Case report

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Acquired Ileal Atresia in a Preterm Infant

Hyun Ju Lee, M.D., Jae Yun Na, M.D., Tae Kyung Ha, M.D.*, Hyun-kyung Park, M.D., and Chang-ryul Kim M.D.

Departments of Pediatrics and General Surgery*, College of Medicine, Hanyang University, Seoul, Korea

Acquired ileal atresia is a rare but life-threatening complication that can occur after recovery from necrotizing enterocolitis in premature infants. We present an unusual case of acquired ileal atresia in the distal ileum injured by ischemia after intestinal perforation in an extremely low-birth-weight infant. A 900 g (25 weeks gestation) premature infant developed a pneumoperitoneum without the radiologic manifestations of necrotizing enterocolitis on day 9 of life. Primary peritoneal drainage without further need for surgery was performed in the neonatal intensive care facility. Gastrointestinal gastrografen studies confirmed normal intestinal continuity and regular stools. Several weeks later, while the patient was receiving all nutrition orally with no medical problems, the patient's condition suddenly deteriorated, along with clinical signs of intestinal obstruction including emesis of bilious contents and stools. Laparotomy (on day 45 of life) revealed ileal atresia with V-shaped gap mesenteric defect.

Key Words: Acquired ileal atresia, Preterm infants, Intestinal perforation

Introduction

Vascular compromise in early gestation may be responsible for congenital bowel atresia in the jejunum, ileum, and colon. Acquired atresia of the bowel in preterm infants is a severe late complication that arises as a consequence of cicatricial healing of intestine injured by ischemia¹⁾. While partial intestinal obstruction of the colon due to stricture is well-documented as a late complication after resolution of acute necrotizing enterocolitis, ileal atresia is a rare consequence of granulation and fibrosis by the ischemic bowel injury. Atresia refers to the complete obliteration of the bowel lumen, and should be distinguished from stenosis, which refers to luminal narrowing. Acquired intestinal atresia has been reported to occur in premature infants after they recover from necrotizing enterocolitis, and the segment of intestine most often affected is the colon, especially the portion lying distal to

the intestinal stoma^{2,3)}. We present an unusual case of acquired ileal atresia (AIA) after intestinal perforation in the distal ileum injured by ischemia in an extremely low-birth-weight infant.

Case report

A female infant of gestational age 25 weeks and weight 900 g was born to a 33-year-old mother by cesarean section. The pregnancy and delivery were uneventful. There was no evidence of intra-uterine ischemic insult including placental abruption, preeclampsia, intrauterine growth restriction, chorioamnionitis and placental infarction. Apgar scores were 2 and 5, at 1 min and 5 min, respectively. The patient was hemodynamically stable and did not receive inotropic support but did receive artificial surfactant replacement therapy for mild respiratory distress syndrome. Antibiotic therapy with ampicillin and gentamicin was

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Correspondence to : Hyun-Kyung Park, M.D.

Department of Pediatrics, College of Medicine, Hanyang University, 17 Haengdang-dong, Seongdong-gu, Seoul 133-792, Korea

Tel: +82-2-2290-8391, Fax: +82-2-2290-8300, E-mail: neopark@hanyang.ac.kr

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started empirically for 3 days. The umbilical artery catheter was routinely removed at day 7 to prevent vascular insufficiency in the lower extremities, or thrombosis, and total parenteral nutrition was continued through a percutaneous central venous catheter inserted in the right cephalic vein. Oral enteral feeding was slowly advanced to 2 mL every 3 hours without abdominal distension, and bowel gas pattern was normal in abdominal radiographs by day 8.

On day 9, gastric residuals were noted and recurrent apnea developed. Laboratory findings, including complete blood count, serum electrolytes, and liver function, were normal, except for mildly elevated C-reactive protein (CRP; 0.7 mg/dL). The patient developed a pneumoperitoneum without prodromal clinical symptoms and radiographic findings of necrotizing enterocolitis (NEC) (Fig. 1). We decided to use primary peritoneal drainage (PPD) as the treatment of choice without further need for surgery because of a suspected focal intestinal perforation in the absence of NEC. PPD was performed in the neonatal intensive care facility by inserting a Penrose drain on left side of the lower quadrants of the abdomen to evacuate air and fecal contents. The drain was removed after 7 days. Gastrointestinal gastrograffin studies

confirmed normal intestinal continuity and regular stools (Fig. 2). The patient's condition stabilized and oral feeding was advanced to full enteral feeding (130 mL/kg), with no abdominal complications.

On the 42nd day of life (33rd postoperative day), while receiving all nutrients orally with no medical problems, the patient suddenly suffered abdominal distension and elevated pregavage residuals. Because of this sudden deterioration, along with clinical signs of intestinal obstruction including emesis of bilious contents and stools, we performed an exploratory laparotomy on day 45 of life. An abdominal radiograph revealed total absence of bowel gas in the small bowel, colon, and rectum (Fig. 3), and a complete blood cell count gave a white blood cell (WBC) count of $9,500/\text{mm}^3$ (neutrophils, 25%; lymphocytes, 45%; and monocytes, 20%), a hemoglobin level of 11.4 g/dL, and a platelet count of $230,000/\text{mm}^3$. CRP had increased to 2.7 mg/dL. Recurrent apnea and concomitant bradycardia necessitated mechanical ventilation. During surgery (on day 45 of life) 10 cm of the terminal ileum appeared scarred and contracted, and laparotomy revealed dense adhesions and ileal atresia with a V-shaped mesenteric gap (Fig. 4). Histopathologic examination

Fig. 1. Pneumoperitoneum indicated by the presence of free air. Arrows indicate the football sign for free gas in the peritoneal cavity in the absence of pneumatosis intestinalis.

Fig. 2. Gastrointestinal gastrograffin study suggesting normal intestinal continuity.

Fig. 3. Supine abdominal radiograph showing paucity of small bowel gas and gas-filled stomach.

Fig. 4. Ileal segment displaying atresia with a V-shaped mesenteric gap.

showed chronic inflammation consistent with an ileal atresia. No necrosis was detected. Resection of the involved ileum and an ileostomy were performed. The postoperative course was uneventful and favorable, and the patient was discharged from hospital on day 101. Weight at that time was 2,350 g (10th percentile).

Discussion

Although advances in neonatal intensive care have improved the survival of extremely low-birth-weight (ELBW) infants, mortality resulting from intestinal injury has not declined over the past decade^{4,5}. Very small and ill preterm infants are particularly susceptible to enterocolitis resulting from inflammation and ischemia of the colonic mucosa. Kosloske et al.³ reported that six of 31 surviving infants (19%) developed late ischemic stricture of the colon after resolution of acute NEC. However, AIA is rare in premature infants, and only one report has been published to date⁶. Both describe features of intestinal atresia after a NEC episode. Intestinal stricture is part of the reparative process and occurs after an acute decrease in flow through the intramural blood vessels. Although strictures can occur after both medical and surgical treatment for NEC, and in both functional and dysfunctional bowels, the segment of intestine most often affected after small bowel resection and formation of a diverted ileostomy is the colon, especially the portion lying distal to the intestinal stoma.

Reports of non-NEC-related AIA in preterm infants are also limited. In our case, the infant did not meet the classic criteria of NEC, although a histological study was not performed to confirm this. Before the perforation occurred, the patient did not exhibit prodromal clinical symptoms or radiographic findings associated with NEC, such as evidence of abnormal bowel loops, thickened small bowel walls, or pneumatosis intestinalis, and the patient's general condition improved immediately after peritoneal drainage. Unfortunately, no bowel tissue was available for histological comparisons, and the differential diagnosis of NEC and focal intestinal perforation was presumed by plain abdominal radiography and laboratory analysis. Focal intestinal perforation is an emerging disease that lacks the classic features of NEC. Kubota et al.⁷ proposed that use of umbilical catheters, administration of indomethacin or steroids, and congenital defects of the intestinal wall can contribute to ischemic changes of the intestine and, subsequently, intestinal perforation in extremely low-birth-weight infant. Glotzer et al.⁸ demonstrated that if a circulatory disturbance occurs in the intestine, the mucous membrane is damaged first, and, if the condition

persists, perforation can follow. We postulate that in our case the umbilical artery catheter hindered the intestinal circulation, with resultant mesenteric ischemia and loss of intestinal integrity leading to intestinal perforation. In the presence of moderate ischemia and collateral flow, restoration of the blood supply allows healing of this segment, with subsequent cicatrix formation and resorption of the devitalized segment. Depending on the extent of intestinal ischemia or gangrene in premature infants, partial occlusion and atresia of the bowel can occur after recovery from mesenteric ischemia. Severe strictures have been characterized by replacement of the submucosa and muscularis by granulation tissue and fibrosis, resulting in obliteration of the lumen³⁾. Ratan et al.⁹⁾ described two patients, aged 3 months and 4 months, in whom episodes of diarrhea caused unusual acute development of atresias in the distal ilea after brief symptomatology of approximately 14 days. The authors proposed that a thromboembolic event depending on the collateral blood supply to that segment may underlie intestinal stenosis or atresia presenting a few weeks after birth. We speculate that chronic inflammation with subsequent healing and the decreased intestinal circulation caused by severe adhesions may have contributed to the slow progress of the atresia in the present case. Histologic finding of chronic inflammation without necrosis also supports this speculation. Furthermore, the acquired atresia in the present patient may have progressed slowly because the passage of intestinal content through the damaged focal intestinal segment protected the luminal patency from completely interrupting the continuity of the intestine.

In general, the outcomes for patients with focal intestinal perforation are more favorable than for those with NEC. In some centers, PPD without further surgery is the preferred treatment for intestinal perforation without clinical evidence of NEC. However, the efficacy of peritoneal drainage rather than laparotomy as the definitive treatment for ELBW infants with NEC or intestinal perforation remains to be established. Some studies have demonstrated that although PPD is useful for rescuing and stabilizing perforated NECs, most infants ultimately require laparotomy^{10,11)}.

AIA is a rare, but life-threatening, complication that is almost unreported. Recognition of the late sequelae of intestinal per-

foration in preterm infants is becoming critical for successful outcome for these patients as survival rates increase. In patients with focal intestinal perforations who are otherwise healthy after peritoneal drainage but later develop problems with feeding, AIA should be considered as a severe complication and further examination performed. Early surgical management can rapidly return such patients to a normal state and prevent prolonged morbidity.

한글요약

미국아에서 후천성 회장 폐쇄는 드물게 괴사성 장염과 연관하여, 특히 대장에 발생하는 매우 위험한 후기 합병증이다. 이는 손상된 허혈성 장관 조직의 회복과정 중 반흔모양의 형태로 변한 장관의 완전 폐쇄를 초래하는 것으로 단순 협착과 달리, 수술을 요하는 응급상황의 심각한 임상 악화를 초래할 수 있다. 저자들은 초극소 저체중 출생아에서 괴사성 장염의 임상적 증거 없이 초래된 장관공을 일차적 복강 배액술로 완치하고 몇 주 후에 발생한 후천성 회장 폐쇄증 1례를 발견하였고 성공적으로 치료하였기에 이를 보고하고자 한다.

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