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Case Report

Acute Segmental Necrotising Enteritis, Presenting with Transient Protein Losing Enteropathy and Bleeding Diathesis in a Child Managed Surgically: A Case Report

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Abstract

Background: Necrotizing enteritis is a rare cause of small intestinal obstruction reported mainly from Asian sub-continent. The clinical presentation is of acute gastroenteritis followed by features of bowel obstruction. We present a child who had a rare complication of necrotizing enteritis, unreported in literature.

Case Characteristics: Five year old girl presented with 1 month history of worsening vomiting, altered bowel habits and recently developed petechiae and pedal edema. Child had features of small bowel obstruction and after resuscitation underwent emergency surgery.

Intervention/Outcome: Laparoscopy showed features of necrotizing jejunitis and a formal laparotomy with resection of diseased segment lead to rapid recovery.

Conclusion: High index of suspicion is needed to diagnose necrotizing enteritis. Though most cases can be managed conservatively, clinical deterioration and development of complications may necessitate surgical intervention.

Introduction

Necrotizing enteritis is a rare cause of small intestinal obstruction reported mainly from Asian sub-continent. The clinical presentation is of acute gastroenteritis followed by features of bowel obstruction. Conservative management is successful in majority of cases and surgical intervention may need in complicated disease. In the past, the disease had high mortality and morbidity and in recent years the prognosis has improved significantly.

Case Report

A 5 year old Indian girl living overseas (Dubai) was admitted for evaluation of sub acute bowel obstruction (abdominal pain and non bilious vomiting) for the past 1 month, which followed an episode of acute diarrhea. Her symptoms got worsened recently, vomiting became bilious and she developed malena. Clinically the child was dehydrated with anemia and had petechiae and echymoses over the body. There was mild abdominal distension with tenderness in the epigastric and umbilical region with absent bowel sounds. Laboratory investigations showed anemia, leukocytosis (with shift to left), hypoalbuminemia, prolonged Prothrombin time, thrombocytopenia and electrolyte disturbances. There was no evidence of henoch schonlein purpura and blood cultures were negative for organism. The plain X-ray abdomen was unremarkable, except for ground glass appearance suggestive of peritonitis. The ultrasound and CT scan of abdomen showed focal thickening of a segment of upper jejunum. The child was resuscitated with parenteral fluids, fresh frozen plasma, vitamin K and broad-spectrum antibiotics were started. The child's condition worsened and developed features of acute intestinal obstruction necessitating an emergency surgical exploration. On diagnostic laparoscopy there was thickening and hyperemia of entire circumference of a short segment of proximal jejunum. The mesenteric vessels appeared uninvolved. A formal laparotomy [Figure 1] with resection of a 2-inch segment of necrotic jejunum (completely occluded by sloughed mucosa) and primary anastomosis was performed. Postoperatively the bleeding diathesis resolved within 24 hours and oral feeds were tolerated from day 4. The child recovered dramatically and was discharged home after a week. Histopathology confirmed it to be necrotizing jejunitis [Figure 2]. The child got readmitted after a month with early postoperative adhesions, which resolved

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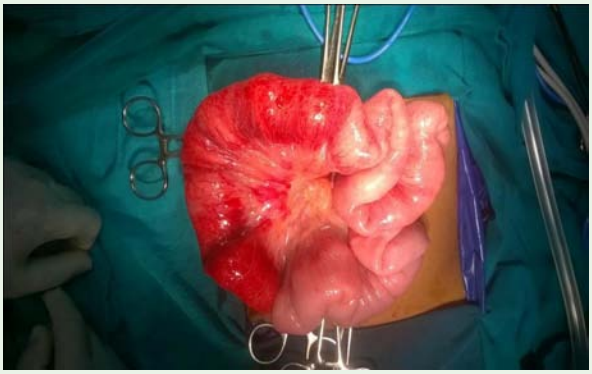

Figure 1: Localised segmental Ischemia of proximal jejunum on laparotomy.

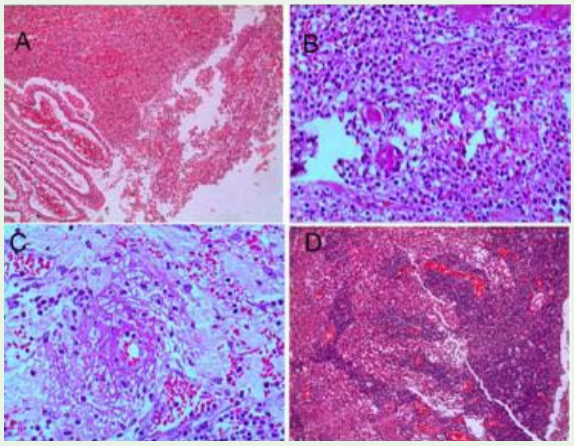

Figure 2: Histopathology of resected jejunum. **A.** Mucosa showing multiple foci of ulceration. **B.** Dense neutrophilic infiltration and few hyaline thrombi in submucosa and vessels. **C.** Submucosal vessels showing fibrinoid necrosis. **D.** Lymph node in 10x power showing distended sinuses and medullary cords with neutrophils.

with conservative management. At 6 months follow up the child is symptom free and thriving well.

Discussion

Acute segmental necrotizing enteritis is also known as necrotizing enteritis [1], non-specific jejunoileitis [2], acute segmental enteritis [3], acute necrotizing jejunoileitis [4], enteritis necroticans [5], pig-bel (in Papua New Guinea) and dermbrand (in Germany) [6], Mainly a disease of adults, it has been described in children especially from Asian sub-continent [5]. Distinct from the Necrotizing Enterocolitis [NEC], which is seen in neonates, this disease affects mainly the jejunum, to a lesser extent ileum and usually spares colon.

The etiology is unclear, but various putative factors like seasonal variation, dietary factors (sweet potato tryps in inhibitor), bacterial toxins (*clostridium welchii*), parasitic infestations (ascariasis, trichinosis) have been implicated [7] and may occur as a sequel to gastroenteritis [8]. In a series of 904 cases of intestinal obstruction reported by Gopi VK *et al*, necrotizing enteritis was the commonest cause [9].

The pathogenesis may be due disturbances of microcirculation occurring during low flow states, as the mucosa and submucosa are predominantly affected with characteristic absence of gross mesenteric vascular occlusion, similar to Neonatal NEC. In the more common Neonatal NEC that occurs predominantly in the very low birth weight and/premature infants, the intestinal mucosa is vulnerable to ischemia and immature immune function predisposes to infection [10] The initial insult is low flow states/hypoxia (Diving reflex theory) [11] and further ischemia may be potentiated by release of vasoconstrictive substances with secondary bacterial infection leading to bowel necrosis. Our patient was healthy before the episode without any such predisposing factors except for diarrhea. Hypersensitivity reaction (type 1 or type 3) to bacterial or parasitic infestation has also been incriminated [12] as the disease is more common in developing nations, where parasitic infestation are more common. Enteritis necroticans also called as pig-bel is endemic to Papua New Guinea and has been attributed to nutritional deficiency, eating half cooked pork meat and sweet potato. The necrotic intestinal lesions are probably due to beta enterotoxin from Cl. Welchii infection [6]. None of these risk factors were evident in our index case. The pathogenesis in our case is obscure as the laboratory findings were negative for any specific cause. The inciting factor could have been the gastroenteritis with subsequent mucosal injury/edema causing intestinal bleed and protein loss. Protein losing enteropathy in segmental jejunitis has also been described previously [7], which could due to protein loss from edematous intestinal mucosa and subsequent lymphatic edema and obstruction. Our patient also had petechiae and echymoses with prolonged prothrombin time, which was not attributable to any specific bleeding diathesis. This sort of presentation in necrotizing jejunitis has not been previously reported and we suspect it to be due to endotoxemia-induced coagulopathy. The blood culture failed to grow any organism as the child was already on antibiotics.

Grossly the intestinal lesions are mainly confined to segmental areas of jejunum and to a lesser extent ileum. The bowel appears congested, edematous and often hemorrhagic with sparing of mesenteric vessels. Microscopically there is necrosis of mucosa and congestion of submucosa, which can be full thickness in advanced cases and pan cellular inflammatory infiltrates in the submucosa. Smaller submucosa arteries may show vasculitis with perivascular infiltration and fibrinoid necrosis. (Suggestive of type 3 hypersensitivity reaction).

Clinical course of disease is variable. As the diagnosis can only be established after surgical exploration [4], mild disease may go unnoticed. Our index patient was initially managed conservatively, but underwent surgical exploration due to worsening obstruction, protein losing enteropathy and generalized bleeding tendency.

When necrotizing jejunitis is suspected clinically, the management should be conservative, but surgical treatment in the form of resection of segment involved is usually curative in children with advanced disease as the mortality is high [13]. Our patient had features of complete bowel obstruction with protein losing enteropathy and bleeding diathesis, which resolved following surgical resection. Rare instances of recurrence after surgery are known [4] and late intestinal strictures attributable to unrecognized acute episodes or after treatment are also reported [4,14].

Conclusion

Segmental necrotizing enteritis is a rare cause of intestinal obstruction in children reported mainly from Asia. As definite

etiological factor is unknown, a high index of suspicion is required for early diagnosis, when a child with acute gastroenteritis develops features of acute abdomen. Majority of patients can be managed conservatively when other causes of mechanical obstruction have been ruled out and surgical treatment is indicated in patients with failed medical management, worsening of obstruction and development of complications.

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