

Case Report

Rare gastrointestinal perforations in the neonatal period: Meckel diverticula and acute appendicitis; Two case reports

Yenidoğan döneminde nadir görülen gastrointestinal perforasyonlar: Meckel divertikülü ve akut apandisit; İki Olgu Sunumu

Ezgi Yangın Ergon¹, Rüya Colak¹, Ferit Kulalı¹, Senem Alkan Özdemir¹, Meral Yıldız¹, Aytaç Karkıner², Malik Ergin³, Tülin Gökmen Yıldırım¹, Şebnem Çalkavur¹

¹İzmir Dr Behçet Uz Children's Disease And Surgery Education Research Hospital, Neonatology, İzmir, Türkiye ²İzmir Dr Behçet Uz Children's Disease And Surgery Education Research Hospital, Pediatric Surgery, İzmir, Türkiye ³İzmir Dr Behçet Uz Children's Disease And Surgery Education Research Hospital, Pathology, İzmir, Türkiye

ABSTRACT

Neonatal gastrointestinal perforation (GIP) is a pathology that can be observed in a highly heterogeneous group ranging from premature and low birth weight babies to healthy term babies. Here, we report two rare cases of GIP in the neonatal period with different pathologies. Our first case presented with spontaneous intestinal perforation in the first week of life is the perforated Meckel's diverticulum. The second case presented with intestinal abscess and perforation is perforated appendicitis. Although there are similar risk factors in the etiology, the GIP pathologies in the neonatal period are very different from each other and their common characteristics are that they present with intestinal perforation in the first weeks of life. The correct diagnosis before the operation is very challenging due to insufficient laboratory and imaging methods. Careful clinical observation and timely surgical intervention protects against morbidity and mortality due to perforation.

Keywords: Meckel's diverticulum, appendicitis, intestinal perforation, necrotizing enterocolitis, newborn

ÖZET

Yenidoğan gastrointestinal perforasyonu (GIP), prematüre ve düşük doğum ağırlıklı bebeklerden sağlıklı term bebeklere kadar oldukça heterojen bir grupta görülebilen bir patolojidir. Burada yenidoğan döneminde farklı patolojilere sahip iki nadir GIP olgusunu sunuyoruz. Yaşamının ilk haftasında spontan intestinal perforasyon ile başvuran ilk olgumuz perfore Meckel divertikülüdür. Bağırsak apsesi ve perforasyonu ile başvuran ikinci olgu perfore apandisit. Etiyolojide benzer risk faktörleri olmasına rağmen yenidoğan dönemindeki GIP patolojileri birbirinden çok farklıdır ve ortak özellikleri yaşamın ilk haftalarında intestinal perforasyon ile ortaya çıkmalarıdır. Sınırlı laboratuvar ve görüntüleme yöntemleri nedeniyle ameliyat öncesi doğru tanı koymak oldukça zordur. Dikkatli klinik gözlem, zamanında cerrahi müdahale, perforasyona bağlı morbidite ve mortaliteye karşı korur.

Keywords: Meckel divertikülü, apandisit, barsak perforasyonu, nekrotizan enterokolit, yenidoğan

Received: 07.11.2022 · Accepted: 29.12.2022 · Published: 30.04.2023

Correspondence / Yazışma: Ezgi Yangın Ergon · İzmir Dr. Behçet Uz Çocuk Hastalıkları ve Cerrahisi Eğitim Araştırma Hastanesi, Yeni Doğan Kliniği, İzmir, Türkiye · yanginezgi@yahoo.com

Cite this article as: Yangın Ergon E, Colak R, Kulalı F, Alkan Özdemir S, Yıldız M, Karkıner A, et al. Rare gastrointestinal perforations in the neonatal period: Meckel diverticula and acute appendicitis; Two case reports. *Pediatr Acad Case Rep.* 2023;2(2):39-43.

INTRODUCTION

Neonatal gastrointestinal perforation (GIP) is a pathology that can be observed in a highly heterogeneous group ranging from premature and low birth weight babies to healthy term babies. In a term infant with GIP, surgical congenital GIS pathologies, such as intestinal obstruction, malrotation, or rarely, Hirschsprung's disease (HD) come into mind first, while in a premature infant, acquired necrotizing enterocolitis (NEC) and spon-

taneous intestinal perforation (SIP) are considered primarily (1). The factors that predispose to neonatal GIP are perinatal hypoxia, congenital absence of gastrointestinal wall muscles, corticosteroid therapy in the antenatal/postnatal period, use of maternal cocaine and exchange transfusion for hemolytic disease (2). Here, we report two rare cases of GIP in the neonatal period with different pathologies.

CASE REPORT

Case report 1

The case that was born at the 30th gestational week (GW) due to preeclampsia and premature rupture of membrane weighing 1530 grams was hospitalized due to prematurity and respiratory distress. Apgar was 5/8 in the delivery room; spontaneous breathing was present but weak. After the first 24-hour-follow-up in nasal continued positive airway pressure, abdominal distension developed on the second day of life. The patient had meconium defecation. The abdominal X-ray was consisted of intestinal perforation with free air in the abdomen (Figure -1). Ampicillin and gentamicin were started as empirical antibiotic on the first day of life due to prematurity and respiratory distress. Also, Metronidazole was added for anaerobic agents on the 2nd day. The patient was diagnosed with SIP and exploratory abdominal surgery was performed. Bilirubin fluid containing bile was observed in the abdomen, a broad-based Meckel's diverticulum (MD) of 20 cm proximal to the ileocecal valve and perforation area was detected on the diverticulum. The abdomen was closed after intestinal resection and end-to-end anastomosis on the 2nd day of life. Antibiotherapy was discontinued on the 7th day of the case without culture growth. Pathology report was consistent with perforated MD (without ectopic tissue) (Figure-2,3). The patient with symptomatic MD in the neonatal period was started on feeding on the 7th postoperation day and discharged as 2050 grams on the 34th day of life with complete recovery. The consent was obtained from the parents of this patient for this case study.



Figure 1. Free air below the diaphragm

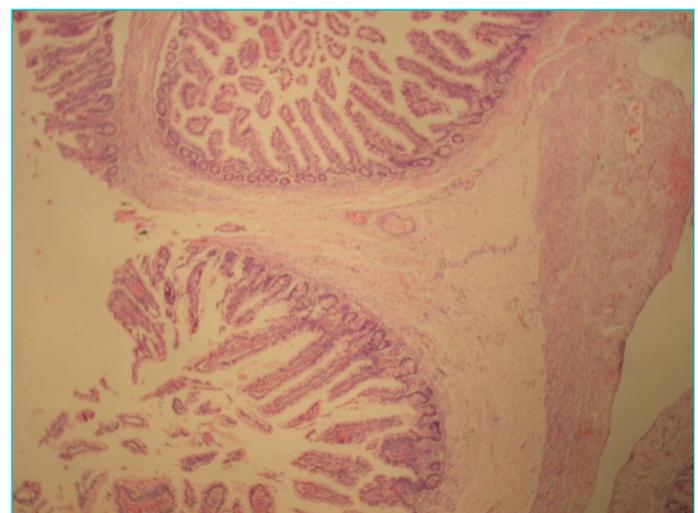


Figure 2. H&E 40x meckel diverticulum

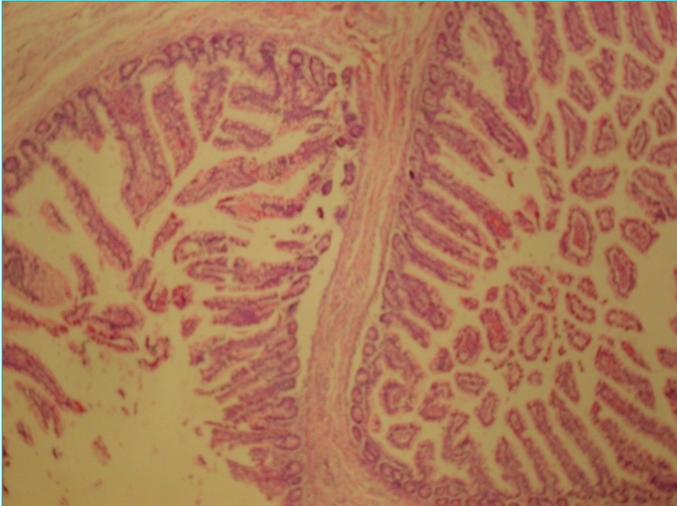


Figure 3. H&E 100 meckel diverticulum

Case report 2

A male infant born at the 40th GW with 3800 grams using cesarean section was hospitalized with the preliminary diagnosis of upper respiratory infection (URI) and late neonatal sepsis on the 5th day of life. In his antenatal follow-up, it was learned that his mother received antibiotic therapy in the last trimester due to a urinary tract infection. The patient case was administered empirical antibiotherapy (ampicillin and gentamicin). However, he had weakness of sucking, an increase in fever, restlessness, and acute phase reactants in the follow-up period. The ampicillin dose was increased due to increased signs of sepsis, but when clinical worsening continued at the 48th hour, empirical antibiotherapy was changed to vancomycin piperacillin-tazobactam. On the 4th day, in addition to sepsis findings, abdominal distention and tenderness developed except for vomiting, and piperacillin tazobactam was changed to meropenem for abdominal findings. No feature was detected in outpatient abdominal radiography. When the increase of echogenicity in mesenteric fat plan in the right lower quadrant, edema in the ileal loops, and fluid loculations between the loops were detected in abdominal ultrasonography (US), metronidazole for anaerobic agents was added to the treatment with the preliminary diagnosis of intraabdominal abscess and intestinal perforation, and pediatric surgery team evaluated the case. Contrast-enhanced abdominal computerized tomography confirmed ultrasonographic findings. An exploratory abdominal surgery showed perforated appendicitis and adhesion on the ileal loops and an appendectomy was performed on the 5th day of the follow-up. Microscopic findings were consistent with macroscopy, and in the specimen taken, ganglion cells

were present in the nerve plexuses; therefore, HD was eliminated (Figure -4). The vancomycin and metronidazole treatments of the case with ESBL (+) *Escherichia coli* growth in the blood culture were discontinued on the 7th day, and the meropenem treatment was completed 14 days after two clean cultures. The patient case with perforation due to neonatal acute appendicitis (NAP) was started on feeding on the 5th postoperation day and was discharged at 4100 grams on the 24th day with a complete recovery. Written informed consent was obtained from the patient's parents, who participated in this case study.



Figure 4. H&E 40 appendicitis

DISCUSSION

Neonatal GIP occurs with different and sometimes sporadic causes. MD is the most common congenital anomaly of the gastrointestinal tract, which may be complicated by approximately 2-4% (3-5). In childhood, MD is most commonly seen after two years of age, with a two-fold increase in males compared to females, with bowel obstruction, gastrointestinal bleeding, acute inflammation and umbilical abnormalities (6-9). MD is caused by incomplete obliteration of the omphalomesenteric or vitelline sac that occurs at about the 5th week of pregnancy, it is a true diverticulum containing all layers of the normal bowel wall (10). There are heterotropic tissues in 60% of the MD and 60% of them have gastric tissues; therefore, in the presence of symptoms, acid secretion and painless GIS bleeding due to ulceration are the most commonly seen (7).

Intestinal obstruction and ileal volvulus due to inflammation are most common in the neonatal period compared to all other age groups, whereas symptomatic MD is rare in this period (11, 12). Symptomatic neonatal MD often occurs with bowel obstruction. MD-re-

lated perforation is, on the other hand, a rare condition. MD, perforated in the first two weeks of life, has been reported in the literature, with only 13 cases in the last 30 years (13). These cases are usually premature and they have been reported to be four times more frequent in male cases than in female cases (13, 14). MD perforation may be caused by inflammation, ulceration, congenital muscle defect, or the development of secondary perforation of HD, or it can be seen spontaneously (2, 14). Chang et al. described the first spontaneous perforated neonatal MD case with progressive abdominal distention and pneumoperitoneum at the 29th hour of life, where MD perforation did not cause peritonitis, meconium did not contaminate the abdominal cavity (14). Although not containing inflammation or ectopic mucosa in its pathology, our patient is in the second case in the literature as neonatal MD with spontaneous perforation at a very early age.

Spontaneous intestinal perforation (SIP), in other words, isolated-focal bowel perforation, is a rare neonatal GIP (15) in which the adjacent intestinal tissues are intact. It is seen in the rate of 3-8% in infants whose birth weights are below 1000 grams during the first weeks of life (15). While focal ischemic necrosis area was detected in the pathology, due to the absence or thinning of the muscularis propria in the perforation area in the pathogenesis, the perforation of the intestine is demonstrated as the cause since it could not tolerate intramural pressure with the increase of bowel movements around the 7th day of life (16). In the pathogenesis of neonatal MD, the fact that congenital muscle defect is stated as the reason and similar risk factors trigger the perforation raises the question of "Is spontaneous perforated neonatal MD a rare form of SIP?" An increase in the cases reported in the literature will be useful to clarify this situation.

Acute appendicitis is common in children and adults, whereas acute appendicitis in the neonatal period is rare. There have been about 50 cases reported in the literature in the last 30 years (17). The rarity of AP in the neonatal period is associated with the presence of fluid diet, supine position, lower rate of GIS infections due to breast milk, and large appendix base with rare lymphatic hyperplasia (18, 19). There are three theories of NAP etiology; "Is it an isolated form of NEC in the first two weeks of life? Is it caused by obstructive cecal distention due to HD? Is it caused by vascular insufficiency?" (20, 21). Karaman et al. retrospectively analysed 121 NAP cases and stated that 50% of the cases were preterm and 75% of the cases were male. In the preterm

cases NAP was accompanied by comorbidities, such as HD, cystic fibrosis and inguinal hernia. On the contrary, in the term patients comorbidities were rare (18). Since the beginning of the 1900s, the mortality rate of NAP has decreased from 78% to 28% both with the increase in the quality of care and the widespread use of antibiotics (18). However, in case of late diagnosis and delayed intervention, one out of the four patients still die. Our patient here is a rare NAP case in the literature, being a term infant with no expected comorbidities but clinical symptoms of URI and late neonatal sepsis.

One premature and one-term neonates with rare causes of GIP in the early weeks of life are presented above. The common features are that they are presented with point intestinal perforation in the first weeks of life. It is very challenging to establish the correct diagnosis before the operation because the laboratory and imaging methods can be inadequate. The important point is that clinical observation should be made carefully in the presence of a neonatal pre-diagnosis of GIP, surgery intervention be performed in a timely and accurate manner, and thereby, the infants be protected from morbidity and mortality due to the late intervention of the perforation.

Patient Consent Form / Hasta Onam Formu

The parents' of this patient consent was obtained for this study.

Conflict of Interest / Çıkar Çatışması

The authors declared no conflicts of interest with respect to authorship and/or publication of the article.

Financial Disclosure / Finansal Destek

The authors received no financial support for the research and/or publication of this article.

REFERENCES

1. Aguayo P, Fraser JD, St Peter SD, et al. Perforated Meckel's diverticulum in a micropremature infant and a review of literature. *Pediatr Surg Int* 2009; 25: 539Y541.
2. Kumar P, Ojha P, Singh UK. Spontaneous perforation of Meckel's diverticulum in a neonate. *Indian Pediatr* 1998; 35: 906Y908.
3. Mackey WC, Dineen P. A fifty year experience with Meckel's diverticulum. *Surg Gynecol Obstet* 1983; 156: 56Y64.
4. Moore TC. Omphalomesenteric duct malformations. *Semin Pediatr Surg* 1996; 5: 116Y123.
5. Shalaby RY, Soliman SM, Fawy M, et al. Laparoscopic management of Meckel's diverticulum in children. *J Pediatr Surg* 2005; 40: 562Y567.
6. Seagram COF, Louch RE, Stephen CA, et al. Meckel's diverticulum-a 10 year review of 218 cases. *Can J Surg* 1968; 11: 369Y373.

7. Weinstien EC, Cain JC, ReMine WH. Meckel's diverticulum: fifty-five years of clinical and surgical experience. *JAMA* 1962; 182: 131Y133.
8. Rutherford RB, Akers DR. Meckel's diverticulum: a review of 148 pediatric patients, with special reference to the pattern of bleeding and to mesodiverticular vascular bands. *Surgery* 1966; 59: 618Y626.
9. Vane DW, West KW, Grosfeld JL. Vitelline ducts anomalies. Experience with 217 childhood cases. *Arch Surg* 1987; 122: 542Y547.
10. Moore KL, Persaud TVM. *The Developing Human. Clinically Oriented Embryology*. Philadelphia, PA:W.B. Saunders Company; 1993: 255Y256.
11. Benson CD. Surgical implications of Meckel's diverticulum. In: Ravicth MM, Welch KJ, Benson CD, eds. *Pediatric Surgery*. Chicago, IL: Year Book Medical Publishers; 1978:955Y960.
12. Soltero MJ, Bill AH. The natural history of Meckel's diverticulum and its relation to incidental removal. *Am J Surg* 1976; 132: 168Y171.
13. Bertozzi M, Melissa B, Radicioni M, et al. Symptomatic Meckel's Diverticulum in newborn. *Pediatric Emergency Care*, 2013; 29:9.
14. Chang Y, Lin J, Huang Y. Spontaneous perforation of Meckel's diverticulum without peritonitis in a newborn: report of a case. *Surg Today* 2006; 36: 1114Y1117.
15. Wadhawan R, Oh W, Hintz SR, et al. Neurodevelopmental outcomes of extremely low birth weight infants with spontaneous intestinal perforation or surgical necrotizing enterocolitis. *J Perinatol* 2014; 34(1):64–70.
16. Gordon P, Rutledge J, Sawin R, et al. Early postnatal dexamethasone increases the risk of focal small bowel perforation in extremely low birth weight infants. *J Perinatol* 1999; 19: 573-77
17. Schwartz KL, Gilad E, Sigalet D, et al. Neonatal acute appendicitis: a proposed algorithm for timely diagnosis. *J Pediatr Surg* 2011; 46: 2060–4.
18. Karaman A, Cavuşoğlu YH, Karaman I, et al. Seven cases of neonatal appendicitis with a review of the English language literature of the last century. *Pediatr Surg Int* 2003; 19:707-9.
19. Lin YL, Lee CH. Appendicitis in infancy. *Pediatr Surg Int* 2003; 19: 1-3.
20. Arliss J, Holgersen LO. Neonatal appendiceal perforation and Hirschsprung's disease. *J Pediatr Surg* 1990; 25:694–5.
21. Van Veenendaal M, Plotz FB, Nikkels PG, et al. Further evidence for an ischemic origin of perforation of the appendix in the neonatal period. *J Pediatr Surg* 2004; 39: e11–12.