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Case report

A case report and approach to management in pneumatosis intestinalis☆

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ABSTRACT

Pneumatosis Intestinalis (PI) is an uncommon condition in which there is gas present within the wall of the gastrointestinal tract. PI is usually found in the large bowel, but can less commonly occur in the small bowel, and gas may be present in either the subserosal or submucosal layer of the intestine. Its unfamiliarity often means it is under-recognised and not considered as a differential diagnosis when assessing a patient with abdominal symptoms.

The spectrum of conditions that produce PI is varied and ranges from the non-urgent to life-threatening. Early appreciation of the overall clinical picture is therefore paramount to enable the practitioner to distinguish between the benign to the fatal cases of PI and enable precise decision-making regarding its management.

The challenge facing the clinician is twofold; firstly the accurate identification of the diagnosis of PI, as opposed to other causes of peritoneal gas and secondly judging whether operative or non-operative management should ensue. We present a case of a patient presenting on two separate occasions with PI, abdominal symptoms and radiological signs of acute abdominal pathology which demonstrates the wide spectrum of difficulties faced with this uncommon condition.

1. Introduction

Pneumatosis Intestinalis (PI) is a rare condition with an incidence of 0.03% worldwide [1] and refers to the presence of gas within the wall of the intestine. PI was first reported in 1754 [2] and ranges from an incidental finding with no clinical symptoms, to a life-threatening condition with peritonism [2]. The aetiology is unclear but is thought to be multi-factorial in origin. Two main theories have been proposed. A mechanical theory suggests that intestinal obstruction increases luminal pressure which in turn allows gas to enter the submucosal space. A bacterial theory suggests that gas-producing bacteria, particularly anaerobes, invade the submucosal layer and produce gas within the intestinal wall. In addition, it is recognised that long-term corticosteroid therapy can induce atrophy and fibrosis of the intestinal mucosa [3].

PI tends to be associated with other conditions including pulmonary disease, inflammatory bowel disease, diabetes, drugs (for example steroids and chemotherapeutic agents), and collagen vascular disease such as systemic sclerosis. The severity varies depending on the underlying condition but it is accepted that PI associated with collagen disease, particularly systemic sclerosis, has an increased risk of perforation with resultant poor outcome [4]. Systemic sclerosis is an autoimmune disease of the connective tissue and the systemic manifestations are diverse. Although death normally results from involvement of the heart, lungs and kidney, as many as 90% have some degree of gastrointestinal involvement [5].

The management of PI is not well documented. Operative intervention had been considered the mainstay of management [6], although recently there has been a trend to a more conservative approach, as patients will often recover with non-surgical management [7,8]. The challenge for the clinician is to identify those patients in whom non-operative management is failing and those with peritonitis or abdominal sepsis who require surgical intervention. Significantly, the level of serum lactate acid appears to be a prognostic predictor; patients who have levels of > 2mmol/L have an overall mortality rate of greater than 80% [9]. We present a case of a patient presenting on two separate occasions with PI, abdominal symptoms and radiological signs of acute abdominal pathology which demonstrates the wide spectrum of difficulties faced with this uncommon condition. This work has been reported in line with the SCARE criteria [10].

1.1. Presentation of case

A 69 year-old gentleman was referred to clinic with a paraumbilical hernia. In addition, he reported worsening abdominal distension, diarrhoea and weight loss of 9.5 kg over a 3–4 month period. On

☆ This Case Report has been reported in line with the SCARE criteria and guidelines.

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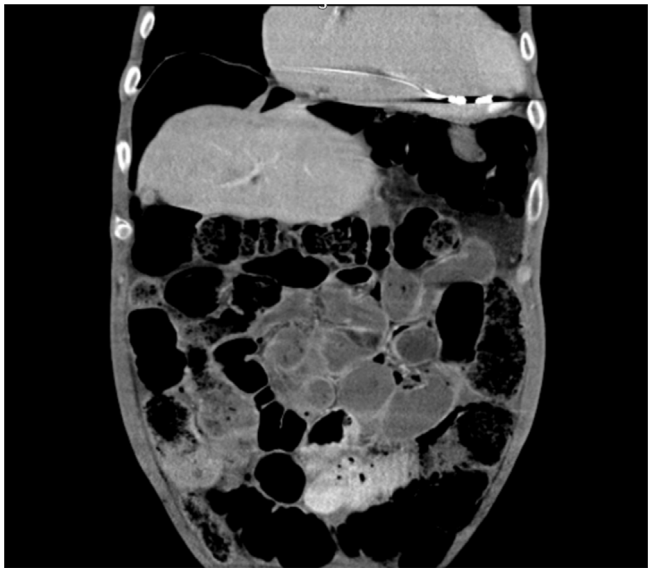

Fig. 1. CT scan showing free air and fluid.

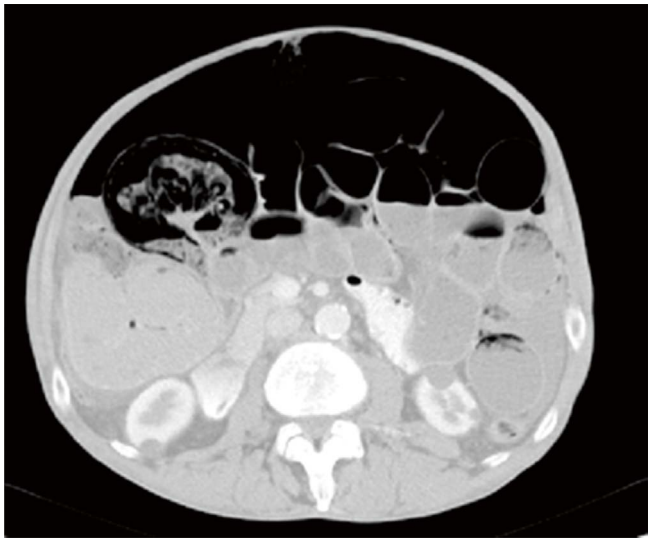

Fig. 2. CT scan showing possible area of bowel necrosis.

examination, he had a markedly distended, tympanic abdomen and a 2cm reducible paraumbilical hernia. His relevant medical history included lung fibrosis, obstructive airway disease, atrial fibrillation, cardiomyopathy, heart failure (treated with a biventricular pacemaker) and systemic sclerosis, which was treated with steroids and methotrexate. Given the atypical presentation of a paraumbilical hernia, an urgent CT (Fig. 1) was arranged. This showed small loops of distended bowel with free fluid and air in the abdomen and pelvis, indicative of perforation with a possible area of ischaemic bowel necrosis, likely secondary to ischaemia (Fig. 2). The patient was then admitted for further assessment. He remained cardiovascularly stable with no change to his clinical examination; haematological investigation was unremarkable apart from a slightly elevated CRP of 34 (n $<$ 5 mg/L) and an arterial lactate of 1.2 mmol/L (n $<$ 2 mmol/L). Given the CT findings of a small bowel ischaemia and perforation, the patient underwent an emergency laparotomy.

At operation, performed by Mr Williamson, the small bowel appeared highly unusual throughout its length, with multiple thickened patches associated with pockets of subserosal gas, suggesting a diagnosis of PI (Fig. 3). Although the colon was preserved, the majority of the small bowel was affected and two areas of probable self-sealed

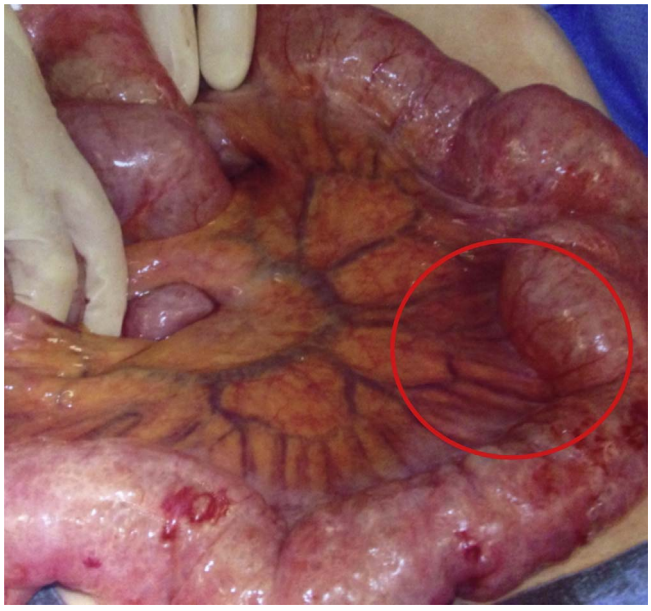

Fig. 3. Findings at laparotomy – pockets of air on surface of small bowel.

perforations were identified; it was not possible to resect these two areas and maintain a viable length of intestine in continuity, so the abdominal cavity was irrigated, drained and the hernia repaired. Postoperatively, he was admitted for a short admission to intensive care, prior to a prolonged stay on the ward for nutritional support secondary to ileus. He was discharged on oral antibiotics and had an outpatient review with the Rheumatology team; given systemic sclerosis was the likely causative factor for his PI.

Unfortunately, the patient represented one month later with a 12-h history of severe epigastric pain and metabolic acidosis (pH 7.33 and lactate of 4.6 mmol/L). On examination, he was tachycardic (106 bpm), tachypneic (respiratory rate 34) but normotensive. Repeat blood tests showed a white cell count of 14 10^9/L (n 4–11 10^9/L), haemoglobin 168 g/L (n 120–150 g/L), creatinine 133 μmol/L (n 45–84 μmol/L) and INR 5.3 (n 0.9–1.2). Urgent CT confirmed ongoing PI of the small bowel, with diffuse areas of hypoperfusion and free air within the abdomen. Management options were considered, and a conservative approach was agreed based on his previous operation findings, alongside a p-possum score reporting a morbidity of 99% and mortality of 67.5%. However, the patient deteriorated, becoming more acidotic with a pH of 7.1 and a lactate of 10.2 mmol/L, and a decision to re-operate was made, although he sadly died before this point. Post mortem findings confirmed small bowel perforation secondary to PI.

2. Discussion

PI is an unusual condition caused by a myriad of underlying pathophysiological processes that range from benign to life-threatening conditions. PI typically affects the large bowel (46% of cases) but can also affect the small intestine (27%); the incidence of both the colon and small intestine combined is only 7% [11]. It can be managed conservatively in well patients with no signs of peritonism or sepsis; operative intervention for colonic disease usually involves colectomy, with or without anastomoses to restore continuity. Management is not clear but it is generally accepted that asymptomatic patients should be managed in a conservative manner [11], but surgical management is an alternative depending on the extent of the affected bowel. In addition, it has been reported that operative manipulation of a bowel affected by systemic sclerosis tends to result in prolonged ileus [12] and for that reason alone, should be avoided, if possible, in this group of patients. Studies have predominantly concentrated on the management of large

bowel PI, as this is the most commonly affected, so there is little evidence in the optimal management in regards to small bowel PI, as found in this patient.

PI, although uncommon, is seen more often in patients with underlying co-morbidities. In particular, collagen vascular disease is a risk factor for developing the condition [13]. Immunosuppression seems to be the common point of most of the diseases or their treatment related with PI [14]. The gentleman discussed in this case had a background of established systemic sclerosis. In addition, he had other recognised risk factors including pulmonary disease and long-term steroids. These combined pathologies most likely predisposed him to PI. The literature also proposes PI associated with systemic sclerosis results in poorer outcomes [4]. Given this, this patient was at risk of developing complications and consequently requiring surgical management.

PI is a challenging condition to identify and manage. Although this gentleman initially presented with symptoms suggestive of bowel pathology, he was clinically stable. However, contradictory to his presentation, his initial CT scan was indicative of ischaemic changes and bowel perforation, which heavily influenced the decision to operate. A laparotomy was preferred over a laparoscopic approach; many authors advocate the use of laparoscopy when the patient is stable [14]. This would have been favourable as a more diagnostic and less invasive procedure but in view of the radiological findings there was concern regarding significant pathology. In contrast, his representation was more in keeping with underlying perforation and abdominal sepsis. The decision for expectant management during the second presentation was guided by the previous operation, which found no viable surgical options to manage the abnormalities found (apart from removal of the small bowel in its entirety). As the patient also presented in extremis and had multiple co-morbidities, there was an extremely high risk of peri-operative mortality, as indicated by his p-possum score.

These two sequential admissions illustrate very clearly how PI is difficult to assess and undertake the appropriate management strategy. Interestingly, the predictive role of lactate levels being associated with increased mortality reported in the literature would support the outcomes witnessed in this case [15,16]; the initial presentation with a lactate of 1.2 mmol/L was associated with survival, whilst the representation with a lactate of 4.6 mmol/L and subsequently > 10 was not survivable. The strength of this case report is that it describes a rare situation and shows how it is therefore prudent to ensure a serum lactate is performed with any suspected or confirmed case of PI to help guide management.

3. Conclusion

PI is a rare condition, especially when it affects the small bowel, which is often associated with underlying disease. PI presents a challenge to the clinician in terms of identification, as it has numerous vague symptoms. Due to the wide range of possible causes and outcomes from this condition, patients should be managed expectantly with operative intervention reserved for those with signs of perforation, peritonitis or abdominal sepsis. Arterial lactate seems to be a valid marker for significant disease in whom prognosis is poor.

Ethical approval

An ethical approval was not required.

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Author contribution

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Other contributors: Dr Emma Pope, Mr James Williamson.

Conflict of interests

None.

Trial registry number

N/a.

Guarantor

Dr Manon Jenkins.
Mr James Williamson.

Consent of patient

Written informed consent was obtained from the patient for publication of this case report and accompanying images.

Research registration unique identifying number (UIN)

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