Open Journal of
Clinical & Medical
Case Reports

Volume 2 (2016)
Issue 3

ISSN 2379-1039

Pigbel: The Under-Recognized Cause of Bowel Ischemia

Arvand Elihu, MD; Catherine Chung, BS; Eugene P. Ceppa, MD*

***Eugene P. Ceppa**

Department of Surgery, Indiana University School of Medicine, 545 Barnhill Dr., EH 541, Indianapolis, IN 46202 USA.

Tel: 317-948-8358, Fax: 317-278-4897; Email: eceppa@iupui.edu

Abstract

Pigbel is a pathologic entity not found commonly throughout the world. The term "pigbel" originated from the native dialect, Tok Pisin, of Papua New Guinea. It describes the violent abdominal pain and distension suffered by patients after consuming large quantities of pork during tribal festivals. *Clostridium perfringens* was described as the infectious agent responsible for causing pigbel, or enteritis necroticans, in Papua New Guinea. The immediate recognition of a patient with necrotic bowel is paramount for patient survival. Diagnosis is confirmed by histopathological findings in the setting of no clear vascular or mechanical etiology. A few cases have been reported in developed countries, in which involved patients had predisposing comorbidities. We report a novel case of pigbel in North America in a previously healthy patient.

Keywords

Pigbel; Enteritis necroticans; Mesenteric ischemia; *Clostridium perfringens*; Acute mesenteric ischemia

Abbreviations

EN: Enteritis necroticans; CT: Computed tomography; CP: *Clostridium perfringens*; AMI: Acute mesenteric ischemia

Introduction

Enteritis necroticans (EN), used synonymously with pigbel, is a fatal illness resulting in spontaneous necrosis of the small bowel. A similar illness was first described in Germany as *Darmbrand* following a post-World War II epidemic [1]. Pigbel became a recognizable public health problem following an outbreak, involving 17 patients, in Papua New Guinea during the 1960's [2]. This allowed for detailed documentation of the clinical presentation, depiction of the histopathological findings, and characterization of the mechanism by which *Clostridium perfringens* (CP) causes EN.

CP consists of five types (A-E), which are responsible for producing 12 total antigenic protein toxins. CP type A releases the α-toxin resulting in "gas" gangrene in postoperative and posttraumatic wounds [3]. The type A enterotoxin ranks as the third most common source of foodborne illness in the United States [4]. EN is another well documented disease related to CP type C via the β-toxin [5]. Not limited to Papua New Guinea, cases of EN caused by CP have been documented in industrialized nations in patients with relative immunosuppression or with diabetes [6-8]. This case report describes a previously healthy patient who is afflicted by EN following consumption of pork and sweet potatoes.

Case Presentation

A 52-year-old Caucasian female was evaluated in the emergency department for complaints of acute onset epigastric abdominal pain, which was preceded by nausea, emesis, and episodes of explosive bloody diarrhea for six hours. Upon further investigation, the patient admitted to consuming a meal of large portions of pork and sweet potatoes the evening prior to symptoms at a local restaurant. Her past medical history was unremarkable for coronary artery disease, arrhythmia, or peripheral vascular disease. Of note, she denied any recent weight loss or fear of food, and admitted to completing a seven-day course of Bactrim for a urinary tract infection and consuming soy to relieve her hot flashes. Past surgical history included a combined cholecystectomy and appendectomy via a paramedian incision 30 years ago. Physical examination was pertinent for profound hypotension, soft abdomen, and pain out of proportion to examination. The patient was treated with volume resuscitation, and her blood pressure responded appropriately. Computed tomography (CT) revealed diffuse thickened loops of small bowel with a large amount of free fluid in the pelvis without signs of calcifications of the visceral segment of the aorta (Figure 1). The patient became hypotensive again with a rising lactate in spite of aggressive intravenous resuscitation.

The patient was emergently taken to the operative theater for an exploratory laparotomy. Upon entering the peritoneum, the presence of necrotic bowel was evident (Figure 2). The jejunum and proximal ileum was hemorrhagic, necrotic appearing, and non-edematous. A clear line of demarcation was visible at both the proximal and distal margins. No signs of significant adhesions, abdominal wall hernias, internal hernias, or other mechanical causes were evident to the surrounding small bowel. Strong pulses were palpated at the root of the mesentery as well as at the arcuate arteries feeding the hemorrhagic segments of bowel. No other intra-abdominal pathology was noted upon inspection. A segmental resection of 165cm of necrotic small bowel was completed with a linear stapler and reconstructed with a stapled small bowel anastomosis.

The postoperative period was notable for extensive hypercoagulability evaluation by hematology. Laboratory studies included serum evaluation for protein C/S deficiency, anticardiolipin antibody, lupus antibody, and hyperhomocyteinemia; these studies were all unremarkable for any abnormality. The microscopic findings of the surgical pathology revealed full thickness necrosis with viable margins of normal appearing bowel and no other pathologic findings. Gram stain revealed gram positive rods distributed along the brush border of the small bowel with a morphology consistent with *Clostridium Perfringens*. She recovered well following surgery and was discharged home on a regular diet on postoperative day seven.

Discussion

Enteritis necroticans is a devastating infectious disease resulting in necrosis of small bowel with few warning signs. Symptoms associated with EN mimics acute mesenteric ischemia (AMI) and should be considered a form of non-occlusive AMI. Interestingly, 15-20% of all cases of AMI are considered non-occlusive typically a result of a low flow state, vasoactive drugs, or unknown etiology [9]. In this case, this patient's initial presentation was consistent with AMI. However, the intraoperative findings and postoperative clinical evaluation failed to demonstrate a viable cause of mesenteric ischemia. Upon further investigation of the patient's history, gross specimen, and gram stain, it was evident that

Clostriudium perfringens was responsible for this patient's small bowel necrosis, and may be one cause of the "unknown" cases of AMI.

The etiology of AMI is extensive but is simplified by dividing causes into two categories: occlusive and nonocclusive. Occlusive ischemia results from either embolism or thrombosis of a major branch of the splanchnic vasculature. Embolic sources include myocardial infarction and atrial fibrillation, and thrombotic sources include atherosclerosis and thrombophilic processes. Nonocclusive ischemia results from preferential vasoconstriction of the splanchnic vascular bed during a low flow state secondary to shock in critical patients. However, most algorithms fail to include infection as a cause of AMI.

CP is a naturally occurring bacterium lining the gastrointestinal and genital tracts. EN is a well-documented disease process caused by CP type C β-toxin. Animal studies have shown that protein-starved animals inoculated with *Clostridium welchii* type C and natural trypsin inhibitors from sweet potatoes into the duodenum died with histopathological changes consistent with EN. Meanwhile, animals treated with autoclaved sweet potato were unaffected and were found to have β-antitoxin without previous vaccination [10]. Thus, proteolysis by the gut detoxifies the β-toxin of CP. However, if infected pork is consumed with trypsin inhibitors or if a patient is protein malnourished, providing suppressed secretion of trypsin, then proteolysis of the β-toxin of CP will not occur. In a study by Rackis et al, studies found consuming a large pork meal in addition to sweet potatoes, known trypsin inhibitors, led to the development of typical symptoms of EN [11]. In this case, the patient previously consumed a large meal of sweet potatoes and pork. Additionally, she was chronically ingesting raw soy, which contains the highest concentration of active soybean trypsin inhibitors. Both of these factors contributed to the patient's development of EN caused by the failure of CP type C β-toxin proteolysis.

The clinical features after inoculation include a latent period of hours to days before the onset of symptoms, which include severe upper abdominal pain, vomiting, and bloody diarrhea. These symptoms can be followed by constipation, intestinal obstruction, and shock. The gross pathologic features are described as segmental patches of necrosis along the antimesenteric border of the involved small bowel [12]. Areas involved are typically the jejunum with some involvement of the ileum. There are not any reports of involvement of the colon in cases of EN. The histopathologic findings include a heavy polymorphonuclear cellular response, thrombi and endothelial proliferation visible within the mucosal venules, and infarction of mucosal epithelium [13]. The remaining layers of the small bowel wall remain intact. Small bowel villus tip necrosis and the presence of adjacent organisms are visible by gram stain or electron microscopy [14].

Conclusion

In conclusion, patients who have an atypical pattern of ischemic small bowel without history, physical exam, or diagnostic studies demonstrating an embolic, thrombotic, or nonocclusive etiology of AMI should be further assessed for an infectious etiology. Intraoperatively, gross specimens should be sent for anaerobic culture and gram stain in addition to other conventional microbiologic and histopathologic studies.

Figures

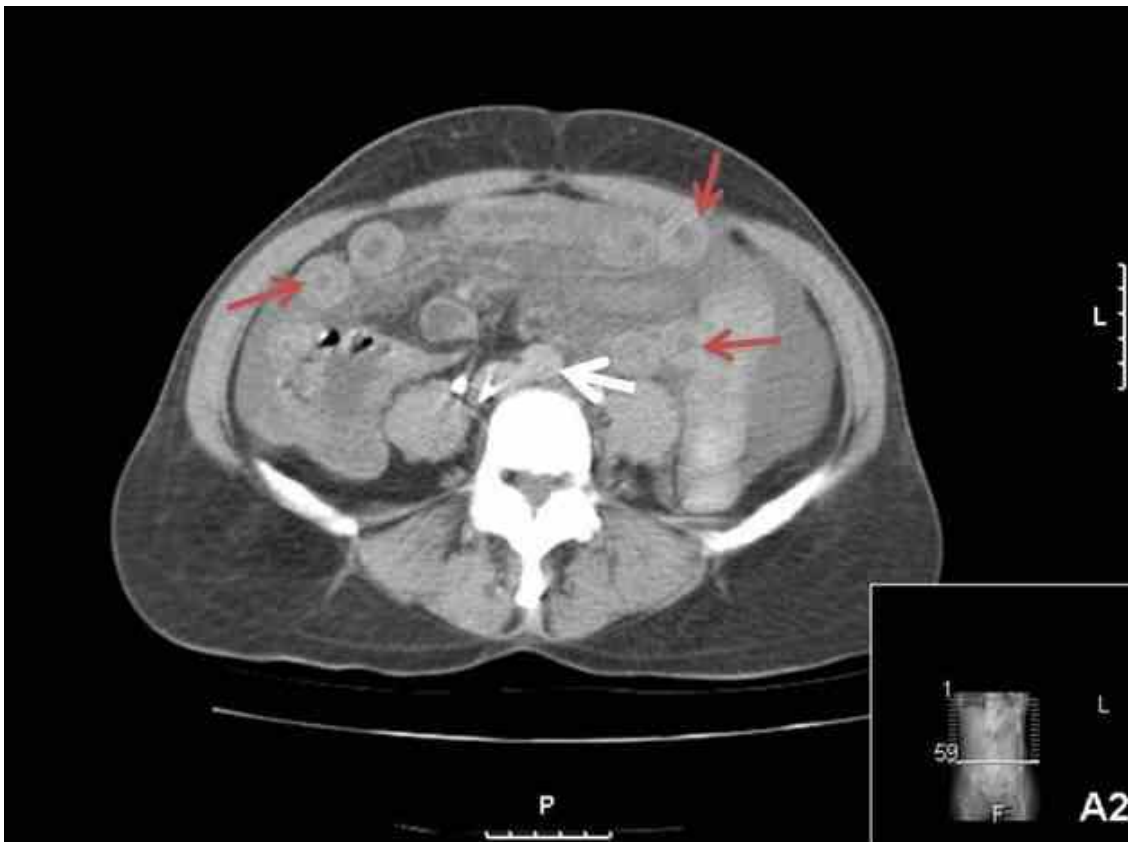

Figure 1: Axial view of CT scan of the abdomen without contrast. The red arrows indicate the thickened loops of small bowel. No signs of calcifications of the aorta are noted (white arrow).

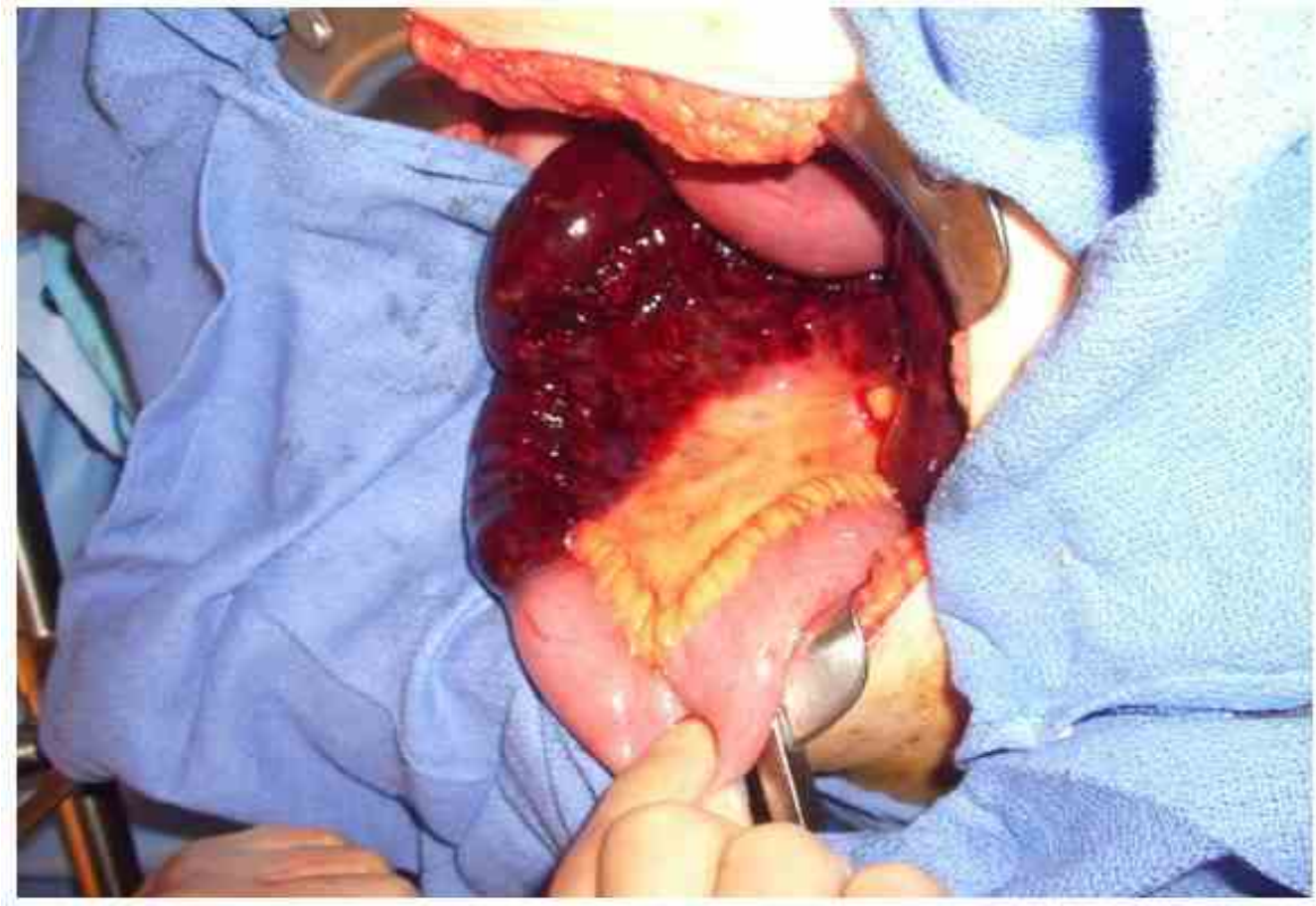

Figure 2: Intraoperative image of small bowel. Clear line of demarcation is noted between necrotic proximal ileum bowel and normal appearing distal ileum. Affected bowel was hemorrhagic, necrotic appearing, and non-edematous as shown.

1. Hansen K, Jecklen E, Jochims J, Lezius A, Meyer-Burgdorff H, Schutz F. Darmbrand-Enteritis Necroticans. Stuttgart: George Thieme (1949).

2. Murrell TGC, Roth L. Necrotizing jejunitis: a newly discovered disease in the highlands of New Guinea. Med J Aust. 1963; 1: 61-69.

3. MacLennan JD. The histotoxic clostridial infection of man. Bacteriol Rev. 1962; 26:177-276.

4. McClane, BA. Clostridium perfringens, p 351-372. In MP Doyle, LRBeuchat, and TJ Montville (ed), Food microbiology: fundamentals and frontiers, 2nd ed. ASM Press, Washington DC.

5. Egerton RJ, Walker PD. The isolation of Clostridium perfringens type C from necrotic enteritis of man in Papua New Guinea. J Pathol Bacteriol. 1964; 88: 275-78.

6. Petrillo TM, Beck-Sague CM, Songer JG, Abramowsky C, Fortenberry JD, Meacham L, et al. Enteritis necroticans (pigbel) in a diabetic child. *N Engl J Med*. 2000 Apr 27;342(17):1250-3.

7. Gui L, Subramony C, Fratkin J, Hughson MD. Fatal enteritis necroticans (pigbel) in a diabetic adult. Mod Pathol. 2002 Jan; 15(1): 66-70.

8. Sobel J, Mixter CG, Kolhe P, Gupta A, Guarner J, Zaki S, et al. Necrotizing enterocolitis associated with clostridium perfringens type A in previously healthy north american adults. J Am Coll Surg. 2005 Jul; 201(1): 48-56.

9. Yashura H. Acute mesenteric ischemia: the challenge of gastroenterology. Surg Today. 2005; 35(3): 185-95.

10. Lawrence G, Walker PD. Pathogenesis of enteritis necroticans in Papua New Guinea. Lancet. 1976; 1: 125-6.

11. Rackis JJ, McGee JE. Effects of Soy Proteins Containing Trypsin Inhibitors in Long Term Feeding Studies in Rats. J. Am Oil Chemists Soc. 1979; 56: 162-68.

12. Murrell TG, Walker PD. The pigbel story of Papua New Guinea. Trans R Soc Trop Med Hyg. 1991 Jan-Feb; 85(1): 119-22.

13. Cooke R. The pathology of *pigbel*. P N G Jed J. 1979; 1: 35-8.

14. Walker PD, Murrell TGC, Nagy LK. Scanning electronmicroscopy of the jejunum in enteritis necroticans. J Med Microbiol. 1980; 13: 445-50.

Manuscript Information: Received: December 12, 2015; Accepted: February 12, 2016; Published: February 16, 2016

Authors Information: Arvand Elihu, MD[1]; Catherine Chung, BS[2]; Eugene P. Ceppa, Md[2*]
[1]Department of Surgery, Kaiser Permanente Woodland Hills Medical Center, USA
[2]Department of Surgery, Indiana University School of Medicine, USA

Citation: Elihu A, Chung C, Ceppa EP. Pigbel: the under-recognized cause of bowel ischemia. Open J Clin Med Case Rep. 2016; 1078

Journal: Open Journal of Clinical and Medical Case Reports is an international, open access, peer reviewed Journal focusing exclusively on case reports covering all areas of clinical & medical sciences.

Visit the journal website at **www.jclinmedcasereports.com**

For reprints & other information, contact editorial office at **info@jclinmedcasereports.com**