Case Reports

Neutropenic Enteropathy

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Neutropenic enteropathy (NE) is used to describe the inflammation of the bowel in neutropenic patients under aggressive chemotherapy, mainly for Iymphoproliferative and hematologic malignancies .Surgical intervention may be required in patients with the advent of the disease. We report our experience in 7 children with NE who had to be treated surgically. Absolute neutrophil counts were less than 1000/mm^3 in all, with positive blood cultures in five patients. Four patients recovered with rapid resolution of neutropenia, while three patients died with persistent neutropenia.

Key words: *Intestinal perforation, Neutropenic enteropathy, Typhlitis.*

Neutropenic enteropathy (NE) is used to describe the inflammation of the bowel (mostly the cecum and the ascending colon) in neutropenic patients under aggressive chemotherapy, mainly for lymphoproliferative and hematologic malignancies(1,2). Although, the initial treatment of choice is nonoperative treatment with bowel rest, decompression, nutritional support and appropriate antibiotics, operative intervention is needed in patients with the advent of the disease(3,4). We treated 7 such children during last 5 years.

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Case Report

Table I highlights the clinical details, laboratory findings, intervention and outcome of seven cases of neutropenic enteropathy. All patients were undergoing cancer chemotherapy and required pediatric surgery consultation because of abdominal pain, vomiting and abdominal distension. The neutrophil counts were less than 1000/mm^3 in all children at presentation.

All patients were treated with bowel rest, nasogastric decompression, parenteral nutritional support and antibiotics. Plain abdominal radiographs and abdominal ultrasound revealed dilated, thick walled small bowel loops with air-fluid levels, free intraperitoneal fluid (n = 6) and free intraperitoneal air (n = l). In two patients, surgery was indicated immediately after suspicion of the intestinal perforation. In others, progressive and/or persistent clinical and radiological findings let us to explore the abdominal cavity, within 3-5 days.

Free intraperitoneal fluid, hemorrhage, marked edema of the bowel wall, patchy inflammation and localized abscess were the main operative findings. The lesions were mostly localized at terminal ileum and the ascending colon. In one patient, perforation site was the posterior wall of the stomach with diffuse gastrointestinal fungal plaques.

The histopathological findings were mucosal and transmural hemorrhagic ulceration of the bowel (and stomach), with

TABLE I–*Details of Children with Neutropenic Enteropathy.*

Case	Age at diagnosis	Sex	Primary disease	Neutrophil count/mm^3	Blood culture	Operative findings	Operation	Course
1	6yrs	M	NHL	800	+	Multiple ileal perforations	ileostomy	died
2.	9 mo	F	NHL	600	+	Multiple ileal perforations	anastomosis	died
3.	5 yrs	F	ALL	100	+	ileal performation	ileostomy	alive
4.	1 yrs	F	AML	300	-	segmental necrosis jejunum	anastomosis*	died
5.	8 yrs	M	NHL	6400	-	perforation of terminal ileum	ileostomy	alive
6.	6 yrs	F	HL	3200	+	gastric perforation	primary repair	alive
7.	16 yrs	M	AML	1900	+	patchy inflammation of terminal ileum	appendec -tomy	alive

* required ileostomy at a second operation due to anastomotic dehiscense
NHL: NonHodgkin lymphoma, ALL: Acute lymphoblastic leukemia, AMI: Acute myeloblastic leukemia, HL: Hodgkin lymphoma.

perforation in 6 patients. In surviving children, neutropenia improved following surgery.

Discussion

Cooke was the first to describe submucosal hemorrhage and appendiceal perforation in children with leukemia(5). Later, autopsy reviews demonstrated pathological findings of the bowel in patients who died during induction or consolidation therapy. A disease process, called “typhlitis”, “neutropenic enterocolitis” or “ileocecal syndrome” is usually found in the terminal ileum, ascending colon and cecum. Although its exact pathogenesis is not clear, it is thought that chemotherapy may damage the gastro-intestinal tract (whether infiltrated with the primary disease or not) by destroying the rapidly dividing mucosal cells, which when coupled with neutropenia allows bacterial invasion of the bowel wall(2,6). A close relationship between the use of cytosine arabinoside and subsequent perforation has also been reported by several authors(1,4,7). Arabinoside-C was being used in 4 of our patients.

Recovery of the leucocyte count is fundamentally related with the survival of patients. Prolonged leukopenia may allow continued bacterial invasion of the bowel wall

with persistence of the bowel lesion, followed by necrosis and perforation(4,8). We recognized the clinical findings of neutropenic enteropathy on an average of 4 days after the onset of chemotherapy-induced neutropenia. Persistence of neutropenia should also be noted in our patient in whom surgical intervention did not provide regression of the process. The ongoing ileus with impaired vascularity, severe cell mediated immune defect with or without infection with *Candida albicans* or other opportunistic organisms, would not permit healing of the anastomosis.

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Hereditary Sensory Autonomic Neuropathy Type IV

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Hereditary sensory autonomic neuropathy Type IV is an autosomal recessive disorder due to lack of maturation of small myelinated and unmyelinated fibers of peripheral nerves, which convey sensation of pain and temperature, therefore, resulting in self mutilation. There is anhidrosis due to lack of innervation of normal sweat glands resulting in recurrent episodes of hyperpyrexia. The clinical presentation of two children with this rare disease is described.

Key words: *Hereditary sensory autonomic neuropathy, Insensitivity to pain, Self-mutilation.*

Hereditary sensory and autonomic neuropathy (HSAN) is a rare syndrome

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