CASE REPORT

Adult necrotising enterocolitis—pig-bel disease: a Pacific disease in London

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SUMMARY

Adult necrotising enterocolitis secondary *Clostridium perfringens* type C—pig-bel disease—is rarely seen outside of Pacific populations, with the highest incidence being in Papua New Guinea. We present the first reported case of pig-bel disease in a previously well patient without diabetes following food poisoning since 1996. In this case, the enterotoxin-induced disease necessitated emergency laparotomy and bowel resection following the failure of medical treatment and worsening septic shock. We hypothesise that obstruction secondary to a sigmoid diverticular stricture complicated by infection-related oedema led to colonic stasis and rapid progression of disease and bowel loss. Following bowel resection, several parenteral antibiotic agents and prolonged intensive care, this patient survived this severe infective process and was discharged home.

BACKGROUND

Symptoms of vomiting, diarrhoea and abdominal pain are common presenting symptoms in emergency departments worldwide. In many cases, simple gastroenteritis is the cause, whether bacterial or viral. In other cases, significant medical or surgical pathology may be the cause of these symptoms necessitating hospital admission. Rarely, patients may present with these symptoms and progress to systemic sepsis and shock as a result of the underlying pathology. We present an exceptionally rare case of this type where a patient presented with severe intra-abdominal sepsis, refractory to medical therapy and requiring extensive abdominal surgery and prolonged intensive care due to bacterial necrotising enterocolitis associated with *Clostridium perfringens* type C—also known as pig-bel disease.

CASE PRESENTATION

A 54-year-old man presented to the emergency department with a 48-hour history of central abdominal pain, profuse diarrhoea and vomiting followed by cessation of diarrhoea but continued severe vomiting for a further 18 hours. His symptoms began shortly after ingesting a chicken curry from a takeaway 3 hours previously. Other family members had also eaten the same dish but at a smaller quantity. They described similar symptoms but less severe.

On examination, he was in septic shock with a tachycardia of 121 bpm, a blood pressure of 102/64 mm Hg and was hypothermic at 35.6. He was peripherally vasoconstricted with prolonged capillary refill time of 4 s. His abdomen was distended,

tense and tympanic, with diffuse tenderness but no peritonism.

He had no medical, family or social history of significance.

INVESTIGATIONS

An arterial blood gas showed a pH of 7.12, base excess −15, HCO_3^- 14.3, pCO_2 5.9 and lactate of 6.8.

Biochemistry showed a degree of kidney injury, with creatinine of 100 an estimated glomerular filtration rate of 68 from a previously normal baseline.

His C reactive protein was raised at 189 with a normal white cell count. Liver function tests showed an albumin level of 23.

He proceeded to have an erect chest radiograph, which showed no evidence of pneumoperitoneum and an abdominal radiograph that demonstrated globally distended loops of large bowel.

A CT abdomen and pelvis showed large bowel dilation to the distal descending colon/proximal sigmoid colon, which appeared thickened. The small bowel was also dilated with no transition point. There was a small amount of free fluid but no free intra-abdominal gas. The exact aetiology was not clear from CT, but infection/inflammation was the most likely cause of the findings noted—neither malignancy nor ischaemia could be ruled out.

DIFFERENTIAL DIAGNOSIS

1. Infectious colitis with perforation leading to intra-abdominal sepsis
2. Ischaemic colitis with perforation leading to intra-abdominal sepsis
3. Perforated peptic ulcer
4. Appendicitis with perforation and abdominal sepsis

TREATMENT

He was treated for combined septic and hypovolaemic shock with aggressive intravenous fluid resuscitation and intravenous antibiotics—cefuroxime and metronidazole. His condition was refractory to fluid resuscitation and he remained persistently hypotensive despite 5 L of crystalloid infusion. He was transferred to the intensive care for further resuscitation, invasive monitoring (central venous line and arterial line) and inotropic support with metaraminol and norepinephrine. He was treated with low-dose intravenous hydrocortisone and sodium bicarbonate. His acidosis improved but despite maximal medical therapy his lactate continued to rise. His abdomen became increasingly distended and tender such that his respiratory

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Figure 1 Intraoperative flexible sigmoidoscopy demonstrating classic necrotic mucosa associated with pig-bel disease.

function became compromised secondary to diaphragmatic splinting. At this point, he was started on meropenem and linezolid in place of cefuroxime and metronidazole.

Fourteen hours into his admission he was taken to theatre based on a failure of medical treatment. A flexible sigmoidoscopy was initially performed and showing an impassable ischaemic stricture at 40 cm obstructing the sigmoid colon (figure 1). The patient rapidly and progressively deteriorated during the flexible sigmoidoscopy. A subtotal colectomy and long-segment ileal resection was performed rapidly with a stapling device to save time. An end ileostomy was formed. Intraoperatively, macroscopic findings were as follows: patchy necrosis of the colon up to the obstructing diverticular stricture in the sigmoid and patchy necrosis in the small bowel up to 170 cm from the duodenojejunal flexure. There was also disease in the remaining small bowel as evidenced by sloughing of the mucosa at the level of the end ileostomy postoperatively, even though externally the small bowel left behind looked healthy at a serosal level.

OUTCOME AND FOLLOW-UP

He received inotropic and respiratory support for 5 days postoperatively, after which he was extubated. The hospital was contacted by Public Health England, as toxin-producing *C. perfringens* type C was isolated from the bowel resection specimen. The histopathology report of the bowel resection specimen was consistent with a diagnosis of *C. perfringens* necrotising enterocolitis. Two days postoperatively his white cell count remained high (42 units) causing concern about ongoing infection and further bowel necrosis. At this point, metronidazole was started in addition to meropenem. After this he recovered well and was discharged from hospital after a 16-day stay. He was reviewed in clinic to manage his initial high output stoma with antidiarrhoeal agents and intermittent infusion of magnesium. He will undergo an elective reversal of his ileostomy with restoration of intestinal continuity in due course. With 170 cm of small bowel and 40 cm of colon remaining he is expected to have reasonable bowel function. Our patient's severe symptoms and clinical deterioration resulted in an extensive resection of bowel with a prolonged hospital stay due to a severe *C. perfringens* infection with toxin C, known as pig-bel disease.

DISCUSSION

Pig-bel disease is a condition characterised by necrotising enterocolitis associated with *C. perfringens* type C.¹ Although rare worldwide, it classically affects Pacific populations. Indeed, pig-bel comes from a pidgin English term for abdominal pain after a pig feast.² Papua New Guinea in particular has the highest incidence of pig-bel disease and may be secondary to the ritual eating of contaminated pork.³ It has a predilection to affect those with severe protein malnutrition and diabetics and has a high mortality, especially in children.⁴ It is incredibly rare in the West, with only a few reported cases in the UK, Japan and the USA, the majority of these in patients with diabetes mellitus.⁴⁻⁵ Interestingly, our report is the second to describe one of the rarest presentations of pig-bel disease—in a previously fit and healthy patient without diabetes following food poisoning in the UK, with only one other reported case in a healthy vegetarian woman.⁶

C. perfringens causes a variety of diseases in humans and is associated with several gastrointestinal-specific infections. The pathogenesis of *C. perfringens* gastroenteritis is via proliferation of the bacterium within the gastrointestinal tract and the subsequent production of toxins that have profound catastrophic local and systemic effects.⁷ Of all of the toxins produced by *C. perfringens*, *C. perfringens* β toxin is the most important for intestinal disease, while α and θ are key mediators of systemic septic shock.⁸ Locally, *C. perfringens* type C infection causes haemorrhagic necrosis of the intestinal mucosal epithelium that then progresses to all layers of the intestinal wall. Progressive inflammation and infection leads occlusion of the superficial arteries and veins of the lamina propria and submucosa occurs leading to further necrosis.⁹

We hypothesise that the toxin produced by this bacteria was able to cause extensive patchy necrosis necessitating extensive bowel resection, in a pathogenesis not dissimilar to necrotising fasciitis, due to a mechanically obstructing sigmoid lesion that prolonged the time spent in direct contact between the toxin and the mucosa of the bowel. In other reported cases of pig-bel disease resulting in the same pathological process, patients had used excessive quantities of loperamide and anti-diarrhoeal agents resulting in effectively the same stasis of infected stools.

In this patient, we changed antibiotics early in our management despite not having culture results back, when we suspected poor response to surgical resection. This was based on high inotropic requirements and ongoing requirement for multiorgan support.

Learning points

- ▶ Abdominal pain, vomiting and diarrhoea—although often benign, can be associated with significant septic shock requiring urgent medical and surgical intervention.
- ▶ Pig-bel disease is a rare cause of abdominal sepsis and is most commonly found in Pacific populations and patients with diabetes mellitus.
- ▶ Severe infective colitis that is refractory to medical therapy may require surgical review for consideration for bowel resection in the presence of worsening sepsis.
- ▶ Infective colitis in the presence of mechanical bowel obstruction can lead to rapid deterioration of bowel integrity and perforation.

Contributors All authors have contributed equally to the preparation and submission of this case report. JCRW and FM drafted the manuscript. JCRW and SD reviewed and edited the drafts. NB provided expert input and reviewed final draft prior to submission. All authors were directly involved in the care of the patient.

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