

Nonspecific Necrotizing Jejunitis: A New Challenging Entity

S H MANSOOR* U F YOUSAF** B A SULEMAN***A I QURESHI*

*Department of Paediatric Surgery, Services Hospital, Lahore. ** Department of Paediatrics, Sheikh Zayed Hospital, Lahore. *** Department of Pathology, Sheikh Zayed Hospital, Lahore.

Correspondence To: Syed. Haroon Mansoor

Non-specific Necrotizing Jejunitis (NNJ) is an uncommon paediatric surgical problem. Nine cases of NNJ presenting as acute abdomen were studied. Exploration revealed ischemic changes in small intestine especially in jejunum varying from punctate haemorrhagic spots in seromuscularis to generalized dusky areas of impending perforation. In 2 cases ileum was also involved along with jejunum. Histopathological examination of enlarged mesenteric lymph nodes and resected segment of gangrenous gut showed non-specific changes. Simple exploration of abdomen and mesenteric lymph node biopsy was performed in 7 cases, whereas in 2 cases of impending perforation resection and anastomosis had to be done. Culture of peritoneal fluid was inconclusive. Hospital stay was shorter in cases undergoing resection and anastomosis than those having simple exploration. The pathology appears to be a kind of local hyperimmune reaction provoked by endotoxins in the segment of bowel involved. It is the first series of NNJ ever reported in Pakistan. Clinicians are encouraged to be aware of this unusual entity.

Key words: Enteritis, jejunitis, Necrosis, Endotoxins.

Non-specific Necrotizing Jejunitis is a relatively new clinicopathological entity characterized by abdominal pain, fever, bilious vomiting and other features of acute intestinal obstruction¹. The child may have bloody diarrhoea prior to development of constipation. Roentgenography of abdomen in upright posture reveals features of small bowel obstruction. Circumferential intestinal wall inflammation varying from oedema to severe congestion, haemorrhage, necrosis and gangrene with skipped areas of normal bowel are the features of NNJ². Local vasculitis giving rise to thrombosis of micro circulation of bowel leading to ischaemia is the probable pathogenesis of the disease. It tends to follow a self limiting course and can usually be managed by supportive measures³.

Patients And Methods

The study was conducted at Paediatric Surgical unit Mayo Hospital Lahore in the year 1992 where 8 cases were received during 7 weeks of extreme summer. Then there was gap of 5 years and 9th case reported in November 1997 at Services Hospital Lahore. Out of 9 children, there were 7 males and 2 females. Age ranged from 5 to 12 years (Table - I). Detailed history of the disease was taken in all cases and complete physical examination was performed in each patient. Investigations included complete blood examination for Hb%, T.L.C., D.L.C. & ESR, Urine analysis, estimation of serum electrolytes and Widal test. Roentgenography of the abdomen in upright posture was also done in all cases. All the patients were rehydrated and laparotomy performed in each case. Peritoneal fluid was sent for bacteriological culture. Mesenteric lymph nodes and segments of resected intestine were examined histopathologically. In 2 cases anastomosis of jejunum was performed after resection of gangrenous segments of 6 inches and 2 feet respectively. Ampicillin, Metronidazole and aminoglycosides were the antibiotics used.

Results

For the first three cases no definite diagnosis could be made pre-operatively. However in later cases it was made on clinical grounds alone but laparotomy had to be done to confirm the diagnosis and avoid missing any other serious condition like gangrene, perforation etc. About 88% patients presented during June and July whereas only one case (12%) reported in the month of November. All patients belonged to poor socioeconomic class. There were no special dietary habits. The diet consisted mainly of cereals, pulses and vegetables with occasional intake of meat. Duration of symptoms preceding admission ranged from 3 to 7 days. Presenting symptoms of the patients are shown in Table - II. Plain X-rays of abdomen in upright posture showed multiple air fluid levels. Widal test was negative in all cases.

Gross operative findings were (i). Straw coloured peritoneal fluid containing fibrinous flakes ranging in amount from 200cc to 1 litre (ii). Normal mesenteric vascular tree (iii). Inflamed small bowel. Proximal jejunum of variable length was the site of involvement in 7(77.8%) cases whereas in 2 cases (22.2%) entire jejunum alongwith a portion of ileum were involved. The pattern of involvement was the discontinuous areas of different grades of bowel inflammation with normal bowel in between. Pathologically the lesions could be graded as follows:

Grade - 1.

Involved bowel segment was inflamed with areas of punctate haemorrhage and minimal peritoneal fluid. 2 cases.

Grade - 2.

Bowel segment congested and thickened with patchy areas of necrosis and ulceration covered with thick fibrinous material. 5 cases.

Grade - 3.

Frankly gangrenous areas of impending perforation. 2 cases.

Non specific Necrotizing Jejunitis

Histopathologically, areas of bowel segment showed intense congestion and heavy infiltration of acute inflammatory cells in mucosal, submucosal and serosal layers. Focal as well as diffuse areas of necrosis of variable bowel depth were found. Vessels of mucosal and serosal layers revealed fibrinoid eosinophilic thrombi in their lumina. Sections of mesenteric vessels of resected segment showed marked dilatation and congestion with focal inflammatory cell infiltrate. Mesenteric lymph nodes only showed non-specific reactive changes. No granulomas were seen. Culture of peritoneal fluid was sterile in 6 cases whereas mixed growth of bacteria was obtained in 3 cases. The facility of anaerobic culture was not available. There was no mortality. Average hospital stay of children with simple exploration was 18 days whereas it was 10 days in patients undergoing resection and anastomosis.

Discussion

Presentation of NNJ bears similarity to Darmbrand⁴ of Germany, Pigbel⁵ of New Guinea, Ischemic enteritis of Thailand⁶ & Goa⁷, Enteritis necroticans of Uganda⁸. Segmental necrotizing jejunitis is said to be a disease of developing countries⁹. It has also been called Pasini disease¹⁰ and regional jejunitis³. Epidemic and sporadic forms have been reported from India^{1,2,11}.

Clinical manifestations of the disease usually include abdominal pain, vomiting, passage of blood per rectum, abdominal distension and constipation in children 5 - 15 years of age. Varying degrees of shock is also present in majority of patients. Stages of bowel necrosis is the characteristic feature of the disease. No etiological agent has been found but it is thought to be caused by endotoxemia due to gram negative bacilli¹. Intestine may act as portal of entry for endotoxins which later on triggers the hypersensitivity reaction. Vasculitis and obstruction of micro circulation by fibrinous thrombi explains Arthus type of reaction¹. Microscopically, lesions show non-specific acute inflammatory reaction with predominance of neutrophils⁹. Differential diagnosis includes enteritis necroticans, Crohn's disease and Henoch-Schonlein purpura⁹. Enteritis necroticans is a disease caused by clostridium perfringens type C and can be diagnosed by serological studies or bacteriological culture of blood and peritoneal fluid¹². Granulomas of Crohn's disease are absent in NNJ. However Henoch-Schonlein purpura may be difficult to define before the appearance of characteristic rash.

The clinical course of NNJ is not as fulminant as neonatal necrotizing enterocolitis. Most cases can be salvaged by supportive measures like nasogastric aspiration, bowel rest, administration of intravenous fluids and antibiotics. Corticosteroids have also been tried⁶. Surgery should be carried out where complications like perforation or gangrene are suspected or when diagnosis is not confirmed. We had to operate on all patients due to

unawareness of relatively new clinical entity in Pakistan. Second look operation can be performed in patients not improving on conservative treatment after first exploration¹. Resection and anastomosis of localized segments results in early recovery.

Mortality reported by Sharma et al¹, was 32% in patients treated conservatively whereas it was 49.2% in those undergoing surgery. Rai et al³ reported 30%-40% mortality rate in patients who underwent surgical resection, however no mortality in the present study may be due to relatively smaller number of cases in the series. Although recurrence appears to be rare but has been reported by Taufiq et al⁹.

We believe that major surgery can be avoided in most cases of NNJ if the diagnosis is confirmed beforehand. This can be achieved by ultrasonography of thickened bowel loops, endoscopy of jejunum showing typical skip lesions and histological confirmation of jejunal biopsy. Diagnosis can also be confirmed by laparoscopic vision of abdomen which also excludes gangrene and perforation of bowel. However without laparoscopy most of them may end up in laparotomy. It is our intention to generate increased awareness of the entity to other investigators so that more insight may be gained into what at present is an enigma.

References

1. Sharma Ak, Shekhawat N S, Belhari S, Chandra S, Sogani KC: Non-specific jejunitis - A challenging problem in children. *Am J Gastro enterol* 1986; 81: 428-31.
2. Narayanan R, Bhargava B N, Karba SG, Sangal BC: Segmental necrotizing jejunitis [Letter] *Lancet* 1987; 2: 1517-8.
3. Rai AN, Prasad PR, Prasad SN, Tiwari RK. Epidemic regional jejunitis: a new clinical entity [Letter] *Lancet* 1987; 2: 1020.
4. Zeissler J, Rassfeld-Sternberg L. Enteritis necroticans due to *Clostridium welchii*- type F. *Br Med J* 1949; 1:267.
5. Murrell TGC, Roth L, Egerton J et al. Pigbel, Enteritis necroticans. *Lancet* 1966; 1:217-22.
6. Welch TP, Sumitswan S. Acute segmental ischaemic enteritis in Thailand. *Br J Surg* 1975; 62: 716-19.
7. Gupta SD, D'Silva V, Salelkar Av, Reys M. Acute ischemic enteritis in Goa. *Br J Surg* 1977, 64: 420-3.
8. Wright DH, Stanfield JP. Enteritis necroticans in Uganda. *J Pediatr* 1967; 71: 264-68.
9. Taufiq S, Hernanz-Schulman M, Wheeler AV, Morgan WM, Teague MD, Polk DB. Recurrent necrotizing jejunitis in a child. *J pediatr* 1996; 128 (2): 246-9.
10. Knezevic S. Pasini's regional jejunitis [Letter] *Lancet* 1987; 2: 1327.
11. Puyari BD, Deodhare SG. Necrotizing enteritis. *Br J Surg* 1980; 67: 254-6.
12. Clarke LE, Diekmann-Guiroy B, Mc Namee W, Java DJJ, Weiss SM. Enteritis necroticans with mid gut necrosis caused by *Clostridium perfringens*. *Arch Surg*. 1994; 129 (5): 557-60.