

Journal of Pediatric Sciences

BOWEL PERFORATION IN NEUTROPENIC CHILD

Necrotising enterocolitis or not a case of necrotising enterocolitis

Basant Kumar, Vijay D Upadhyaya, Pradeep Kumar, Rajat Gupta

Journal of Pediatric Sciences 2011;3(6):e112

How to cite this article:

Kumar B, Upadhyaya VD, Kumar P, Gupta R. Bowel perforation in neutropenic child. Necrotising enterocolitis or not a case of necrotizing enterocolitis. Journal of Pediatric Sciences. 2010;3(6):e112

CASE REPORT

Bowel perforation in neutropenic child: necrotizing enterocolitis or not a case of necrotizing enterocolitis?

Basant Kumar, Vijay D Upadhyaya, Pradeep Kumar, Rajat Gupta

Abstract:

Neutropenic enterocolitis (NE) or typhlitis is a life-threatening, necrotizing process of intestine; occurring most frequently after intensive chemotherapy in acute leukaemias. Its presentation and diagnostic criteria are heterogeneous and even the histological diagnosis shows a considerable diversity. We report an unusual presentation of bowel perforation in a child with suspected NE.

Keywords: Necrotizing enterocolitis, parietal abscess, acute leukemia

Received: 15/10/2011; **Accepted:** 22/11/2011

Neutropenic enterocolitis (NE) is a well known life threatening entity in patients who have severe neutropenia; especially when complicated by bowel perforation [1,2]. It occurs most often in patients receiving intensive chemotherapy for acute leukemia, but it has been observed in patients who are neutropenic from other causes as well. [1-3]. Usually the condition is localized to the terminal ileum, caecum or ascending colon but in rare circumstances; more distal bowel may be involved [1]. Various clinical and radiological presentations are well described in literature. The diagnostic criteria are heterogeneous and even the histological diagnosis shows a considerable diversity [4]. The clinical picture is usually characterized by the triad of fever, abdominal pain and diarrhea, but other complications may present these same signs and symptoms, and thus a definite diagnosis may be difficult to establish [5]. We present our experience of an unusual presentation of bowel perforation in a child with suspected NE; presented as parietal wall abscess with enterocutaneous fistula without bowel obstruction or obvious peritonitis.

CASE REPORT

A 10 years old male child weighing 20 kg, presented with high grade fever and pain in right lower abdomen; after 7 days of chemotherapy (BFM-95 MR protocol, 1st cycle) for acute lymphoblastic leukemia (ALL). On examination, abdomen was soft with palpable bowel loops in right iliac fossa. Mild to moderate tenderness and guarding was

present at right iliac fossa but generalized sign of peritonitis was absent. There was restriction and pain during flexion of the right hip joint. He was anemic and total leucocytes count (TLC) was 1200/ μ L. Ultrasound showed bowel wall thickening(> 4 mm) with minimal free fluid in the right iliac fossa. He was treated with bowel rest, nasogastric drainage, and broad spectrum antibiotics / antifungal along with supportive therapy and granulocyte colony-stimulating factor. Over the next few days, erythema and indurations were evident over the right

Kumar B^{1*}, Upadhyaya VD¹, Kumar P², Gupta R¹

¹Department of Pediatric Surgery, Sanjay Gandhi Post Graduate Institute of Medical Sciences, India

|

²Department of Hematology, Sanjay Gandhi Post Graduate Institute of Medical Sciences, India

Corresponding Author:

*Basant Kumar, Department of Pediatric Surgery, Sanjay Gandhi Post Graduate Institute of Medical Sciences, Raibareli Road, Lucknow, U.P., India
E-mail: kumardrbasant@yahoo.com
Phone: 91-800- 4904839

lateral abdominal wall without guarding or rigidity. Review ultrasound showed parietal wall abscess and thickened bowel loops with minimum free fluid in the right iliac fossa. Latter, overlying skin was necrosing with discharge of sero-purulent fluid. Serial abdominal X-rays showed dilated bowel loops without air-fluid levels or free intra-peritoneal gas or pneumatosis intestinalis. But, there was persistent huge parietal wall gas shadow in the abdominal films. Urgent computed tomography (CT) scan was done which showed thickened (> 4 mm) collapsed large bowel, parietal wall abscess with gas in parietal muscles and minimal fluid in ileo-caecal region (**Figure-1**). Fistulous communication between the bowel and abscess cavity could not visualized. Child was explored via lower midline incision under GA. Operative findings were dilated small bowel, local adhesion with minimal fluid, collapsed perforated caecum communicating with large abscess cavity opening at skin. The bowel, lying in the vicinity of abscess cavity: were thickened and inflamed while rests of the bowel were healthy. Appendix was thickened but not perforated. After debridement and vigorous cleaning, closure of caecal perforation with appendectomy was done and proximal loop ileostomy was created. Biopsy was taken from the perforated site. Wound infection and superficial wound dehiscence in post operative period was managed by conservative means (**Figure-2**). Histopathology showed transmural necrosis with mixed inflammatory cells. Focal areas showed granulation tissue. Child is under follow-up and is doing well.

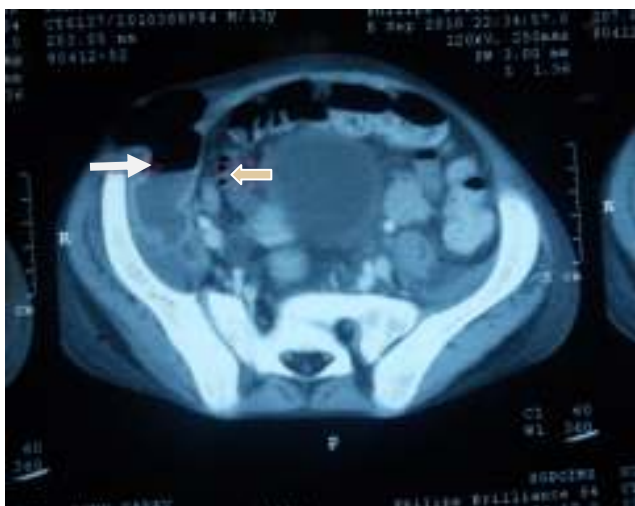


FIGURE – 1: CT-scan abdomen showing collapsed caecum (left arrow) and abscess cavity with gas shadow (right arrow).



FIGURE –2: Postoperative day 15th showing granulations in wounds with ileostomy.

DISCUSSION

The true incidence of neutropenic enterocolitis is unknown, but may be 5.3 % or more (0.8 % to 26 %) among adult patients receiving chemotherapy for solid malignant tumors [4,6]. The incidence is reported to be slightly lower in children [6]. Mortality rates are reported currently to be between 30 % to 50 % or even more (≥ 50 %) [3,4,6]. This case presentation is one in which NE was diagnosed after chemotherapy for hematological malignancy. Management of these conditions is challenging. The initial management for patients who do not have gastro-intestinal bleeding, peritonitis or intestinal perforation; typically entails bowel rest, decompression, antibiotics, nutritional support including administration of granulocyte colony-stimulating factor [1-6]. Surgical management is generally reserved for patients with perforation or those whose condition deteriorates clinically. Shamberger *et al* [7] proposed objective criteria for immediate surgical treatment: (a) persistent gastrointestinal bleeding in spite of the resolution of neutropenia, thrombocytopenia or clotting abnormalities; (b) free intraperitoneal perforation; and (c) clinical deterioration suggesting uncontrolled sepsis [7]. NE is characterized by transmural inflammation and edema of bowel wall followed by ulceration, necrosis and perforation. The exact pathogenesis is also unknown. Gut mucosal ulcerations may result from direct drug related cytotoxicity or from neutropenia itself; facilitates

microbial invasion in the bowel wall [3,6]. Macroscopically, the involved bowel segments show edematous or thickened walls with varying degree of ulceration and hemorrhage; and perforation occurs in 5% - 10% of cases [5]. Prominent microscopic findings include sub mucosal edema, hemorrhage and necrosis with inflammatory infiltrate [5].

The classical clinical presentation includes fever, abdominal pain and neutropenia but diagnosis is often hindered by subtle or non-specific clinical findings [4]. Although ultrasonography and radiological imaging including computed tomography are excellent modalities to diagnose early complications; but some time; it is not possible to find radiological image early in the course of disease, as occurred in our case. The term "neutropenic enterocolitis" contains following elements: (a) the patient must be neutropenic, (b) there is an inflammation, (c) this inflammation involves the gut, (d) an infection is causative, (e) the infection is invasive and accordingly and (f) *C. difficile* is at least not the leading pathogen [4]. The suggested diagnostic criteria for neutropenic enterocolitis includes: (a) *presence of fever* (axillary temperature >38.0°C or rectal temperature >38.5°C); (b) *abdominal pain* (at least degree 3 determined by the patient using a visual analogous scale pain score ranging from degree 1 to 10); (c) demonstration of the *bowel wall thickening* of more than 4 mm (transverse scan), over more than 30 mm (longitudinal scan) in any segment by US or CT [4].

The survival of patients with NE has been linked to neutrophil recovery [2]. In our patient, aggressive medical therapy including granulocyte infusion, failed to prevent delayed bowel perforation. The typical signs of perforation were absent and emergent surgery resulted in a satisfactory outcome even at low TLC count (2300/ μ L). Despite of successful treatment, diagnosis of NE is still in doubt. It is very clear that the patient developed an abscess and presented with flexion deformity at right hip joint without obvious abdominal pain and peritonitis; and latter enterocutaneous fistula was developed. The only evidence of NE are neutropenia and bowel wall thickening in US and CT-scan; but during surgery, it was found that much of the patient's bowel was "healthy" away from the abscess. Was it NE leading to abscess and enterocutaneous fistula or parietal wall abscess causing bowel wall inflammation and thickening? The wide spectrum of clinical presentation requires an individualized approach to therapy. This case exemplifies the need for early operative intervention in this life threatening situation.

In conclusion, it is advisable that a high index of suspicion is needed for all patients who present with fever and abdominal pain in the setting of neutropenia and is also advisable to surgeons for early surgical intervention which

is mandatory for peritonitis, perforation and in persistent gastrointestinal hemorrhage.

REFERENCES

1. Katz JA, Wagner ML, Gresik MV, Mahoney DH, Fernbach DJ. Typhilitis. An 18-year experience and postmortem review. *Cancer*, 1990; 65 (4): 1041-1047.
2. Wade DS, Nava HR, Douglass HO Jr. Neutropenic enterocolitis. Clinical diagnosis and treatment. *Cancer*, 1992; 69 (1): 17-23.
3. Wilson DB, Rao A, Hulbert M, Mychaliska KP, Luchtman-Jones L, Hill DA, *et al.* Neutropenic enterocolitis as a presenting complication of acute lymphoblastic leukemia: An unusual case marked by delayed perforation of the descending colon. *J Pediatr Surg*, 2004; 39 (7): e18-e20.
4. Gorschluter M, Mey U, Strehl J, Ziske C, Schepke M, *et al.* Neutropenic enterocolitis in adults: systematic analysis of evidence quality. *Eur J Haematol*, 2005; 75: 1-13.
5. Gomez L, Martino R, Rolston K V. Neutropenic Enterocolitis: Spectrum of the disease and comparison of definite and possible cases. *Clin Infect Dis*, 1998; 27: 695-699.
6. Ullery BW, Pieracci FM, Rodney JR, Barie PS: Neutropenic Enterocolitis. *Surg Infect (Larchmt)*, 2009; 10 (3): 303-314.
7. Shamberger RC, Weinstein HJ, Delorey MJ, Levey RH. The medical and surgical management of typhilitis in children with acute nonlymphocytic (myelogenous) leukemia. *Cancer*, 1986; 57: 603-609.