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An approach to pneumatosis intestinalis: Factors affecting your management



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ABSTRACT

Pneumatosis Intestinalis (PI) is defined as the presence of extra-luminal gas confined to the bowel wall. PI is an ominous condition often requiring emergent surgery. The management can be challenging in some circumstances, as the choice of surgery versus medical treatment can be difficult.

In this study, we first report the case of a seventy-seven year old woman presenting to the emergency department with the presence of PI on computed tomography of the abdomen. Secondly, we review the existing literature regarding the management of PI and we suggest a treatment algorithm based on clinical, laboratory and radiological findings.

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1. Introduction

In 1730, Pneumatosis Intestinalis (PI) was first described in the literature by Du Vernoi as the presence of gas within the wall of the small or large bowel [1]. The exact prevalence of this pathology in the general population is not known. One autopsy series reported the prevalence of PI as 0.03% in the general population [2]. However, most clinicians believe this to be an underestimate as a proportion of cases are asymptomatic and thus go unreported. PI can be subdivided into two distinct groups: Primary PI, representing 15% of cases, and secondary PI, representing 85% of cases [3]. In Primary PI, the intramural gas is cystic and benign in nature, in contrast to secondary PI where the gas accumulates as linear collections and reflects a pathological condition [4]. Secondary PI has been attributed to endoscopic procedures, immunological disturbances, bowel mucosal disruptions and intra-abdominal pathologies.

It is often difficult to differentiate primary from secondary PI, as they can present with similar symptoms. However, management of primary and secondary PI differ; primary PI requires medical treatment while secondary PI requires surgical intervention. In this study, we evaluate factors that affect management of PI. We first report the case of a seventy-seven year old woman presenting to the emergency department with the presence of PI on computed

tomography of the abdomen. Secondly, we review the existing literature regarding the management of PI and we suggest a treatment algorithm based on clinical, laboratory and radiological findings.

2. Presentation of case

Mrs. M is a 77 year-old woman who presented to the emergency department (ED) because of longstanding abdominal pain with intermittent vomiting, which had worsened that day. On history, she is known for panhypopituitarism secondary to Sheehan Syndrome, hypertension and a recent diagnosis of gastro-esophageal reflux disease (GERD). She had no surgical history.

On arrival to the ED, the patient was hemodynamically stable and afebrile. Abdominal exam showed a distended and tympanic abdomen without tenderness or guarding. Her laboratory tests were unremarkable except for an elevated serum lactate, of 2.1 mmol/L. To further investigate this pain, an abdominal series was requested to rule out an abdominal obstruction. The X-rays showed several air-fluid levels with slightly dilated small bowel (Fig. 1). A computed tomography (CT) of the abdomen with intravenous (IV) contrast was then requested in order to identify the cause of this possible small bowel obstruction. The CT scan showed the presence of mild to moderate small bowel obstruction with a transition point toward the mid bowel. There was extensive pneumatosis intestinalis involving small bowel distal to the transition point. The CT scan confirmed patent arterial vascularity and a small amount of gas was noted in the portal venous system (Fig. 2). Of note, a CT abdomen with IV contrast taken 15 months prior to rule out obstruction was unremarkable. Given the patient's pain, the new findings on her CT and her slightly elevated lactate, the

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Fig. 1. Abdominal X-ray series.

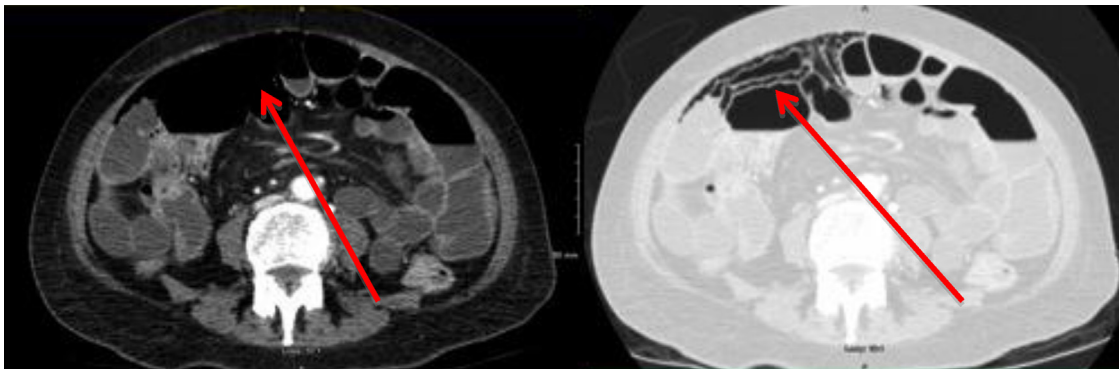


Fig. 2. CT-scan abdomen C+ (axial)–pneumatosis intestinalis.



Fig. 3. Intra-operative finding of PI.

decision was made to proceed with an exploratory laparotomy. Our intraoperative findings showed extensive mesenteric emphysema in a large part of the small intestine with no small bowel compromise and no identifiable transition point (Fig. 3).

Given there was no sign of bowel compromise, it was decided that small bowel resection was not necessary and the incision was closed. Post-operatively, the patient failed to improve, complained of abdominal pain and was eating a minimal amount. The gastroenterology service was consulted for their opinion, but no cause was identified to explain patient's symptomatology. The patient refused all medical interventions and she was in a decompensated physical

state. After discussion with the family, the treating team decided to apply her to a long-term care facility. From the time of the application she continued to deteriorate; she became increasingly frail, and about six weeks post-operatively she passed away.

3. Discussion

The pathophysiology of PI remains controversial and as such, its management can be challenging for clinicians. Emergent surgery is required in as many as 66% of cases [5]. Hence, for the treating team, the most important step in managing a patient with PI is

Table 1
Clinical & laboratory findings associated with morbidity when PI present.

Study	Acute abdomen	Hypotension	Elevated lactate	Bicarbonate ≤ 20 mmol/L	Age ≥ 60	Elevated creatinine
Lee [15]	4.73 ^a (1.79–12.45)	3.79 ^a (1.67–8.60)	–	2.54 ^a (1.21–5.32)	–	2.43 ^a (1.17–5.05)
DuBose [6]	4.7 ^b (2.2–10.3)	5.1 ^b (2.0–13.0)	4.3 ^b (2.2–8.7)	–	–	3.4 ^b (1.5–7.8)
Duron [16]	9.35 ^b (1.85–47.14)	4.38 ^a (1.52–12.65)	2.29 ^b (1.17–4.51)	–	2.88 ^a (1.27–6.50)	–
Wayne [17]	9.5 ^a (1.7–52.1)	–	13.8 ^a (1.6–122.0)	–	–	–
Greenstein [8]	–	7.0 ^a (1.2–40)	9.0 ^a (1.1–71)	6.7 ^a (1.2–3.6)	18.4 ^b (1.3–261)	–
Hawn [18]	–	–	30.37 ^b (7.31–126.20)	–	–	3.05 ^a (1.25–7.42)

Odds ratios and 95% CI.

^a Univariate analysis.

^b Multivariate analysis.

the decision to operate. Many studies have investigated the use of risk factors as predictors of compromised bowel and the need for surgery.

In 2013, the Eastern Association for the Surgery of Trauma (EAST) pneumatosis study group published the largest retrospective multicenter study to date involving 500 patients diagnosed with PI and explored different risk factors for pathologic PI. In their study, they identified risk factors, which best predicted pathological PI. The risk factors included the presence of hypotension or

vasopressor use, peritonitis, a lactate level >2 mmol/L, acute renal failure, and active mechanical ventilation [6]. A memorial Sloan-Kettering cancer center study published in 2013, analyzed risk factors classifying oncology patients having PI as “worrisome” or “benign”. Patients were placed in the worrisome group according to previously studied laboratory and clinical risk factors including: elevated serum lactate or the presence of guarding or rebound tenderness on physical exam. Forty four percent of the cohort (37 patients) were designated as worrisome according to these fac-

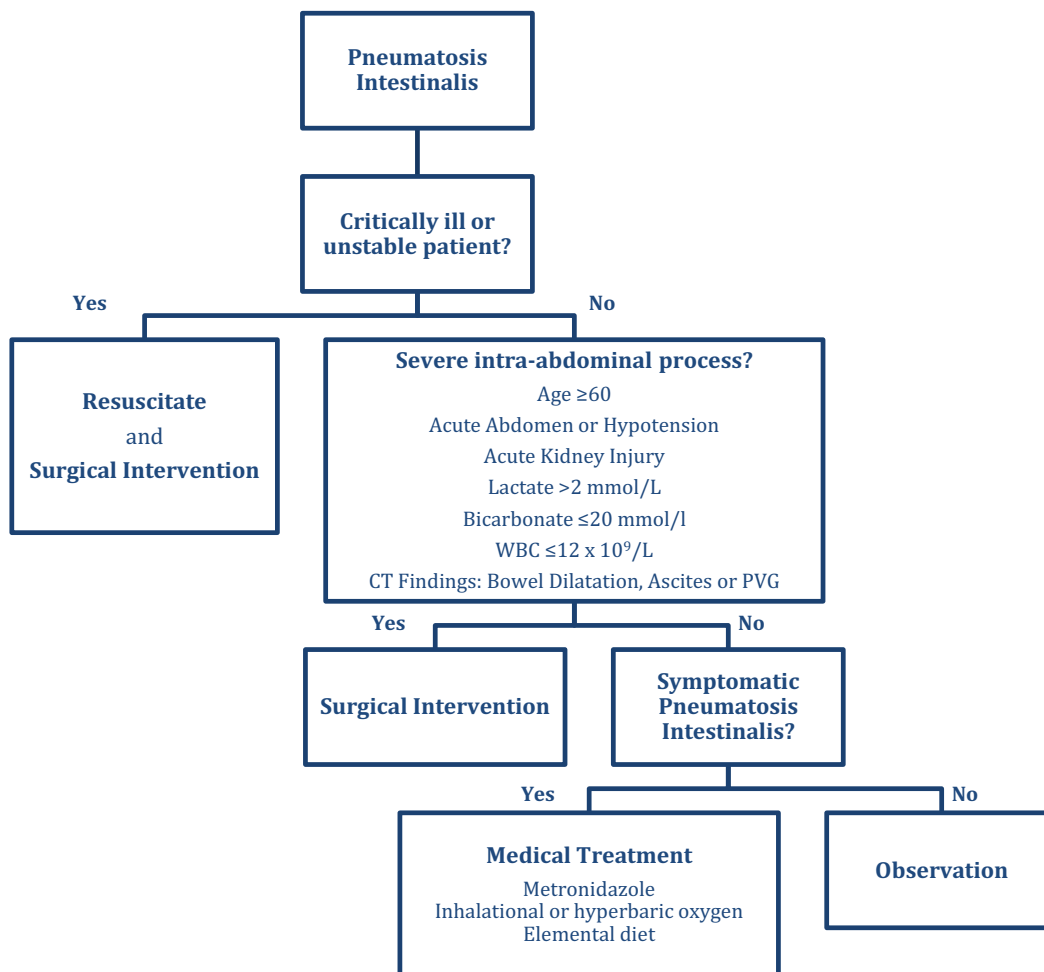


Fig. 4. Treatment algorithm for PI.

Table 2
Radiological factors associated with morbidity when PI present.

Study	PVG	Bowel dilatation	Ascites
Lee [15]	3.71 ^a (1.78–7.71)	–	4.80 ^a (1.67–13.80)
Duron [16]	3.19 ^a (1.29–7.87)	13.19 ^b (2.81–61.93)	2.21 ^a (1.06–4.62)

Odds ratios and 95% CI.

^a Univariate analysis.

^b Multivariate analysis.

tors, of which 86% (32 patients) were surgically and pathologically proven to have bowel perforation, bowel infarction, colitis, and/or bowel obstruction. CT findings that most significantly correlated with worrisome PI included small (>3 cm) or large (>6 cm) bowel dilatation, bowel wall thickening (>0.5 cm), mesenteric stranding, ascites and PVG. Interestingly, pneumoperitoneum was present in greater than 50% of either group and did not inherently suggest a life-threatening process [7]. In 2007, a retrospective study of 40 patients at Mount Sinai Medical Center had investigated several of the same factors associated with surgical management later studied by the EAST group in 2013. Risk factors that were associated with pathological process were presence of PVG, age ≥ 60 years, emesis, bicarbonate ≤ 20 mmol/L and WCC $\geq 12 \times 10^9/L$ (OR 3.6). On multivariate analysis however, none of the factors presented by the EAST group approached statistical significance (8). Those studies aimed to help clinicians identify patients who need surgery when presenting with PI. In these studies, the most common factors associated with the need to undergo surgery were: (1) presence of hypotension, (2) presence of an acute abdomen on physical exam, (3) elevated serum lactate, (4) low serum bicarbonate, (5) elevated white cell count, (6) age ≥ 60 years (7) acute rise of creatinine, and (8) presence of ascites, portal venous gas or bowel dilatation on CT-scan of the abdomen (Tables 1 and 2).

Once the need for emergent surgical intervention is ruled out, according to the patient's clinical condition and predictive risk factors, a trial of medical treatment can be attempted. Antibiotics have been investigated as a mainstay treatment for PI. One regimen, which has proven to have some success, is metronidazole 500 mg per os (PO) three times daily (TID) for up to 3 months [9]. It is theorized that antibiotics reduce the amount of gas produced by bacteria and alleviate obstructive symptoms.

Also, inhalational oxygen therapy is believed to alleviate symptoms by acting as a toxin to anaerobic bacteria in the gut [10]. In primary PI, the oxygen content of cysts is typically low and the hydrogen content can be as high as 50%. It is theorized that inhalational oxygen or hyperbaric oxygen therapy can decrease the partial pressure of non-oxygen gases in the venous system and promote diffusion of gases out of the bowel wall. Hyperbaric oxygen treatment can be used to reduce oxygen toxicity in patients by using 2.5 ATM of oxygen 90 min a day, continued until 2 days after the disappearance of cysts, to reduce the risk of recurrence [11–13].

Finally, an elemental diet has also been proposed as a treatment for symptomatic PI. Theoretically, elemental diet should be totally absorbed in the small intestine and thus decrease production of gas by colonic flora. One case report showed that two patients with incapacitating symptoms of one year duration, unresponsive to other treatments, had complete resolution of cysts on colonoscopy within two weeks and symptomatic relief in as little as four days following an elemental diet [14].

4. Conclusion

When managing a patient with PI, their risk for bowel wall compromise should be assessed according to the presence of acute

abdomen, hypotension, elevated serum lactate, white cell count or creatinine, low serum bicarbonate, age above 60 and complemented by radiological findings such as bowel dilatation, ascites or PVG. In the presence of these factors, surgical exploration is warranted. In the absence of these red flags, a regimen of antibiotics, elemental diet and oxygen therapy can be attempted to relieve symptoms. In this study, we present a management algorithm to assist clinicians in determining if emergent surgical intervention is necessary (Fig. 4).

Conflict of interest

Nothing to declare.

Sources of funding

Nothing to declare.

Ethical approval

Not applicable.

Consent

Written consent was obtained.

Authors contribution

Mehdi Tahiri: Data collection, data analysis for the review of the literature. Writing of the paper. Jordan Levy: Data collection, data analysis for the review of the literature. Writing of the paper. Saud Alzaid: Data collection, data analysis for the review of the literature. Writing of the paper. Dawn Anderson: Supervision of the project mainly on the correction and helping of the paper.

Guarantor

Mehdi Tahiri MD.

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