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CASE REPORT

Pneumatosis intestinalis – an illusive disease

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Summary
Pneumatosis intestinalis (PI) is characterised by pathological gas infiltration into the submucosa and subserosa of the gastrointestinal tract, sometimes with an unclear pathogenesis. The clinical presentation of PI varies, with the diagnosis established via computed tomography (CT), where PI manifests as linear or bubbly gas patterns within the bowel wall. Management often necessitates surgical intervention to address potential life-threatening causes like mesenteric ischemia or bowel necrosis. This case report discusses a 69-year-old male who presented with abdominal pain and distension alongside worrisome radiological features indicative of extensive PI, who underwent an exploratory laparotomy that revealed no pathological findings and with an eventual uneventful recovery.
Keywords: pneumatosis intestinalis, intestinal ischemia, portomesenteric pneumatosis, computed tomography

Case report

A 69-year-old male patient presented to the emergency department of our hospital with complaints of progressively worsening abdominal pain, accompanied by abdominal distension and an absence of bowel movements over the preceding five days, while still passing flatus. On further enquiry, he admitted to an initial episode of non-bloody, non-mucoid diarrhoea, followed by a brief period of normal stool passage, after which constipation ensued. The patient's medical history was notable for tobacco use and arterial hypertension controlled using a single agent (angiotensin receptor blocker). The patient was afebrile with normal vital signs and his abdominal examination revealed diffuse tenderness, with no signs of peritonitis. Laboratory analyses revealed a white blood cell count of 16 000/microliter, comprising 77% neutrophils, a C-reactive protein level of 48 mmol/dL and lactate dehydrogenase (LDH) of 180 IU/L. Coagulation values and renal and liver function tests were within normal limits. Arterial blood gas analysis on room air indicated a pH of 7.37, pO_2 of 87 mmHg, pCO_2 of 40 mmHg, bicarbonate (HCO_3) of 22 mmol/L and a serum lactate concentration of 1.5 mmol/L.

A three-phase computed tomography (CT) of the abdomen with intravenous contrast revealed diffusely oedematous bowel loops and intramural air extending through at least 100 cm of proximal jejunum (Figure 1a). Air was observed within the portal venous system, extending to the mesenteric vein and left hepatic vein (Figure 1b). During the arterial phase of imaging, an atheromatous stenosis of up to 50% was identified at the ostium of the superior mesenteric artery

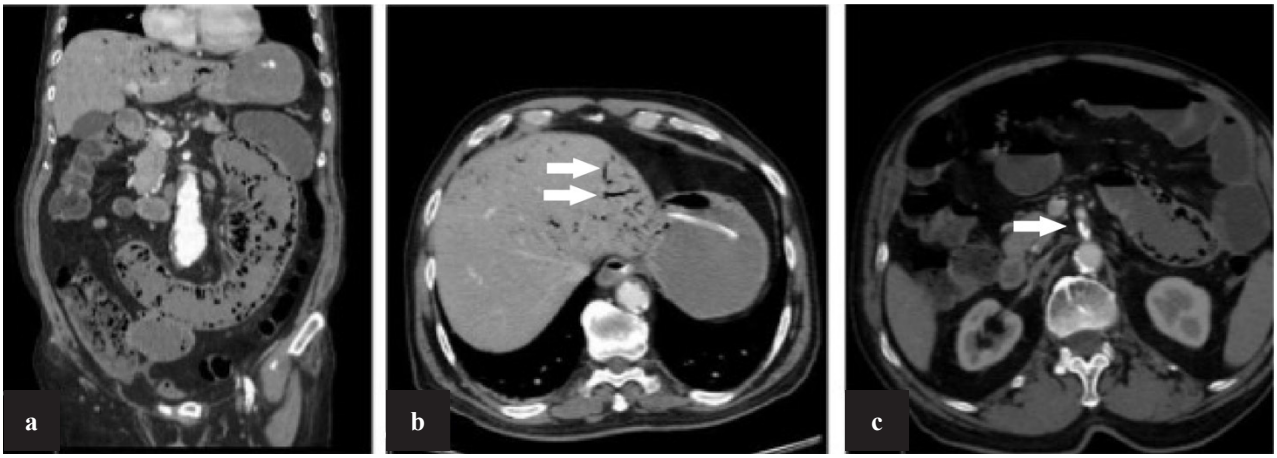

Figure 1: CT of the abdomen with intravenous contrast – 1a. Oedematous bowel loops with presence of intramural air for at least 100 cm of jejunum; 1b. Presence of air in the portal system (white arrows); 1c. Atherosclerosis of the origin of SMA (white arrow)

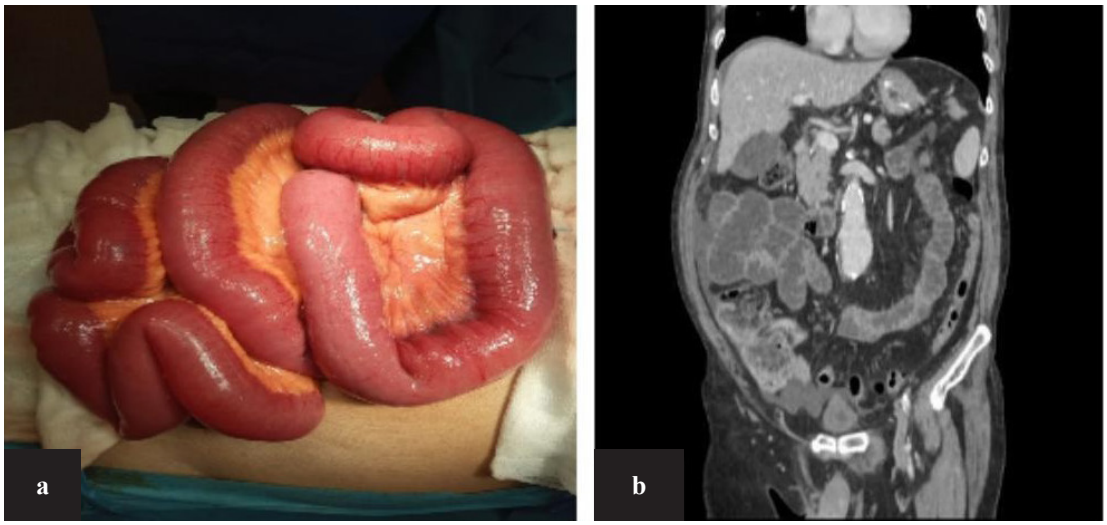

Figure 2: 2a. Intraoperative image of the bowel with no signs of ischemia or other pathology; 2b. Postoperative follow-up abdominal CT with regression of intramural and portal venous system air

(SMA), whereas the inferior mesenteric artery was poorly visualised (Figure 1c).

Given the severity of the radiological findings, an exploratory laparotomy was deemed imperative to investigate the possibility of bowel ischemia. Upon examination of the abdominal cavity, no ischemic bowel changes or other pathological findings were observed (Figure 2a). A vascular surgeon was consulted to provide expertise on vascular integrity and to assist with any necessary vascular assessments. The patency of the SMA was assessed through direct visual and tactile evaluation. The absence of ischemic bowel changes suggested adequate collateral blood flow, thus no SMA pressures were measured, and no further surgical interventions were undertaken. Postoperatively, the patient was managed with a regimen of bowel rest, intravenous administration of piperacillin-tazobactam for antimicrobial coverage for a duration of 7 days, and anticoagulant therapy to mitigate the risk of thromboembolic events. Repeated blood cultures were taken during the patient's hospital stay, all of which returned negative results. The postoperative period was uneventful, with a gradual reintroduction of oral intake. The patient began passing stools on the third postoperative day, initially soft and later normal. A follow-up abdominal CT scan, conducted on the seventh postoperative day, demonstrated a resolution of the previously noted intramural and portal venous air (Figure 2b). The patient was discharged on the 10th postoperative day, remaining asymptomatic at 8 months of follow-up.

Discussion

Pneumatosis intestinalis (PI) is a rare condition that is defined as pathological gas infiltration into the submucosa and subserosa of the gastrointestinal tract. It has been found to affect 0.03% of the population.[1] The pathophysiology of PI remains elusive, with the most prominent theories suggesting a multifaceted interplay involving increased luminal or intramural pressure (mechanical theory), bacterial invasion and intramural gas production (bacterial theory), and the migration of gas from the pulmonary system (pulmonary gas theory).[2] It affects patients with a mean age of 56.6 ± 19.4 years with a male to female ratio of 1.13:1.[3] The clinical presentation of PI varies, with abdominal distension being the most observed sign at 54%, followed by constipation and peritonitis, reported at 19% and 17%, respectively.[3] It is noteworthy that, in a large multicentre study, a significant proportion of patients, (21%) were unable to undergo comprehensive evaluation at the time of diagnosis due to factors such as sedation or altered mental status.[3] PI exhibits a preference for certain regions of the gastrointestinal tract, with the colon being the most frequently involved site at 57%, followed by the ileum and jejunum, at 30.7% and 30.5%, respectively.[3]

The diagnosis of (PI) is primarily established through CT, where PI characteristically manifests as low-density linear or bubbly gas patterns within the bowel wall.[4] CT imaging not only helps in distinguishing PI from other radiographic findings, but also aids in identifying underlying causes. Additional CT findings such as bowel wall thickening, either absent or intense mucosal enhancement, bowel dilation, arterial or venous occlusion, ascites, and hepatic portal or portomesenteric venous gas should raise the suspicion of PI stemming from an underlying life-threatening cause.[1] The differential diagnosis of underlying conditions presenting with PI mainly involves intestinal ischemia and bowel necrosis but also includes a variety of pathological conditions such as inflammatory bowel disease, malignancy, chemotherapy, immune deficiencies, trauma, or medical procedures.[5] It is noteworthy that in our case clinical and radiographic findings suggesting severe pathology were in contrast to unremarkable operative findings and the patient's uneventful clinical course. The radiographic evidence of SMA stenosis in our patient may have contributed to the development of PI by means of a transient bowel ischemia superimposed on chronic mesenteric ischemia. The presence of collateral circulation possibly facilitated the prompt resolution and favourable clinical outcome observed in this patient.

The management of PI poses significant challenges, especially when it comes to the critical decision regarding vascular and/or surgical intervention. To assist clinicians in this decision-making process, several predictive factors for bowel compromise and a worrisome patient course have been proposed.[3,6-8] A recent review by Tropeano et al. categorised risk factors based on their correlation with the presence of bowel ischemia at surgical exploration (Table I).[8]

Table I: Risk factors associated with bowel compromise. Adapted from Tropeano G, Di Grezia M, Puccioni C, et al.[8] (reprinted with written permission from the authors)

Risk factors	
Systemic	Hypotension or vasopressor use
	Acute renal failure
	Requirement for mechanical ventilation
Clinical signs	Peritonitis
	Guarding or rebound tenderness
	Emesis
Anamnestic	Vascular disease
	Atrial fibrillation
Major laboratory risk factors	Lactate > 4 mmol/L
	LDH > 400 UI/L
	Blood Urea Nitrogen > 50 mg/dL
	pH < 7.31
Minor laboratory risk factor	WBC > 15.000/L
	Creatinine > 2 mg/dL
	HCO_3- < 18 mmol/L
	Potassium > 5.5 mmol/L
Radiological	Portomesenteric pneumatosis
	Free peritoneal fluid
	Pneumoperitoneum

Based on these factors, treatment algorithms have been proposed primarily aiming at identifying patients with life-threatening underlying conditions associated with PI.[2,6,8] These cases, representing 40% of total PI cases, are associated with a significantly adverse prognosis, with a reported mortality rate of up to 72%.[3,9] For cases deemed benign, a conservative treatment approach is recommended, which includes observation, hyperbaric oxygen therapy, and antibiotic administration.[10] Hyperbaric oxygen therapy, in particular, is noted for its efficacy in alleviating symptoms by lowering the partial pressure of gases in the venous system, facilitating gas expulsion from the bowel wall, and inhibiting the growth of anaerobic bacteria in the intestinal tract.[11] In situations where non-surgical management is the initial course of action, it is crucial to closely monitor and continuously reevaluate the patient for response to treatment, with consideration for exploratory surgery in the absence of improvement or a deterioration in the patient's condition.[12] For cases where surgical treatment is indicated, laparotomy allows for a thorough exploration of the abdominal cavity, providing direct and immediate access to the abdominal organs for rapid intervention. This approach offers valuable tactile feedback, which is crucial for assessing the viability of the affected bowel and the patency of vessels. However, laparotomy has certain drawbacks, including the risk of an extended hospital stay, increased postoperative discomfort, and a longer recovery period compared to laparoscopic exploration. Despite these risks, the superior access and feedback provided by laparotomy were deemed essential in this case for effective management.

Conflict of interest

The authors declare no conflict of interest.

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Ethical approval

Informed written consent was obtained from the patient for being included in the study.

This article does not contain any studies with human or animal subjects.

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