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Pneumatosis Cystoides Intestinalis as Incidental Finding with Ischemic Ileum: A Case Report

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Abstract

Purpose: Pneumatosis Cystoides Intestinalis (PCI) can be innocuous or fatal. Distinguishing pathologic etiology from benign is problematic. Physicians must cumulatively evaluate patient's vital signs, clinical signs and symptoms, lab data and radiographic image to reach a management plan.

Methods: We present a case of an 83-year-old man with presentation of abdominal pain. We provided a review of the clinical manifestations, imaging data of this case, and the literature related to the PCI.

Results: A male patient had a history of cecum adenoma post with laparoscopic assisted right hemicolectomy 13 years ago, recurrent interstitial pneumonitis related to Nivolumab under methylprednisolone 4 mg use, and diabetes under acarbose control. This patient was admitted to the emergency department with right upper quadrant abdominal pain. A Computed Tomography (CT) scan showed PCI and ischemic colitis. Emergent explore laparotomy was performed and subtotal colectomy and small bowel resection due to incidental bowel ischemia under Indocyanine Green (ICG) image. Patient's condition continued to deteriorate and expired on post-operative day 81.

Conclusion: For acute abdomen or sepsis condition, operation should be arranged due to suspicion of ischemia or necrosis. Laparoscopy can be considered if certain risk factors are presence. If ischemic colon is noted, small intestine should be assessed carefully, especially the ileum, due to higher possibility of small intestinal pathologic PCI.

Keywords: Pneumatosis Cystoides Intestinalis • Ischemic colitis • Small bowel ischemia • Indocyanine green

Abbreviations: PCI: Pneumatosis Cystoides Intestinalis • CT: Computed Tomography • ICG: Indocyanine Green • RUQ: Right Upper Quadrant • ICU: Intensive Care Unit • POD: Post Op Day

Introduction

Pneumatosis cystoides intestinalis (PCI) was first described in autopsy by Du Vernoy in 1730 as presence gas-filled cysts in intestinal submucosa and subserosa. PCI is not a disease but merely a radiographic sign. Although its true incidence is unknown and its cause is multifactorial, an increasing trend of incidence is noted as Computed Tomography (CT) exams became more readily available. PCI is divided into primary and secondary type. Primary PCI is seen as radiologically cystic collection of air, usually located in colon; generally benign in nature but may be complicated with obstruction or bleeding. Secondary PCI is characterized by linear or circumferential collection of air that is related to necrotic or non-necrotic gastrointestinal and pulmonary pathology, as described by Koss' review in 19521 [1-3]. A multicenter study reported that 62% of patients had benign etiology and 38% had pathologic etiology based on presence of transmural ischemia in surgery or in autopsy [4]. Pathologic PCI occurs more frequently in the small intestine, especially in ileum, and in the presence of ascites, whereas asymptomatic PCI occurs more commonly in the ascending colon. Acute diarrhea is a clinical characteristic of asymptomatic PCI, a benign etiology without indication of invasive procedures [5]. The presence of PCI on radiologic image can be innocuous or fatal so physicians must cumulatively evaluate patient's vital signs, clinical signs and symptoms, lab data and radiographic image to reach a management plan. We describe a case of PCI presenting as acute abdomen related to perforation of colon with small intestine ischemia after ICG use.

Case Report

An 83-year-old man with chronic tobacco use has a medical history of cecum tubulovillous adenoma with focal high-grade dysplasia post with laparoscopic assisted right hemicolectomy 13 years ago, prostate adenocarcinoma, lung adenocarcinoma, recurrent interstitial pneumonitis related to Nivolumab under methylprednisolone 4mg use twice daily, and diabetes under acarbose control for half-an-year. He was presented with right upper quadrant (RUQ) abdominal pain for four days when he visited our emergency department. Vital signs upon admission were BT 37 'C, HR 91 bpm, RR 20 cpm, BP150/129 mmHg, SpO2 96% under room air, with GCS E4M6V5. Physical examination revealed RUQ tenderness and positive Murphy's sign. Laboratory tests revealed normal white blood cell count of 7100/μL (normal range: 4000–10000/μL), elevated neutrophil level of 89.9% (normal range: 39.4-72.6%) and mildly increased total bilirubin level of 1.27 mg/dL (normal range: 0.3-1.2 mg/dL). Under the impression of acute abdomen, abdominal CT was ordered. Abdomen CT showed linear air collection from transverse colon to descending colon with poor enhancement of mucosal and muscular wall and pneumoperitoneum. Pneumatosis cystoides coli was diagnosed and ischemic colitis was suspected (Figure 1). The patient underwent emergent explore laparotomy. Intraoperative finding included massive intraperitoneal free air and numerous air bubbles within the colon wall (Figure 2), extending from ileocolic anastomosis to descendo-sigmoid junction. Subtotal colectomy was performed for microperforation and an approximate 5 cm long small intestinal resection was done due to bowel ischemia under ICG

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image (Figure 3). Pathology report revealed presence of three polypoid lesions in colon with pneumatosis cystoides coli, as well as ischemic and necrotic small intestine without malignancy. Patient was sent to surgical Intensive Care Unit (ICU) after operation and laboratory data upon admission revealed sepsis condition with elevated lactate of 3.8 mmol/L (normal range: 0.5-2 mmol/L) and low sodium bicarbonate level of 15.1 mmol/L (normal range: 21-28 mmol/L). Hence, empirical antibiotic ertapenem was prescribed. His sepsis progressed to shock status on Post Op Day (POD) 3, where vasopressor norepinephrine was prescribed and ertapenem was shifted to Piperacillin/Tazobactam according to ascites culture result. According to his history, interstitial pneumonitis is suspected to be associated to Nivolumab so methylprednisolone was prescribed. Due to unsuccessful extubation, patient was on ventilator and was hospitalized. On POD 60, massive hematochezia and anemia was noted. Emergent abdominal CT revealed hemorrhage in rectum but without active bleeding. Diagnostic angiography also gave negative result. Sigmoidoscopy revealed anastomosis site necrosis with lumen narrowing and much blood clot. Pathology of the necrotic tissue biopsy revealed chronic inflammation with focal granulation tissue. Patient's condition continued to deteriorate and expired on POD 81 due to septic shock.

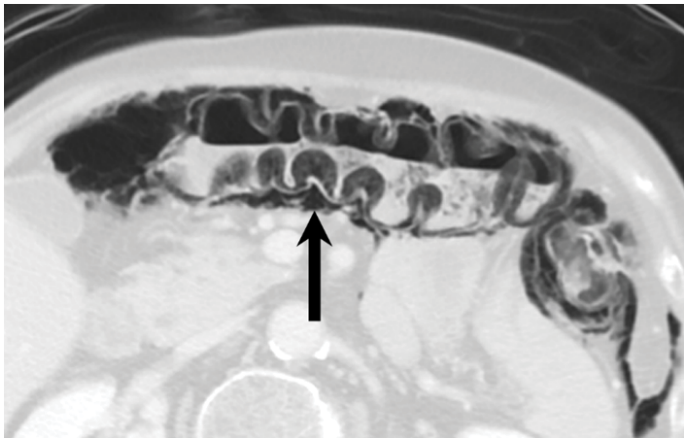

Figure 1. Abdominal CT with Pneumatosis cystoides coli.

Figure 2. The surface of transverse colon.

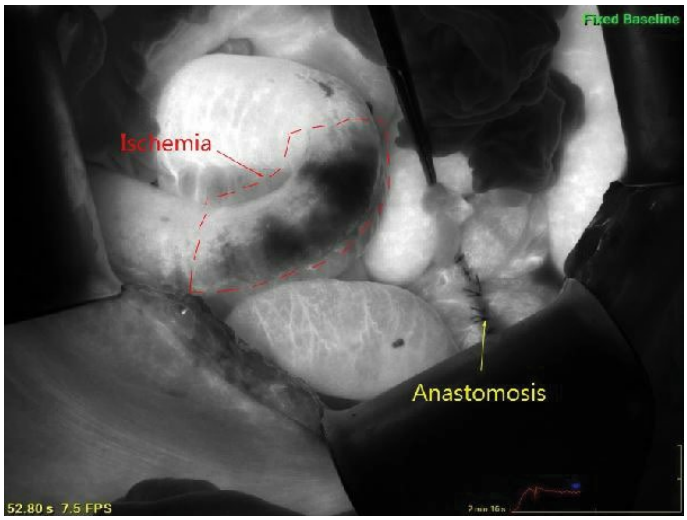

Figure 3. Perioperative ICG image with bowel ischemia.

Discussion

Pneumatosis cystoides intestinalis is rare but should not be overlooked. Nowadays its incidental finding rates had shown to increase as radiological exam becomes more readily available. Retrospective studies showed inconsistency in male-to-female ratio, ranging from 1:1 to 3.5:1. Recently, Sato T, et al [5]. divided this radiographic finding into asymptomatic PCI and pathologic PCI, with male-to-female ratio 2:1 and 0.4:1, respectively. However, the actual incidence and gender ratio is unknown due to underestimated asymptomatic PCI [1-5].

The pathophysiology of PCI is unclear but three hypotheses have been proposed: (1) mechanical theory: overpressure of gastrointestinal gas causing mechanical damage and mucosal rupture, leading to gas migrate into intestine wall; (2) pulmonary theory: in chronic pulmonary disease, air flows out when alveolar ruptures and migrates along aorta and mesenteric blood vessel into intestinal wall; (3) bacterial theory: aerogenic bacteria migration across mucosal barrier and ferments gas in intestinal wall; this is supported by the theory of counter perfusion supersaturation [6,7].

Diabetes medicine, alpha-glucosidase inhibitor, has been reported to be associated with asymptomatic PCI due to suppression of carbohydrate absorption, augmenting intestinal gas production by bacteria [8]. Therefore, prophylactic symbiotics was used in all patients to avoid bacterial overgrowth in Sato T, et al. study [5] though diabetes has not been shown to be correlated with asymptomatic PCI; it is associated with cardiovascular disease risk, including bowel ischemia. Thus, diabetes is one risk factor of symptomatic PCI. Other medications associated with PCI were corticosteroids and chemotherapy. Corticosteroid causes depletion of lymphoid tissue in Payer's patch, lessening resistance for intraluminal gas migration into bowel wall after mucosal disruption [8]. Chemotherapy also elicits gastrointestinal damages, especially PCI and perforation because it interrupts the cell cycle, inducing apoptosis and inhibiting regeneration, which results in loss of mucosal integrity. Molecular target agents, such as tyrosine kinase inhibitor, cause gastrointestinal symptoms. 15% of patients using immune checkpoint inhibitor, such as nivolumab and pembrolizumab, have perforated colitis or enterocolitis that requires ICU admission [9].

PCI presentation can range from asymptomic to acute abdomen. Wu LL, et al. [10] found common initial symptoms in Chinese patients were abdominal pain (53.9%), diarrhea (53%), distension (43.4%), nausea and vomiting (14.3%), bloody stool (12.9%), mucous stool (12%), and constipation (7.8%). However, these are nonspecific.

Nowadays, advancements and conveniences in abdomen CT make PCI more prevalent. Distinguishing pathologic etiology from benign ones is problematic, posing difficulties in subsequent treatment management. In cancer patients, it is more challenging because multiple treatment modalities that is associated with PCI, especially corticosteroids and chemotherapy. For PCI treatment, the uttermost important management is deciding on operation timing. Therefore, many recent studies focused on risk factors related to pathologic etiology that require surgical management. CT findings of cancer patients that give rise to worrisome PCI include portomesenteric venous gas, small bowel dilation >3 cm, large bowel dilation >6 cm bowel dilatation, bowel wall thickening (>0.5 cm), ascites and mesenteric stranding. PCI confined to different location provides hints about its cause. 68% of colon PCI, commonly the right colon, are benign cases, whereas small bowel PCI may be diseased, such as presence of gastrointestinal cancer though this cause is not statistically significant [11]. Linear air pattern on CT is associated with bowel obstruction,

volvulus, intussusception, hemorrhage, and bowel infarction in approximately 90% of cases [12].

Whilst PCI may occur anywhere along the gastrointestinal tract, it is usually seen in the intestine. The distribution of PCI is 46% - 78% in colon, 20% - 27% in small intestine and 2% - 7% in both parts [3,13]. About 80% of asymptomatic PCI is noted in the ascending colon, whereas 41% of pathologic PCI is noted in the jejunum and 82% of pathologic PCI is noted in the ileum. The presence of ileal PCI with ascites has 82.4% sensitivity and 87.1% specificity indicating pathologic PCI [5]. Other strong pathologic etiology indicators, even in patients without peritonitis, include decreased hemoglobin, elevated INR, and lactate level of 2 mmol/L.

PCI is a potential life-threatening condition. Mismanagement can result in devastating result. Due to rarity of PCI, management is traditionally guided by speculation rather than by statistically conclusion. Conservative management in non-operative patients include medical therapies with oxygen supply, hyperbaric therapy, antibiotics and special element diets. In a large series of adult cases, patients who were septic at the diagnosis of PCI had high risk of poor outcome regardless of surgical status. Concomitant presence of emesis, WBC >12000/mm^3, age greater than 60 year-old or CT with portal venous gas favored surgical management [14]. Lee GS, et al. [15] developed a simple radiologic scoring system for predicting mortality and may provide information on preoperative management. A decreased or absent of bowel wall enhancement on CT along with signs of peritoneal irritation predicts mortality risks of 57.7% on day 1 and 59.9% on day 7 after diagnosis. When radiologic image shows no evidence of perforation and portal venous gas, but there is presence of severe clinical signs such as peritoneal irritations, tachypnea and fever, whether to undertake conservative management or surgical intervention is a dilemma. If benefits outweigh the risks upon suspicion, diagnostic laparoscopy should be considered to rule out bowel perforation and ischemia [14].

Comorbidities associated with death were immunosuppression and liver disease. Mortality risks include clinical findings of mesenteric ischemia, hypotension, abdominal tenderness, and peritoneal irritation signs. Laboratory data of serum creatinine ≥ 1.5 mg/dL, albumin < 3 g/dL, lactate ≥2 mmol/l, and bicarbonate <20 mmol/L are also associated with high mortality. CT Image findings indicating high risk in death include presence of PCI in both small bowel and colon, involvement of two segments or more, presence of portal venous gas, presence of free peritoneal fluid, curvilinear shape of the intramural gas and decreased or absent bowel wall enhancement [14,15].

Conclusion

In conclusion, for acute abdomen or sepsis condition, operation should be arranged due to suspicion of ischemia or necrosis. Laparoscopy can be considered if certain risk factors are presence, including high lactate, low sodium bicarbonate, sepsis, small intestine involvement, the use of alpha-glucosidase inhibitor, chemotherapy and corticosteroid, along with abdominal CT showing only PCI in the absence of ascites or portal venous gas. If ischemic colon is noted, small intestine should be assessed carefully, especially the ileum, due to higher possibility of small intestinal pathologic PCI.

Acknowledgement

Availability of data and material

Data related to this case report will become available upon request, if needed.

Author's contribution

The authors that contributed to the study conception and design were Tsung-Jung Tsai and Yu-Yao Chang. Data acquisition, interpretation and first draft of the manuscript were performed by Tsung-Jung Tsai. All authors commented on previous versions of the manuscript and revised it critically for important intellectual content. All authors read and approved the final version of the manuscript to be submitted.

Compliance with Ethical Standards

Conflicts of interest

The authors declare that they have no conflict of interest.

Ethics approval

No ethical approval is required according to institutional policies.

Consent to participate

All patient-related info was de-identified so patient consent was waived.

Consent for publication

All patient-related info was de-identified so patient consent was waived.

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