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Pneumatosis Cystoides Intestinalis in A Patient with Advanced Scleroderma, A Case Report

Shamma Ahmad Al-Nokhatha*, Muddassar Ahmad, Prof Ronan Mullan

Department of Rheumatology, Tallaght University Hospital, Dublin, Ireland

***Corresponding author:** Shamma Ahmad Al-Nokhatha, PO Box.15258, Al Ain, Tawam hospital, UAE.

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Abstract

Background: *Scleroderma is an autoimmune connective tissue disease affecting multiple organ systems. Gastrointestinal features are common, ranging from mild gastroesophageal reflux disease to life-threatening bowel dysfunction. Pneumatosis cystoides intestinalis is a rare intestinal feature of scleroderma, characterized by gaseous cysts in intestinal submucosa, subserosa and surrounding tissues.*

Case presentation: *A 74-year old female fulfilling both anticentromere antibody-positive limited scleroderma and seropositive rheumatoid arthritis diagnostic criteria, presented to surgical service with nausea, vomiting, abdominal pain and constipation. Examination revealed abdominal distension without peritonism. Radiographic imaging showed volvulus with intestinal obstruction. Emergency laparotomy revealed a large portion of dilated bowel without perforation. Bowel resection with end stoma formation was performed.*

Four months later the patient was readmitted with recurrent abdominal discomfort and subacute obstruction. Abdominal CT showed both small bowel pneumatosis and residual small volume pneumoperitoneum attributed to recent surgery. Conservative management with bowel rest, fluid, electrolyte supplementation and parenteral nutrition were instituted with full resolution of symptoms. One-month later, the patient represented with a second episode of subacute small bowel obstruction. Repeat imaging showed interval progression of small bowel pneumatosis and new small bowel dilatation. Rheumatology service was contacted and the patient was managed using high-flow oxygen therapy, rifaximine to manage anaerobic bacterial overgrowth and the somatostatin analogue octreotide for intestinal pseudo-obstruction. The patient's bowel symptoms resolved and she was weaned off oxygen. To date, her symptoms have not recurred.

Conclusion: *An increased awareness of Pneumatosis cystoides intestinalis among treating rheumatologists, along with the prompt institution of conservative management strategies, can prevent the requirement for surgical intervention leading to improve long-term outcomes.*

Keywords: *Scleroderma, pneumatosis cystoides intestinalis, pneumoperitoneum.*

Introduction

Scleroderma is an autoimmune connective tissue disease affecting multiple organ systems. Gastrointestinal (GI) manifestations affect the majority of affected individuals, ranging from mild gastroesophageal reflux disease (GERD) to severe, life-threatening bowel dysfunction. While the pathogenesis of GI manifestations is not fully understood, the hallmark of scleroderma is the replacement of normal intestinal architecture with fibrous tissue resulting in disruption of peristaltic activity.1Dysfunction and failure of the lower GI tract in scleroderma is associated with a poor prognosis. Pneumatosis cystoides intestinalis (PI) is an unusual intestinal feature of scleroderma, characterized by multiple gas cysts into the diseased intestinal wall within the small and/or large intestine which can extend into mesentery of the gut or the peritoneal cavity [1,2]. Here we are reporting a case of patient with PI in the setting of scleroderma.

Case presentation

We report a 74-year-old an Irish female fulfilling diagnostic criteria for both anticentromere antibody-positive limited scleroderma (2008) and erosive seropositive rheumatoid arthritis (2013) successfully managed with rituximab

therapy [3,4] who presented acutely to surgical services with a 5-days history of abdominal pain, vomiting and absence of bowel motions. She had a background history of hiatus hernia, mild diverticulosis, emphysematous chronic obstructive pulmonary disease and atrial fibrillation.

A plain radiography abdominal film showed a large intestinal volvulus at the splenic flexure with features of intestinal obstruction. Emergency laparotomy was performed and revealed a dilated large bowel segment without perforation. The affected section was resected (760mm length, 70mm dilatation), with end stoma formation in the left lower quadrant. Intermittent postoperative obstructive symptoms resolved with conservative measures and the patient was discharged when well.

Four months later, the patient was readmitted surgically with abdominal discomfort, nausea, vomiting, and weight loss and reduced stoma output. A CT abdomen showed pneumatosis in a left quadrant small bowel loop with adjacent small vessel mesenteric venous gas. New small volume pneumoperitoneum was attributed to recent surgery. Figure1A Initial conservative management with IV fluid and NGT feeding failed, and a period of intravenous total parenteral nutrition (TPN) was required before recovery of GI function.

One month after she required a third admission for subacute bowel obstruction. CT of the abdomen showed interval progression of small bowel pneumatosis and new small bowel dilatation (Figure1 B, C). Rheumatology service was consulted and she was successfully managed for PI with high flow oxygen therapy and rifaximine to impede intestinal anaerobic bacterial overgrowth, laxatives and somatostatin analogue octreotide to promote peristalsis, IV fluid and TPN feeding. Over the following days her symptoms resolved, and she was discharged on ongoing rotational antibiotics to prevent reemergence bacterial overgrowth.

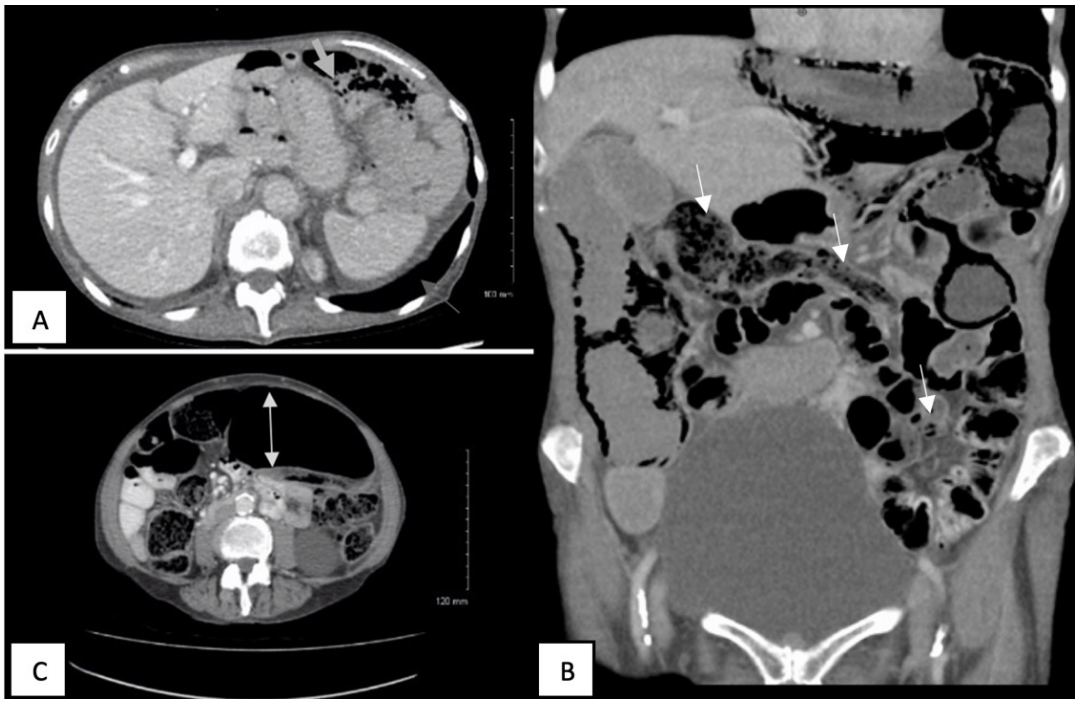

Figure 1(A): CT abdomen: Pneumatosis in a left quadrant small bowel loop with adjacent small vessel mesenteric venous gas (thick arrow). New small volume pneumoperitoneum (thin arrow). (B, C) CT abdomen: Interval progression of small bowel pneumatosis (single head arrows) and new small bowel dilatation (double headed arrow).

Discussion

PI may be defined either as primary (15%) or secondary to an underlying cause (85%). 2,5Secondary PI may occur following as a result of penetrating abdominal trauma, in the setting of GI malignancy, as a complication of surgical procedures (endoscopy, enteric tube placement for volvulus, surgical anastomosis), or in the setting of autoimmune diseases including scleroderma. While the pathogenesis of PI has not been fully established, it has been proposed that the accumulation of gas-producing bacteria in the setting of altered mucosal integrity and reduced peristaltic activity, leads to the accumulation of gas and raised intra-abdominal pressure. PI may occur in both the small and large intestine. Computer tomographic imaging remains the gold standard modality of choice in confirming the diagnosis, superior to plain film or ultrasound.

PI has no pathognomonic clinical signs, with patient presentations ranging from asymptomatic radiographic disease, to non-specific symptoms including abdominal bloating, nausea, abdominal pain or constipation. Complications occurring in 16.3% of cases include bowel distension leading to obstruction or volvulus, rupture of air-filled cysts leading to pneumoperitoneum, or intestinal perforation. A key feature of PI is the relatively benign clinical presentation despite radiographic features of free

abdominal gas or fluid more usually seen in the setting of perforated abdominal viscus or ischemic bowel [2,5,6,7].
PI is often a relatively benign condition, responding to conservative management including bowel rest, fluid and electrolyte supplementation, total parenteral nutrition, and bowel tract decompression for symptomatic relief if medical management has failed. Antibiotics may be used to eliminate gas-forming bacteria from the GI tract. Inhalational oxygen or hyperbaric oxygen therapy has been proposed as an adjunct therapy in refractory PI, where increased oxygen concentrations may act as a toxin to anaerobic gut bacteria and where altered partial pressures may promote the reabsorption of hydrogen and nitrogen from gas-filled cysts [5,8,9]. The somatostatin analogue octreotide is a prokinetic treatment used in scleroderma patients with intestinal pseudo-obstruction to improve intestinal motility and overcome the bowel stasis occurring as a result of progressive fibrosis of the GI tract.

Surgical resection of affected PI bowel in scleroderma is often not recommended due to the risk of prolonged post-surgical ileus and the diffuse nature of GI involvement [10]. Effective treatment of PI therefore requires the exclusion of other causes of pneumoperitoneum so that unnecessary procedures including laparotomy or bowel resection can be avoided.

Conclusion

GI involvement in scleroderma varies, PI has heterogeneous presentations which doesn't necessarily correlate with radiographic findings. An increased awareness of Pneumatosis cystoides intestinalis among treating rheumatologists, along with the prompt institution of conservative management strategies, can prevent the requirement for surgical intervention leading to improve long-term outcomes.

Abbreviations

- **GI:** gastrointestinal
- **GERD:** gastroesophageal reflux disease
- **PI:** Pneumatosis cystoides intestinalis
- **NGT:** nasogastric tube
- **OGD:** oesophago-gastroduodenoscopy
- **CT:** Computed tomography
- **TPN:** total parenteral nutrition

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

Competing interests
The authors declare that they have no competing interests.

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