Case Report

Gastrointestinal perforations in neonates with anorectal malformations

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We describe the presentation and management of gastrointestinal perforation in four neonates with anorectal malformations. Two neonates with high malformation had pneumoperitoneum on X-ray; surgery revealed sigmoid perforation in one patient and transverse colon perforation in the other. Colostomy was done, followed by posterior sagittal anorectoplasty at four months; both recovered satisfactorily. The third neonate had no radiological feature of gut perforation but cecal perforation was found at surgery; the neonate recovered following right hemicolectomy with stoma followed by anorectoplasty at five months. The fourth neonate presented with clinical and radiological features of perforation and recovered satisfactorily after anoplasty and colostomy. [Indian J Gastroenterol 2004;23:107-108]

Key words: Anorectal anomaly, neonatal peritonitis

Gastrointestinal perforation in neonates with anorectal malformations (ARM) is extremely uncommon. To the best of our knowledge, only five such cases have been reported in English literature till date.1-4 The clinical features and management protocol of this entity has rarely been described in Indian literature.4 Delayed patient presentation is an important factor that demands special attention, especially in developing countries like India.1 We report four such cases, to highlight the role of early diagnosis, aggressive resuscitation and early surgical intervention in the management of such high-risk neonates.

Case Reports

Case 1: A 2.25 Kg 3-day-old full-term male was admitted with abdominal distension and absent anal opening. There was no history of meconiumuria. On examination, the neonate was dehydrated, with distended, soft, silent abdomen. There was no edema or erythema of the abdominal wall. Perineal inspection revealed absent anal opening with well-developed natal cleft and normal external genitalia. There was no evidence of meconium at the anal site or the penile tip. Abdomen X-ray and inverteogram suggested large bowel obstruction with high ARM and free intraperitoneal air (Fig). Investigations revealed leukopenia with band forms and normal renal function tests.

After preoperative resuscitation (nasogastric aspiration, IV fluids, antibiotics and blood transfusion) exploratory laparotomy was done. This revealed free intraperitoneal air with approximately 20 mL of meconium in the pelvis and 5-mm perforation in the distal sigmoid colon. The rest of the peritoneal cavity was closed in a single layer. A proximal high sigmoid divided colostomy was done after perineal toilet. The postoperative period was uneventful. Radiological evaluation suggested intermediate anorectal anomaly without evidence of rectourinary fistula or any cardiac, spinal or urinary tract anomalies. Posterior sagittal anorectoplasty (PSARP) at the age of four months followed by colostomy closure two months later resulted in satisfactory recovery. The patient is continent and doing well on follow up for the last six months.

Case 2: A 4-day-old full-term male neonate weighing 2 Kg was admitted with respiratory distress, abdominal distension and absent anal opening. He appeared tachypneic, septic and dehydrated on examination. Abdominal examination revealed a tense, distended abdomen with edema and erythema of the perianal wall. Perineal examination revealed absent anal opening with normal external genitalia and no evidence of meconium at the anal site or the penile tip. Abdomen X-ray and inverteogram suggested free intraperitoneal air, large bowel obstruction and high ARM. Investigations showed leukopenia with band forms, metabolic acidosis, hyperbilirubinemia, positive C-reactive protein, and pre-renal azotemia.

Along with preoperative resuscitation (IV fluids, inotropic support, blood and plasma transfusion), tube laparostomy was done to drain intraperitoneal air. Laparotomy was done after 48 hours. This revealed approximately 40 mL of meconium in the peritoneal cavity with perforation in the mid transverse colon. The perforation was exteriorized as loop transverse colostomy after peritoneal lavage. In the postoperative period the patient

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required phototherapy but recovery was satisfactory. PSARP at the age of 4 months followed by colostomy closure after 1½ months resulted in satisfactory recovery; the patient is doing well on follow up for the last 2 months.

Case 3: A 2-day-old male neonate presented with absent anal opening and abdominal distension. There was no history of meconiumitis. On examination, the patient was dehydrated, with soft, distended, non tender abdomen. There was no peritoneal wall edema or erythema. Peritoneal examination showed absent anal opening. Abdomen X-ray and invertogram suggested high ARM and large bowel obstruction. Hemogram and renal function tests were normal.

After preoperative resuscitation the patient was taken up for colostomy. On opening the peritoneal cavity there was escape of free air, which suggested presence of bowel perforation. Further exploration revealed approximately 5 ml of meconium in the right paracolic gutter with 2 mm perforation in the anterior wall of the cecum. Right hemicolecotomy with end ileostomy and distal mucous fistula resulted in uneventful recovery. PSARP at the age of 5 months followed by ileostomy closure 1 month later resulted in satisfactory recovery. The patient is on regular follow up for the last 3 months and is doing well.

Case 4: A 4-day-old male neonate presented with abdominal distension, failure to pass meconium, and absent anal opening. There was no history of meconiumitis. On examination the patient was dehydrated, febrile and tachypneic. Abdominal examination revealed distended soft abdomen with mild abdominal wall edema and erythema in the left iliac fossa. There was no peritoneal wall edema and bowel sounds were sluggish. There was no other congenital anomaly. Peritoneal lavage showed covered anus without anocutaneous fistula. The external genitalia were normal. Investigations revealed leukopenia with band forms and positive C-reactive protein. Abdomen X-ray and invertogram suggested large bowel obstruction and low ARM without evidence of free intraperitoneal air.

After resuscitation, anoplasty was performed. The patient had poor bowel function in the postoperative period and features of sepsis were observed. On the second postoperative day abdominal examination revealed persistent abdominal distension with increased abdominal wall edema and peritoneal wall edema. Repeat abdomen X-ray suggested pneumoperitoneum. Laparotomy revealed a 0.5 cm perforation in the sigmoid colon, which was exteriorized as sigmoid colostomy after peritoneal lavage. The postoperative period was uneventful. Anal dilatations were continued and colostomy was closed after 2 months. The child is doing well on regular follow up for the last 1 month.

Discussion
Gastrointestinal perforation is a rare clinical entity in neonates with ARM. Lack of awareness makes early diagnosis and timely surgical intervention difficult, thereby resulting in high morbidity and mortality.

The etiopathogenesis of GI perforation in neonates with ARM is explained by a combination of factors. The downstream occlusion results in proximal intestinal dilatation and increase in intraluminal pressure, resulting in tension gangrene. In the presence of various factors like delayed presentation and surgical intervention and absence of rectourethral fistula, the gangrene progresses to GI perforation, with the large bowel being the most common site. The non-visible intestine resulting from tension gangrene may undergo perforation even when the closed loop obstruction has been relieved, emphasizing the role of close clinical observation of such cases in the postoperative period.

A high index of suspicion in neonates with ARM presenting with sepsis and features of neonatal peritonitis like tense, distended, silent abdomen with parietal wall edema and erythema provide a clue to the diagnosis. Although features of pneumoperitoneum on abdomen X-ray have been reported in 60%-70% of neonates with GI perforation, its presence is confirmatory. Contrast studies may sometimes be hazardous and are not routinely recommended in such sick neonates.

The management of GI perforation in neonates with ARM aims at aggressive resuscitation and early surgical intervention. Preoperative stabilization improves the survival rate in such high-risk cases. As evidenced in one of the present cases, preoperative drainage of intraperitoneal air by drain laparostomy is useful especially in unstable neonates or those with severe respiratory distress. The type of surgical intervention depends upon the physiological status of the patient, the site of perforation, the type of anorectal anomaly, and the degree of peritoneal contamination. Although primary closure of perforation may be attempted in selected cases, exteriorization of the perforation as stoma or its primary closure with a proximal diverting stoma remains the treatment of choice.

References

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