Rare disease

Pneumatosis intestinalis presenting with a pneumoperitoneum in a patient with chronic bronchiectasis: a delayed diagnosis of superior mesenteric artery ischaemia

Nicholas Dawe, Sidrah Akhtar

Department of General Surgery, Princess Royal University Hospital, Orpington, UK

Correspondence to Nicholas Dawe, nicdawe@gmail.com

Summary

We describe a case of pneumatosis intestinalis (PI) in a 69-year-old woman presenting to the surgical department with non-specific abdominal pain and a pneumoperitoneum.

PI complicated by pneumoperitoneum was diagnosed on the basis of the characteristic clinical and abdominal CT findings, demonstrating a cystic gas pattern within the small bowel wall. In the context of resolving clinical findings, non-peritonitic abdomen and with no evidence of portal venous gas or perforation the patient was managed conservatively. The findings were considered benign and attributed to her chronic acquired bronchiectasis.

Following recurrence of symptoms after several months, further imaging confirmed chronic mesenteric ischaemia (CMI) involving the superior mesenteric artery. Angioplasty was performed and symptoms resolved.

PI is one of the few imaging signs to occur in CMI. PI presenting as a pneumoperitoneum is rare and may often lead to avoidable surgical intervention. However, no clear algorithm exists to guide surgical management.

BACKGROUND

Pneumatosis intestinalis (PI) is an unusual radiological finding. It is reported in the literature in association with a wide range of clinical conditions and explained by several diverse theories. PI presenting as a pneumoperitoneum is rare, occurring overall in <3% of cases,¹ and may lead to avoidable surgical intervention.² No clear algorithm exists for surgical management.

Since the first clinical review in 1952 by Koss,³ which considered 255 case reports within 213 English literature publications, the finding has been attributed to numerous underlying pathologies. Pear⁴ performed a significant review in 1998 and recognised 'at least 58 causative factors since 1952' linked to PI.

The four established theories for the pathogenesis of intramural gas are bowel necrosis, mucosal disruption, increased mucosal permeability and pulmonary disease. While the majority of cases may be managed conservatively, intramural gas resulting from bowel necrosis—resulting from infarction, ischaemia or volvulus—represents a surgical emergency and requires urgent exclusion.

We describe a case of PI presenting as a pneumoperitoneum, which was initially considered a benign finding secondary to underlying chronic bronchiectasis. Despite resolution of symptoms following initial conservative management, the patient represented several months later with ongoing abdominal pain with further imaging confirming chronic mesenteric ischaemia (CMI).

Patients presenting with either non-specific symptoms or as an acute surgical abdomen, with previously undiagnosed PI complicated by a pneumoperitoneum, are uncommon

and continue to represent both a diagnostic and management dilemma for the surgeon. Case series suggest that in those whose symptoms are eventually attributed to the benign form, up to 27% may undergo avoidable surgery.¹

Surgeons must remain alert to the numerous medical causes of intra-abdominal free gas in the adult and the appropriateness of a conservative approach in benign causes. Our experience highlights the importance of identifying those patients with PI, with or without complications, who fail to respond to non-operative management and necessitate re-evaluation and exclusion of an underlying pathology that may require urgent intervention.

Our case emphasises the difficulties in excluding significant pathologies that may lead to bowel necrosis.

CASE PRESENTATION

A 69-year-old retired Caucasian woman presented via the emergency department to the surgical team with a 3-day history of intermittent, mild and diffuse abdominal discomfort, occurring with increased frequency, with associated anorexia, nausea and bilious vomiting and mild weight loss. She had opened her bowels 1 day previously and continued to pass flatus.

Over the preceding 3 months, similar but less severe and self-resolving episodes had occurred. She was awaiting an outpatient small bowel enteroscopy for suspected distal ileal abnormality identified on an otherwise unremarkable abdominal CT scan.

The patient had a long-term history of atrial fibrillation (managed with warfarin and digoxin) and a 5-year history

Figure 1 Plain anterior–posterior (AP) chest radiograph performed on admission; the film demonstrates gas pattern consistent with free air beneath the right diaphragm.

of reflux oesophagitis not requiring any medical management. Surgical history included a laparoscopic cholecystectomy 3 years previously and right lower lobectomy 10 years previously for severe chronic bronchiectasis of unknown aetiology. The patient had not received oral or inhaled steroid treatment within the preceding 6 months. She was a lifelong non-smoker and drank an average of 5 alcohol units per week.

Examination revealed a soft, mildly distended abdomen with tenderness in the periumbilical area and no associated peritonism. Rectal examination was normal with formed stool and no blood. Respiratory examination revealed fine bibasal crepitations.

INVESTIGATIONS

Routine admission bloods were unremarkable except for a white cell count 12.7×10^9 . The full blood count, serum biochemistry, C reactive protein, arterial blood gas analysis pH and base excess were within normal limits.

Chest plain film radiograph (figure 1) was consistent with left basal atelectasis and demonstrated a gas shadow beneath the right diaphragm. Abdominal plain film (figure 2) supported a finding of herniation of the large bowel between the liver and diaphragm, moderate colonic faecal loading, cholecystectomy clips and no evidence of perforation or obstruction.

TREATMENT

The patient was managed for partial bowel obstruction, placed nil-by-mouth and admitted for intravenous fluid hydration and observation.

During admission, the patient reported some periods of relief of her abdominal discomfort. She began tolerating free fluids on day 2 and continued to pass normal stool. Repeat abdominal plain film radiograph performed on day 4 demonstrated distended bowel loops with maximal diameter of 3.5 cm with free intraperitoneal gas and pneumatosis (figure 3). A retrospective comparison was made with the admission abdominal film confirming pneumatosis intestinalis within the small bowel.

An urgent abdominal CT scan (figure 4) was performed confirming extensive air, in a cystic pattern, within the wall of the small bowel and peritoneal cavity, with fluid filled distended small and large bowel loops and no free intraperitoneal fluid. No focal perforation or strictures were identified. Notably, the CT showed crossing over of the superior mesenteric vein and artery suggestive of small bowel malrotation. A chest CT confirmed bronchiectatic lung disease with mucous-filled bronchi and emphysematous change at the lung bases.

The patient was placed nil-by-mouth, central access obtained with total parenteral nutrition (TPN) and intravenous metronidazole initiated. In view of the patient's

Figure 2 Plain abdominal radiograph on admission; the image demonstrates small bowel within the right upper quadrant and intramural gas within the herniated bowel segment.

resolving symptoms and non-peritonitic abdomen, a conservative approach was adopted. An exploratory laparoscopy was considered an option at this stage should this initial management approach fail. The patient successfully restarted on a light diet after 5 days.

Water-soluble contrast swallow and follow-through studies (figure 5) were performed 7 days after diagnosis failed to demonstrate any strictures but identified luminal narrowing at the terminal ileum.

However, colonoscopy revealed no terminal ileal disease with diverticulae in the descending colon and caecal atrophy.

PI complicated by pneumoperitoneum was diagnosed on the basis of the characteristic clinical and radiological findings. As the symptoms fully resolved following conservative management, the findings were attributed to a benign cause secondary to her chronic bronchiectasis.

OUTCOME AND FOLLOW-UP

Several months following her discharge, the patient represented with ongoing symptoms of central non-specific abdominal pain. A privately organised MRI scan was performed and characterised the vascular anatomy identifying narrowing within the superior mesenteric artery (SMA). Angiography of the mesenteric vessels was performed with balloon dilatation of the SMA, which led to the resolution of her symptoms.

DISCUSSION

PI is not a diagnosis in itself, but a radiological finding secondary to one of numerous causes and has a range of clinical outcomes.

In the context of non-specific abdominal symptoms, the initial radiological findings and associated pneumoperitoneum may mimic an acute abdomen representing a

Figure 3 Repeat (day 4) abdominal plain radiograph; image demonstrates diffuse small bowel distension and pneumatosis intestinalis.

diagnostic challenge for the surgeon. An accepted classification has been to divide PI into a primary type (15%) and those secondary to a pathological process (85%).³ This classification has been challenged throughout the literature as the intramural gas is always secondary to an underlying pathology in some form. Over the past 60 years, as the accepted theories of pathogenesis have emerged, the primary or benign form is considered, in the majority of cases, to exist secondary to pulmonary disease.

The pulmonary theory received initial scepticism following its development during the early 1960s. As numerous case series were published linking cases of PI with a common history of allergy and asthma, Keyting *et al*⁶ presented a case series of PI associated with pulmonary disease and was the first to apply the now established theory that alveolar rupture could result in a pneumomediastinum;

they suggested the potential existed for gas to then dissect via several routes, including a peri-vascular route, to the submucosal and subserosal layers of the mesocolon. Well-recognised pulmonary associations include asthma, cystic fibrosis, chronic obstructive pulmonary disease, barotrauma, thoracic injury and following lung transplantation.

Significantly, Doumit *et al*⁶ reported an isolated case of uncomplicated PI in a 66-year-old woman suffering from chronic bronchiectasis, which resolved after conservative management. This association had not previously been reported within the English literature. In our case, the patient had a history of bronchiectasis of unknown aetiology, characterised by a chronic productive cough and was under long-term respiratory review. She had reported no recent change in symptom severity, which may have led to the accumulation of intramural gas. Ten years previously

Figure 4 CT scan performed on day 4; image confirms radiological diagnosis of small bowel pneumatosis and an associated significant pneumoperitoneum.

she had undergone a right lower lobectomy, indicating the past severity of her symptoms and failure of medical management.

Two features of our case deserve further discussion: the recurrence of symptoms leading to a diagnosis of CMI and the evaluation of a 'medical' pneumoperitoneum.

Following the recurrence of symptoms, further imaging was undertaken with the findings demonstrating the importance of re-evaluating the diagnosis after failure of conservative management. Evidence of PI is often identified following presentation with subacute abdominal symptoms. In this case, chronic SMA ischaemia had led to PI with pneumoperitoneum. Mesenteric ischaemia is an uncommon cause of abdominal pain, accounting for fewer than 5% of intestinal ischaemic events, and producing the well-defined symptoms of postprandial abdominal pain and food aversion leading to weight loss, which become evident when two out of the three splanchnic vessels are involved⁷; PI, if present, is one of the few imaging signs to occur in CMI.

PI is one of many non-surgical causes of intraperitoneal free gas. With 90% of non-iatrogenic pneumoperitoneum

indicating a visceral perforation,⁸ the remaining proportion may be regarded as 'spontaneous' or 'medical' resulting from postoperative, thoracic, abdominal or gynaecological causes.⁹ Intra-abdominal free gas is an uncommon complication evident in up to 10% of patients with small bowel pneumatosis and 2% with large bowel pneumatosis.¹ Symptoms occurring in association with PI and pneumoperitoneum require careful analysis with exclusion of either an ischaemic event or perforation.

An awareness of the pathogenesis of benign PI and other 'non-surgical' causes of pneumoperitoneum empowers the surgeon to pursue a conservative approach with exploratory surgery indicated only in highly selected cases not responding to non-operative management.

Of significance in our case, the patient had not recently received steroid treatment for her bronchiectasis. In the context of immunosuppressive treatment, the surgeon must be alert to the risks of conservatively managing those presenting with pneumoperitoneum and a paucity of clinical findings since the signs of peritonism may be masked. In these situations, and when hollow organ perforation cannot be excluded, many would advocate early exploratory

Figure 5 Water-soluble contrast follow-through of the small bowel following 7 days of conservative management demonstrating otherwise normal gross anatomy.

laparotomy to exclude the presence of bowel necrosis or perforation.¹⁰ In our case, the patient responded well to the conservative approach involving bowel rest, TPN and intravenous metronidazole.

An examination of factors independently associated with a decision to adopt a surgical approach and a greater mortality, in cases of PI, was undertaken by Greenstein *et al*¹¹ In their review of 40 patients with a retrospective diagnosis of PI, they showed those 60 years old or over, and either the presence of vomiting or white cell count greater than 12×10^9 ,

were most likely to undergo surgery, while sepsis was the sole independent risk for death. A single-centre study of 27 patients over a 1-year period, of which 16 underwent surgery, considered surgical management and predictors of outcome.² Those showing the greatest benefit from surgery demonstrated abdominal symptoms associated with acidosis and hyperamylasaemia; however, eight of the patients with ischaemic bowel and PI had 75% mortality despite their surgery. No clear algorithm for surgical management has yet been accepted.

Learning points

- ▶ PI is an unusual radiological finding, although not a diagnosis in itself. It is reported in association with numerous underlying pathologies from the benign to life-threatening.
- ▶ Surgeons must remain alert to the numerous medical causes of intra-abdominal free gas in the adult and the appropriateness of a conservative approach in benign causes. Equally, it is imperative to identify those patients with PI, with or without complications, with underlying pathologies requiring urgent intervention.
- ▶ No algorithm exists to dichotomise patients into conservative and surgical management. However, before attributing the PI to a benign process, urgency must be established based upon symptom characteristics, radiological features, including portal venous gas and pneumoperitoneum, lung pathology, vascular supply to the involved bowel segment and surgical history.
- ▶ Mesenteric ischaemia is an uncommon cause of abdominal pain accounting for fewer than 5% of intestinal ischaemic events.
- ▶ PI, if present, is one of the few imaging signs to occur in CMI.

Competing interests None.

Patient consent Obtained.

REFERENCES

1. **Jamart J.** Pneumatosis cystoides intestinalis. A statistical study of 919 cases. *Acta Hepatogastroenterol (Stuttg)* 1979;**26**:419–22.
2. **Knechtle SJ,** Davidoff AM, Rice RP. Pneumatosis intestinalis. Surgical management and clinical outcome. *Ann Surg* 1990;**212**:160–5.
3. **Koss LG.** Abdominal gas cysts (pneumatosis cystoides intestinorum hominis); an analysis with a report of a case and a critical review of the literature. *AMA Arch Pathol* 1952;**53**:523–49.
4. **Pear BL.** Pneumatosis intestinalis: a review. *Radiology* 1998;**207**:13–19.
5. **Keyting WS,** McCarver RR, Kovarik JL, *et al.* Pneumatosis intestinalis: a new concept. *Radiology* 1961;**76**:733–41.
6. **Doumit M,** Saloojee N, Seppala R. Pneumatosis intestinalis in a patient with chronic bronchiectasis. *Can J Gastroenterol* 2008;**22**:847–50.
7. **Moawad J,** Gewertz BL. Chronic mesenteric ischemia. Clinical presentation and diagnosis. *Surg Clin North Am* 1997;**77**:357–69.
8. **Winck TG,** Mosely HS, Grout G, Luallin D. Pneumoperitoneum and its association with ruptured abdominal viscus. *Arch Surg* 1988;**123**:709–12.
9. **Mularski RA,** Ciccolo ML, Rappaport WVD. Nonsurgical causes of pneumoperitoneum. *West J Med* 1999;**170**:41–6.
10. **Ruh J,** Testa G, Von Deimling F, *et al.* Management of pneumatosis coli with free intraperitoneal gas mimicking abdominal hollow organ perforation in a 13-year-old patient following bone marrow transplantation. *Dig Dis Sci* 2003;**48**:1542–4.
11. **Greenstein AJ,** Nguyen SQ, Berlin A, *et al.* Pneumatosis intestinalis in adults: management, surgical indications, and risk factors for mortality. *J Gastrointest Surg* 2007;**11**:1268–74.

This pdf has been created automatically from the final edited text and images.

Copyright 2010 BMJ Publishing Group. All rights reserved. For permission to reuse any of this content visit <http://group.bmj.com/group/rights-licensing/permissions>.

BMJ Case Report Fellows may re-use this article for personal use and teaching without any further permission.

Please cite this article as follows (you will need to access the article online to obtain the date of publication).

Dawe N, Akhtar S. Pneumatosis intestinalis presenting with a pneumoperitoneum in a patient with chronic bronchiectasis: a delayed diagnosis of superior mesenteric artery ischaemia. *BMJ Case Reports* 2010;10.1136/bcr.01.2010.2622, date of publication

Become a Fellow of BMJ Case Reports today and you can:

- ▶ Submit as many cases as you like
- ▶ Enjoy fast sympathetic peer review and rapid publication of accepted articles
- ▶ Access all the published articles
- ▶ Re-use any of the published material for personal use and teaching without further permission

For information on Institutional Fellowships contact consortiasales@bmjgroup.com

Visit casereports.bmj.com for more articles like this and to become a Fellow