuppression may play a role in the development of PCI [3-5]. Additionally, the use of glucocorticoids in chemotherapy may result in atrophy of the intestinal mucosa, hence predisposing to greater susceptibility of the mucosa to increased intestinal pressure resulting in submucosal gas [6]. Interestingly, the described patient had a background of recent chemotherapy and obstructive airways disease, both of which are associated with benign PCI. His jejunostomy was believed less likely to be contributing to his presentation given it had been 3 months since insertion and the mesenteric gas was predominantly around the right upper quadrant of the abdomen, including the omentum and mesocolon.

The clinical findings of PCI are often non-specific if present at all. A systematic review by Wu et al of PCI within the Chinese population found abdominal pain, distension and diarrhea to be the most common symptoms [7]. Diagnosis has largely been reliant on radiological investigations; however, endoscopy is playing an increasing role allowing for the direct visualization of submucosal cysts which present as polypoid structures. While these cysts can be mistaken for polyps or inflammatory bowel disease, they are readily apparent on en-

doscopic ultrasound [2]. X-ray may be sufficient to visualize PCI given the radio-opaque nature of the intramural cysts, particularly when outlined in the setting of a barium enema, and can identify complications including pneumoperitoneum. CT identifies the intramural gas consistent with PCI, particularly on a lung window and identifies signs of spontaneous perforation including pneumoperitoneum, pneumoretroperitoneum or mesenteric gas. Where CT is of particular utility compared to alternative imaging modalities or endoscopy is in the identification of features suggestive of fulminant PCI [7-9].

Management of PCI relies on distinguishing between benign and fulminant causes with intestinal ischemia being the primary aetiology of concern, particularly in the setting of peritonitis, hemodynamic instability or leukocytosis. Concerning features on CT more consistent with fulminant PCI include free fluid, reduced bowel wall enhancement, bowel wall edema and portal venous gas all of which were absent in the presented case and strongly suggest the need for operative intervention [8, 9].

Asymptomatic patients require no intervention, or may be managed conservatively with supplemental oxygen including hyperbaric oxygen; to increase the partial pressure of oxygen and reduce the partial pressure of other gases in the circulatory system facilitating the diffusion of gas from the cysts into the blood stream; antibiotics to reduce the intestinal gas-producing bacterial load and fluid replacement in the setting of diarrhea [1, 2, 7]. Additionally, endoscopy is increasingly being used therapeutically by facilitating aspiration or excision of cysts [2, 10]. While endoscopic intervention may not be feasible in the often-widespread nature of PCI, it has a role in manag-

ing possible sequelae including intestinal obstruction from large cysts or preventing a volvulus from cysts acting as a fulcrum [10]. The incidence of complication such as perforation or obstruction is reported as 16.3% by Wu et al and as in the presented case, may still be managed conservatively [7]. Surgery should be reserved for patients showing signs of peritonism or sepsis or for cases of persistent bowel obstruction either not amenable to, or recurrent after endoscopic intervention [7].

Ultimately the rate of recurrence is high particularly in primary PCI, and the main stay of treatment is to relieve the underlying cause in secondary PCI such as cessation of chemotherapy as in this instance [1]. In conclusion, by presenting this case we aim to inform clinicians of this uncommon and often benign cause of pneumoperitoneum and highlight the potential for non-operative management in order to reduce the morbidity associated with unnecessary surgery.

Learning points

PCI is an uncommon radiological and endoscopic finding and often follows a benign cause. While bowel ischemia is an important consideration in the setting of a pneumoperitoneum in a patient undergoing chemotherapy, PCI remains an important differential, particularly in the setting of a clinically well patient. Correctly identifying PCI may allow patients to be managed non-operatively thus avoiding the morbidity involved with surgical intervention.

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Financial Disclosure

None to declare.

Conflict of Interest

None to declare.

Informed Consent

Informed consent was gained prior to completion of the case report.

Author Contributions

PWP contributed to writing, editing and conception of the case

report. AF contributed to the conception and editing of the case report.

Data Availability

The authors declare that data supporting the findings of this study are available within the article.

Abbreviations

PCI: pneumatosis cystoides intestinalis; CT: computed tomography

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