Ileo-ileal Knotting: A Ticking Bomb

Ileo-ileal knot is a rare condition which presents like acute abdomen. The treatment is surgical, and should be performed as soon as possible to decrease the incidence of peri-operative mortality and morbidity [1].

We report a 19-month-old boy (weight 9.3 kg, height 80 cm), who presented with multiple episodes of vomiting for few hours. On initial examination, he was irritable with signs of dehydration. Within four hours of admission, the child had multiple episodes of hematemesis. On reviewing the child, he was drowsy with compensated shock. His abdomen was distended, bowel sounds were absent, with guarding and rigidity. Six hours after admission, the hemoglobin reduced to 6.9 g/dL from 9.2 g/dL at admission. Packed red blood cells were transfused. *X*-ray abdomen showed multiple air fluid levels suggestive of intestinal obstruction. Computed tomography (CT) of the abdomen revealed dilated small bowel loops with intramural air consistent with early small bowel ischemia. His Pediatric Risk of Mortality (PRISM) score was 19.

An emergency exploratory laparotomy revealed ileo-ileal knotting; the entrapped loop of ileum was gangrenous, approximately 50 cm in length, extending up to one foot proximal to ileocecal junction. Resection of the gangrenous segment of ileum was performed and continuity of gut restored by end-to-end anastomosis. Immediate postoperative period was uneventful.

On sixth postoperative day, after starting feeds, child had a whitish collection in the peritoneal drain. Biochemical analysis of the fluid was suggestive of chylous ascites; serum amylase (264 IU/L), serum lipase (365 IU/L) and drain fluid triglycerides (277 mg/dL). Child was started on octreotide infusion and fat free diet. Abdominal girth and serial ultrasound monitoring did not reveal any collection. Octreotide infusion was gradually tapered. Child was discharged on seventeenth postoperative day. There no gastrointestinal symptoms or complications after 9 months of follow-up.

In ileo-ileal knotting, one loop of the ileum remains static around which another loop encircles to form a knot [2]. Because of the rarity of the entity, there is no data on age and sex predilection [3]. Ileal knotting caused due to appendix [3] or Meckel diverticulum also has been reported. Once the knot is formed, it sets off a vicious cycle of intestinal occlusion and ischemia due to continuous peristalsis and vascular pulsations, all leading towards to gangrene. When all segments are viable, untying the knot may be enough, since recurrence is uncommon. When irreversible ischemia is present, needle or controlled enterotomy decompression should be done prior to en bloc resection of the congested segments. Manipulation of the knot with intention of untying is not recommended, because of a high risk of perforation. Once the necrotic ileum is extirpated, a primary end to end anastomosis of the small bowel should be done if the distal ileum is not affected. On the other hand, if the remaining segment is closer than 10 cm to the ileocecal valve, end to side ileocolic anastomosis is preferred. The cause of ileo-ileal knotting is not known. Factors such as freely mobile small intestine and redundant sigmoid colon with a long and narrow mesentery have been implicated in ileo-sigmoid knotting [3,4]. The mortality rate is approximately 50%. Treatment should be started as early as possible with aggressive IV fluid resuscitation, insertion of nasogastric tube, broad spec-trum IV antibiotics. Once the patient is adequately resuscitated, emergency laparotomy should be performed. Though, ileoileal knotting is rare, it should be considered in the differential diagnosis of patients with signs and symptoms of small bowel obstruction.

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