Necrotizing Enteritis Simulating Pig-Bel Disease in Northern India

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Abstract

Objective: A clinicopathological study of patients presenting with necrotizing enteritis, similar to the Pig-Bel disease of Papua New Guinea.

Methods: A clinicopathological study of 95 patients presenting in last ten years with acute abdominal pain with diarrhea and/or bleeding per rectum and vomiting and found to have lesions similar to Pig-Bel disease.

Results: 89% of the patients were young adult vegetarians from lower socio-economic strata. 94.5% of cases presented between May and August every year showing a marked seasonal variation. Jejunum was the most common site (95%) involved. Gross examination showed blackish brown ‘skip’ lesions with gangrene and perforations. Histology showed marked necrosis and congestion of all the layers with mucosal ulcerations.

Conclusion: A disease simulating Pig-Bel disease occurs in India. It is more common in young adult vegetarians and may have a different etiology from that proposed in endemic regions.

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Introduction

Necrotizing enteritis (enteritis necroticans) is a rare disease affecting the small intestine, mainly the jejunum. It has been reported from various parts of the world and is a major health problem in the highlands of Papua New Guinea where it has been linked to the consumption of putrid pig meat and is locally known as Pig-Bel. It is thought to be caused by Clostridium welchii type C and active immunization with A toxoid derived from type C clostridial cultures has been shown to provide protection. A few reports are also available from India. We report our experience of 95 such patients seen over a period of ten years.

Methods

A clinicopathological study of 95 cases with necrotizing enteritis, seen during the period 1984-1993, was carried out. Of all the patients presenting with acute abdominal pain with diarrhea and/or bleeding per rectum and vomiting, only those who showed ‘skip’ lesion on gross with classical histological findings of necrotizing enteritis were included in this study. A clinical history with special emphasis on socio-economic status and diet was taken. A detailed clinical examination was performed.

Investigations carried out included total and differential leukocyte counts, stool examination for parasites and occult blood, and an erect plain abdominal X-ray (in 61 patients). In the remaining patients, X-ray could not be obtained due to lack of X-ray facilities during emergency hours. The findings at laparotomy were noted in all cases, with special reference to the part of the bowel affected, the length of segment involved and presence of ‘skip’ lesions. Gross and histological features were also noted. The lesions in necrotizing enteritis are small, necrotic, black patches scattered throughout the affected segment. These lesions help to differentiate it from ischemic enteritis where usually confluent involvement of variable length of gut wall is seen depending upon the site of blood flow occlusion.

Results

Of the 95 patients, 74 (78%) were men. Their mean (±SD) age was 24.2 (±13.9) yr (range 5-50). Four patients were below 10 years of age. Eighty two (86%) patients were from rural areas, 89 (94%) had a low socio-economic status and 84 (89%) were vegetarians. Eighty nine (94%) patients presented during the summer and the rainy seasons (between May and August).
Clinical profile
The mean (±SD) duration of symptoms was 2.9±1.1 days (range 1-5). All patients presented with acute abdominal pain which was aggravated by food or drinks. All patients had been healthy prior to the onset of pain. Ninety-two (97%) patients complained of fever, weakness and lethargy while 35 (37%) and 26 (27%) patients had vomiting and 3-4 loose stools per day, respectively. Nineteen (20%) had frank blood in stools. There was no family history of similar illness in any case.

Examination revealed mild pyrexia (37.2 - 38.3°C) in 55 (58%) patients and dehydration in 60 (63%). Abdominal examination revealed tenderness in 88 (93%) patients and distension in 59 (62%). The bowel sounds were feeble or absent in 82 (86%) patients.

Investigations
Investigations revealed polymorphonuclear leukocytosis in 93 (98%) patients. Stool contained ova of Ascaris lumbricoides, Ankylostoma duodenale and Enterobius vermicularis in 38 (40%), 15 (15.8%) and 6 (6.3%) cases respectively. Occult blood was positive in 59 (62.1%) cases. Abdominal X-rays showed generalized gaseous distension in 35 of 61 (57%) patients in whom it was performed. Air-fluid levels and free air in the peritoneal cavity were seen in only three and two patients respectively.

Treatment and findings at laparotomy
Initial treatment consisted of nil orally, intravenous fluids and antibiotics (ampicillin, gentamicin and metronidazole). All the patients were later subjected to semi-elective laparotomy which revealed a small amount of straw-colored fluid in the peritoneal cavity and severe inflammation, marked congestion and thickening of the bowel wall with gangrene and perforation at places. The diseased bowel was resected and end-to-end anastomosis performed; the length of bowel resected varied from 30 to 180 cm. Fourteen (15%) patients died in the postoperative period. These patients had involvement of a large part (> 100 cm) of bowel (10; 10.5%) or perforation (7; 7.3%) or were brought to the hospital in a toxic state (12; 12.6%).

Gross pathology
Jejunum alone was the most commonly affected site (82; 86.4%), followed by both jejunum and ileum (8; 8.4%) and ileum alone (5; 5.2%). The resected specimens showed intense congestion and thickening of the intestine, with blackish brown serosal surface, in a skip fashion (Fig 1). Perforations were also seen at 1-3 places in seven specimens. The mucosa was denuded at places, with dirty brown material spread over it.

Histology
The predominant histological changes observed were varying degree of necrosis of the intestine with acute (polymorphonuclear) inflammatory exudate (Fig 2). The intestinal wall thickness varied in different specimens and even in the same specimen at different sites. In 72 (75.8%) cases, necrosis was transmural; of these, 7 patients had perforation. In 7 (7.3%) cases, necrosis was limited to the mucosa only while in 16 cases, it was intermediate between these two extremes.

The submucosa was specially searched for presence of hyaline thrombi and gas bubbles (pneumatosis); the former were seen in 3 (3.1%) cases but no case showed presence of gas bubbles. Lymph nodes showed non
specific reactive lymphadenitis either in the form of sinus catarrh or follicular hyperplasia.

Discussion

Necrotizing enteritis, also known as Pig-Bel disease because of its association with pig meat consumption, has been mainly reported as endemic disease from Papua, New Guinea. Although clinically and histologically similar cases have been reported from India and Nepal, their presentation has been somewhat different.

The mean age of presentation in our patients was 24.2 ± 13.3 years and only four of 95 patients were below 10 years of age, in contrast to many of the previous reports where most patients were below 10 years. In another report from India, 73% of the patients were between 20 to 50 years of age. The male : female ratio of 3:5:1 was similar to the previous report of Pujari and Deodhar.

The disease is currently believed to be caused by the beta toxin of Clostridium welchii type C which produces inflammation, necrosis and hemorrhage followed by perforation. This is mainly based on studies in Papua New Guinea, an endemic area, where 50% of the population harbors C. welchii as a commensal in the gut. It is believed that consumption of inadequately cooked pig meat further increases the bacterial load and causes the disease. However, our observation that 89% of the Indian patients were vegetarians casts a doubt on this theory. Other Indian studies have also reported similar findings. Two cases reported from Nepal too did not consume pig meat.

Most of our cases were observed during the summer and rainy seasons (94%). This is similar to the previous Indian studies and suggests an infective etiology. In a previous report from Germany too, seasonal variation was observed.

Lawrence et al suggest that prolonged malnutrition, protein deficiency and presence of trypsin inhibitors in diet may lead to a failure to destroy organisms and their toxins in the gut. Sweet potatoes and raw peanuts are rich in trypsin inhibitors and form a major component of diet in rural areas and among lower socio-economic groups during summer and rainy seasons. Most (86%) of our patients came from rural areas and 94% belonged to the lower socio-economic group. Worm infestation was also common and may increase during summer and rainy seasons. Secretions of round worm may contain trypsin inhibitor. Thus, in India and Nepal, a combination of protein deficiency and worm infestation may be responsible for the causation of this disease by reducing the amount of trypsin available in the gut, leading to overcolonization of the gut by toxin-producing bacteria.

There are minor differences in the histological picture of necrotizing enteritis and ischemic enteritis in the form of thickness of the mucosa involved, presence of hyaline thrombi in submucosal blood vessels, submucosal hemorrhage and gas bubbles in submucosa (pneumatosis). Presence of hyaline thrombi and submucosal hemorrhage is more common in ischemic enteritis than necrotizing enteritis. In the present study thrombi were seen in only 3 (3.1%) cases, similar to the study by Pujari and Deodhar.

Transmural involvement of mucosa was seen in 72 (75.8%) cases. Necrotizing enteritis is more in favor of necrotizing enteritis as ischemic lesions rarely show transmural necrosis. Pneumatosis is a peculiar feature seen in necrotizing enteritis but none of our cases showed it. Despite these relative differences between the two lesions it is not possible to differentiate them only on histological findings in a number of cases. In such cases clinical history and presence of 'skip' lesions on gross examination may be helpful.

References