

Enteritis Necroticans in Nepal

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The presence, in Nepal, of a disease histologically identical to Pig-bel (Enteritis necroticans) has recently been established beyond reasonable doubt.¹ Two more cases of Pig-bel occurring in Gorkha District of Nepal are presented followed by a review of the literature. This review includes a discussion of the pathogenesis, clinical features, medical and surgical management.

Keywords: Pig-bel, Enteritis Necroticans, Clostridium perfringens)

Case 1.

RBT, aged 12, male, was well until 4 days before admission to Amp-Pipal Hospital when he developed a mild fever, epigastric and periumbilical pain which was initially intermittent but had become continuous. He was vomiting food and water from the second day but it contained no frank or altered blood. He had vomited one adult roundworm. His stool was of small amounts and not watery. He had eaten goat meat 5 days previously. Usually he would eat meat once or twice per month.

On examination he was mildly toxic, tachycardic (pulse 120 per minute), dehydrated, with mild generalised tenderness and no distension of abdomen. Rectal examination revealed no tenderness or masses but black liquid stool was seen on the glove.

Investigation results - Hb 11.9 gm/dl,

Total WBC count 18800. Abdominal X-Ray showed no fluid levels. Urinalysis was normal. Stool contained Ascaris Ova and Trichuris Ova. A clinical diagnosis of 'mild Pigbel' (En) was made and treatment started with intravenous infusion, intravenous benzylpenicillin and chloramphenicol and kept NPO. After 2 days of conservative medical treatment the patient's condition deteriorated. His temperature increased, pain and vomiting continued and his upper abdomen distended. Preparations were made for Laparotomy.

At Laparotomy by upper right paramedian incision there was upper intestinal obstruction about 50 cm from the duodenojejunal junction caused by an oedematous section of small bowel. The affected segment was continuous, about 50cm long and reddish-purple in colour (Photo 1). Mesenteric vessels were normal but there were multiple enlarged fleshy mesenteric lymph nodes. The rest of the small bowel was normal.

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There was no perforation. A clinical diagnosis of Pigbel was made and the whole of the abnormal segment was resected. Post operative intravenous treatment with benzylpenicillin and chloramphenicol were continued. Intravenous antibiotics were substituted by oral chloramphenicol after 5 days and oral penicillin V after 8 days.

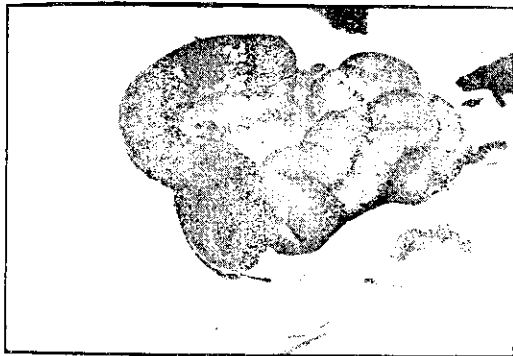


Photo. 1

The appearance of the bowel at laparotomy of the Case 1. This shows upper intestinal obstruction about 50 cm from the duodenojejunal junction caused by an oedematous section of small bowel.

Oral glucose water was started on the 4th day and milk on the 5th post-operative day. The patient was discharged home on the 10th postoperative day.

Histology report "Necrotising enteritis involving the full thickness of the bowel wall. Appearances are consistent with Pigbel".

Case 2.

HBA, aged 6 year was carried the 5 hour journey to Amp-Pipal hospital, 6 days after the onset of his illness. Loose stools and abdominal pain started 6 days previously and the following day stool was accompanied by some blood on 3 occasions. On the third day of illness he was taken to a 'medical hall' and after

vomiting he became drowsy and was transferred to a small hospital. He was treated with intravenous fluids for three days during which his abdomen gradually distended. The day before admission to Amp-Pipal Hospital he had no stool or mucus but did pass a small amount of blood per-rectum. He had eaten goat meat at a wedding 4 days prior to the onset of the illness.

On admission he was very ill, toxic and with a BP of 80/50, pulse rate of 120, dehydrated, drowsy but rousable and had no meningism. Abdominal examination revealed distension, with tenderness only on deep palpation, no masses and scant bowel sounds. Rectal examination was not painful and there was loose brown stool visible on the glove but no frank blood.

Investigations were as follows- Hb. 8.2 gm/dl. total WBC count-12400, urine and stool microscopy negative.

A provisional diagnosis of Pig-bel (enteritis necroticans) was made and treatment started with i.v. fluids, chloramphenicol and benzylpenicillin, and blood transfusion in preparation for laparotomy. Metronidazole was also given pending confirmation of the diagnosis. A single dose of piperazine was also given and then the child was kept fasting.

This regime is in accordance with the guidelines from PNG (Papua New Guinea) for treating Pig-bel.^{2,3}

After resuscitation and transfusion to an Hb of 10.3, laparotomy was performed through an upper midline incision. The whole of the small bowel was abnormal except the first 60cm of jejunum and 30cm of mid small gut. The bowel was purple, oedematous, with multiple necrotic pale green patches (1/2 - 2cm diameter) and scattered black spots (1-3mm diameter). The last 30 cm of ileum was red and slightly oedematous but showed no immediate signs of necrosis. Mesenteric lymph nodes were large and fleshy. The Large bowel was completely normal. To preserve as much bowel as possible a double end to end small

bowel anastomosis was performed, removing all bowel which was in danger of imminent perforation but not eliminating all diseased bowel.



Photo. 2

The resected specimens from case 2. The bowel is purple, oedematous, with multiple necrotic pale green patches (1/2-2cm diameter) and scattered black spots (1-3 mm diameter).

This patient had a very stormy post operative period with continuing fever, toxicity and falling haemoglobin. On the 10th postoperative day his Hb was 5.7 gm/dl and the family refused to give blood. He continued to be febrile, developed a leakage of possible small bowel fluid through a drain site and was eating poorly.

On the 24th postoperative day with continuing anaemia, sepsis and possible intra-abdominal abscess and a low output small bowel fistula and refusal of blood donation and surgery the child was taken home by his father. The prognosis at this stage was dismal.

DISCUSSION

Two cases of 'Pig-bel' (Enteritis Necroticans) successfully treated at Amp-Pipal Hospital, Gorkha District, Nepal, were presented in a recent report in the literature.¹

These were the first reports of histologically confirmed Pigbel in Nepal recorded in the Literature.

Pig-bel (Enteritis Necroticans) is a disease endemic to Papua New Guinea where much of the research has been done into the pathogenesis and treatment. It has also occurred sporadically in India, Korea, Thailand and even Europe.

Until 5 years ago this disease was unsuspected, and undescribed in Nepal, although its presence had been suggested some years before.⁴

Enteritis Necroticans, popularly known as Pigbel, has been widely reported and researched in its endemic setting of Papua New Guinea. However in 1985 a retrospective study, carried out in the two United Mission to Nepal Hospitals based in the western Hills of Nepal analysed the records of 26 cases of non specified small bowel disease variously classified under the headings- 'Intestinal obstruction (upper); Acute abdomen (unspecified); peritonitis; perforation; and enteritis'.

Analysis of the notes in that study showed an alarmingly high mortality of 62%. The conclusion was 'the probable existence of Enteritis Necroticans in Nepal', but 'the need for histological confirmation'.⁵ These two cases demonstrate a number of features about the disease and the difficulties of diagnosis and dilemmas of treatment.

The two cases are very different in their presentation. One presented early had only upper intestinal disease which was very limited in an otherwise healthy young man. Complete resection of the abnormal segment resulted in rapid and full recovery.

The second patient presented late. He was a very sick, malnourished child (weight below the 3rd centile for his age) with anaemia and with very extensive disease. This patient presented a number of surgical dilemmas, double anastomosis to preserve viable bowel, prolonged surgery. The ideal procedure is

join normal bowel to normal bowel in one stage this would have given this child a better chance of survival in the short term but possibly only a mere 30 cm of jejunum to function as small bowel in an already malnourished child. In the absence of parenteral nutrition long term survival would have been unlikely. The widespread problem of blood donation too little and too late jeopardised his chance of survival.

In its endemic setting of Papua New Guinea the disease was linked to the consumption of an unaccustomed protein load

of Pig meat at special festival times and consequently the disease acquired the local name of the 'Pig-bel'.

This term is a misnomer for Nepal as neither of the patients in Amp-Pipal presented above had consumed pig meat but both these two patients had eaten goat meat within the four days preceding the onset of illness. The other reported patients denied eating any meat in the 2 weeks before developing Enteritis necroticans.¹ Current theoretical pathogenesis of Pigbel is shown in Fig. 1.

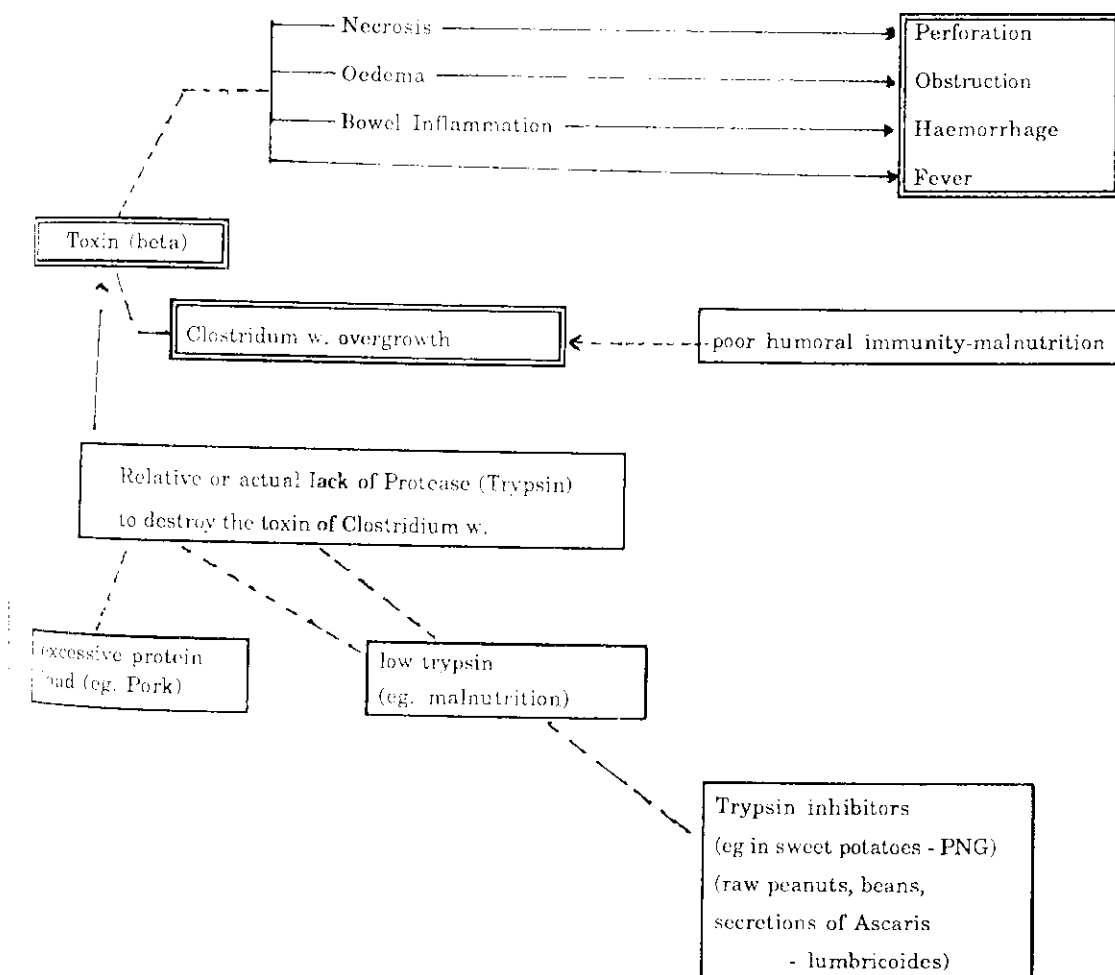


Figure 1: Current theoretical pathogenesis of Pigbel^{7,8}

The underlying pathological process is the production of B-toxin by *Clostridium welchii* type C (perfringens) leading to toxic effects on the bowel and the clinical syndrome of inflammation, oedema, necrosis, haemorrhage, and ultimately obstruction and perforation. Factors leading to the disease are as follows:

Firstly, the population of *Clostridium* producing toxin in the gut may be determined by environmental and dietary factors. In Papua, fifty percent of the population have this organism as a commensal and poorly cooked pig meat also contains a significant bacterial load. Reduced humoral immunity in undernutrition may lead to overcolonisation of the gut.

Secondly, the amount of B-toxin available to cause damage is determined by a number of other factors both agonistic and antagonistic. For example, the protease Trypsin, which destroys the toxin, may be overloaded by an unaccustomed protein load in the diet. Alternatively, Trypsin antagonists (found in sweet potatoes, the secretions of round worms and raw peanuts) may prevent the Trypsin's normal protective effect. Trypsin itself may decrease in malnutrition allowing the harmful effects of the toxin.

An imbalance of these factors, and probably other factors as yet undiscovered determine the onset of the disease.

Treatment

Treatment may be given on the basis of certain definite criteria at presentation (Fig 2) and subsequently changed on the basis of progress or deterioration.

1. Medical treatment - for 'mild' cases (i.e. those without marked distension, toxicity or blood or black flecks in vomitus or gastric aspirate).

- a. intravenous infusion
- b. Antibiotics - benzylpenicillin and chloramphenicol i.v. then oral chloramphenicol if improving.

- c. Piperazine stat then NPO
- d. Nasogastric continuous drainage

If improving after 3 days (distension, pain and vomiting less, bowels functioning and the patient is hungry):

- 4th day stop i/v and give glucose water
- 5th day give milk
- 6th day give food

If not improving or develops 'severe' criteria prepare for Laparotomy.

	Nepal ⁵	Khymer ⁸
Abdominal Pain 12 to 96 hours after a protein meal-colicity.	62%	78%
Distension- mild to marked (*)	72%	-
Vomiting -	62%	73%
Vomit - or aspirate (via NG tube) containing black flecks (*)		9%
Diarrhoea with or without blood followed by constipation		94%
Ascaris may be passed in stool or in vomitus		90%
Toxicity (*)	92%	
Fever	77%	
Bowel sounds absent or diminished		
X-Ray evidence of obstruction	100%	100%

Figure 2: Symptoms and signs - as a guide to management (those marked with * are indicators of 'severe' disease)

2. Surgical treatment-

* Indications

- 'severe' criteria present or developing
- not improving on medical treatment 3 days
- perforation suspected
- persistent bleeding
- other diagnosis suspected and needing surgical intervention (see differential diagnosis - figure 3)

- | | |
|----|---|
| 1. | Dysentery/gastroenteritis |
| 2. | Intussusception |
| 3. | Typhoid |
| 4. | Appendicitis |
| 5. | Other causes of obstruction/perforation/peritonitis |

Figure 3: Differential diagnosis

Principles of surgical treatment

- a. resuscitation and medical treatment as above
- b. Transfusion if Hb less than 10 gm/dl
- c. Operative guidelines

* Upper Midline (or Right Paramedian) incision extended as necessary

* Findings-jejunum is usually involved and sometimes duodenum to ileum. From 50 cm up to 200 cm of intestine will be oedematous, red or plum coloured with possible black or pale yellow necrotic lesion 2-5 cm in diameter or frankly gangrenous sections. The affected section may be continuous or in skip lesions. The mesentery contains oedematous, large fleshy lymph nodes. Sometimes gas bubbles will be seen in the bowel wall. Perforations or abscesses may be found.

* Procedure - decompression, and ideally resection abnormal bowel and anastomosis of normal tissue. However some compromise may have to be made in order to leave some small bowel in very extensive disease. Use peritoneal lavage and drains as needed.

* Post-operative. Avoid haste in reintroducing oral protein. When bowel sounds return and flatus is passed usually by 4th or 5th

day water and then glucose water can be introduced. Thereafter milk and soft carbohydrates can be cautiously reintroduced. Antibiotics should be continued for approximately 10 days post operatively. Intravenous chloramphenicol being substituted by oral when oral fluids are tolerated.

Prognosis 15 - 40% mortality.

Prevention: Immunisation with a b-toxoid is indicated in endemic areas but not in areas of sporadic occurrence like Nepal.

CONCLUSION

Enteritis Necroticans has been positively identified in Nepal and can be treated with the combined efforts of physicians and surgeons according to clear criteria. It is hoped that as it is increasingly recognised in clinical practice we in Nepal can benefit from the experience in management gained by doctors in the highlands of Papua New Guinea.

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