Multiple Intussusceptions in an Extremely Premature Infant

Ha-Su Kim, M.D.¹, Hyun-A Kim, M.D.¹, Sung-Heun Kim, M.D.², Shin-Yun Byun, M.D.³, and Myo-Jing Kim, M.D.¹

Departments of Pediatrics¹ and Surgery², Dong-A University, College of Medicine, Busan, Korea
Department of Pediatrics³, Pusan National University School of Medicine, Yangsan, Korea

Intussusception in premature infants is very rare. Here, we report a case of multiple intussusceptions in an extremely preterm infant, born at 23.5 weeks gestation, who underwent an exploratory laparotomy, for bowel perforation and misdiagnosed necrotizing enterocolitis, at 20 days of life. To our knowledge, this is the most prematurely born baby that has survived with multiple intussusceptions.

Key Words: Intussusception, Extremely premature infant

Intussusception is highly uncommon during the neonatal period and is exceedingly rare in premature infants. The rarity of the condition, and the difficulty differentiating it from a common neonatal disease such as necrotizing enterocolitis (NEC), often delays diagnosis and surgical intervention, which contributes to increased morbidity and mortality.¹,² In this study, we present a case of multiple intussusceptions in an extremely premature infant, born in gestational week 23.5, who was misdiagnosed with NEC. To our knowledge, this is the most premature case that has survived with multiple intussusceptions.

Case report

A female infant weighing 640 g was born at gestational week 23.5 by cesarean section to a 33-year-old mother, who had presented with 33 hours of premature rupture of membranes and confirmed histologic chorioamnionitis. The infant’s Apgar score was 1 at one minute and 4 at five minutes. She had many problems associated with prematurity, including respiratory distress syndrome requiring high frequency ventilator and surfactant replacement therapy, refractory hypotension requiring inotropics and hydrocortisone, thrombocytopenia, suspected sepsis, and patent ductus arteriosus requiring surgical ligation after failed ibuprofen treatment at 8 days of life. At 12 days of life, she exhibited bilious gastric aspirates, and plain radiography showed a gasless abdomen for two days. Abdominal ultrasonography was not performed. Enteral feeding had not started until that time and there was no hematochezia. Laboratory results showed leukocytosis: white blood cell count (WBC) 25,050 (segmented neutrophil 74%)/mm³, hemoglobin 11 g/dL, platelet 161,000/mm³, negative C-reactive protein, pH 7.363, base excess −3.2 mEq/L, bicarbonate 20.9 mEq/L, negative blood culture. NEC was initially suspected, but abdominal distension was minimal, and no pneumatosis intestinalis was found on subsequent
Radiography. Vancomycin and meropenem were treatment. On day 20, we performed an abdominal radiograph due to marked abdominal distension and desaturation, which demonstrated free abdominal air (Fig. 1). Laboratory results were not specific: WBC 19,670 (segmented neutrophil 69%)/mm$^3$, hemoglobin 9.6 g/dL, platelet 144,000/mm$^3$, negative C-reactive protein, pH 7.234, base excess −1.5 mEq/L, bicarbonate 26.3 mEq/L. Our diagnosis was NEC with perforation. A laparotomy was performed immediately. The intraoperative findings were ileo-ileal intussusception with small bowel perforation, requiring ileostomy, approximately 20 cm proximal to the ileocecal valve. The intestinal wall displayed moderate congestion, and two additional intussusceptions, which were successfully reduced manually, at 20 cm distal from Treitz ligament (Fig. 2). No lead point was detected. Upon examination of the gastrointestinal tube, no abnormality was observed. Pathologic evaluation of the specimen revealed inflammation with congestion at the site of intussusceptions. The postoperative course was uneventful. The enteral feeding was started 7 days after the operation, and full enteral feeding was performed at 42 days of life. Repair of the ileostomy was performed successfully with primary end-to-end anastomosis at 5 months of life. The child was discharged in excellent general condition.

**Discussion**

Intussusception occurs very infrequently in the neonatal period, with a reported incidence ranging from 0.3% to 2.7% in the first month of life, and results in less than 3% of all neonatal bowel obstructions. Small bowel intussusceptions occur in less than 10% of all age groups, yet they are more common up to 68% in premature neonates, particularly in the ileum. In Korea, there were three postnatal intussusceptions in prematurity were reported previously. All...
of the patients (25 weeks, 27 weeks, and 28 weeks) were diagnosed to NEC with perforation at first time, but after laparotomy (at 39 days, at 25 days, and at 33 days), they were confirmed to intussusception same as this case.\textsuperscript{6,8} Diagnosis time was slight late compared to this case (at 20 days). Just one case\textsuperscript{6} was performed abdominal ultrasonography, intussusception was not detected by sonography. Two of them\textsuperscript{6,8} were ileo-ileo-colic intussusceptions and one case had no information about that. The etiology of neonatal intussusceptions in premature infants remains unclear. As in the present case, intestinal hypoperfusion or ischemia would result in dysmotility and stasis, but accelerated peristalsis would occur in the early or recovering phase of the event, potentially acting as a functional leading point.\textsuperscript{9}

These clinicopathological characteristics may be responsible for diagnostic confusion with other conditions, particularly with NEC, which is relatively more common in these patients. The mean time from onset of signs and symptoms to surgical intervention was 9.9±11.8 days in reported cases of intussusceptions in neonates.\textsuperscript{9,10} Indeed, Wang et al.\textsuperscript{10} reported that intussusceptions must be highly suspected in a neonate who is diagnosed with NEC, but who has a more stable course than would be expected. Avansino et al.\textsuperscript{11} mentioned that the most common imaging finding in premature neonates with intussusceptions is dilated bowel loops. Similarly, the abdominal radiograph of the patient presented in this study revealed small intestine dilated loops. Pneumatosis intestinalis or portal venous gas was not found. Recent studies reported that abdominal ultrasound was helpful for rapid and accurate diagnosis of intussusceptions, as the clinical picture was not explained by the initially suspected NEC.\textsuperscript{2,9} But this case, we did not performed abdominal ultrasonography due to no doubt to intussusception. The underlying condition was too severe to allow surgery in the patient, but prompt laparotomy following diagnosis is crucial for achieving better outcomes.\textsuperscript{1}

Intussusception in neonates is an extremely rare clinical entity and can often be confused with other causes of intestinal obstruction and intestinal distension. There are neither definite risk factors nor clinical or diagnostic characteristics to definitively differentiate between intussusceptions and NEC in preterm infants. A high degree of suspicion is needed to avoid misdiagnosis.

The authors have no conflicts of interest relevant to this article.

References

초극소 미숙아에서 발생한 다발성 장중첩증

장중첩증은 미숙아에서 매우 드문 질환이다. 본 저자들은 임신 나이 23주 1일로 출생한 초극소 미숙아에서 생후 20일에 괴사성 장염과 관련된 장천공이 의심되어 시험적 개복술을 시행하였으나 장중첩증으로 최종 진단된 증례를 경험하였기에 보고하고자 한다. 본 증례는 다발성 장중첩증으로 진단되어 수술 후 생존한 가장 어린 미숙아로 증례 보고의 의의가 있다.

중심 단어 : 장중첩증, 초극소 미숙아