CONGENITAL DUODENAL ATRESIA ASSOCIATED WITH INTRAUTERINE INTESTINAL VOLVULUS AND MECONIUM PERITONITIS – A CASE REPORT

Svetlana BUKARICA1,2, Jelena ANTIĆ1,2, Nenad ŽAKULA1, Ivana LUKIĆ1,2 and Marina MILENKOVIĆ1,2

Summary

Introduction. Duodenal atresia is one of the most common small bowel anomalies usually diagnosed in utero. We present a case of a newborn with duodenal atresia associated with intestinal volvulus and meconium peritonitis. Case Report. A premature newborn with a birth weight of 1970 g was admitted due to radiological signs of duodenal atresia. Intraoperatively, in addition to duodenal atresia, we found that almost all intestinal loops were adherent to each other and to the liver in the form of conglomerates, as a consequence of intestinal (jejunal) volvulus and meconium peritonitis. A minimal intestinal resection was performed with creation of duodeno-jejunal anastomosis. The postoperative course was prolonged due to serious complications including cardiac tamponade and sepsis. After 77 days, with full enteral feeding, spontaneous bowel movement, and after appropriate weight gain, the child was discharged from the clinic. Conclusion. Treating congenital anomalies of the newborn is a challenge, even for an experienced surgeon, especially when associated with premature birth and low birth-weight. Despite adequate surgical techniques and intensive treatment, complex congenital anomalies are associated with high morbidity and mortality rates.

Key words: Intestinal Atresia; Duodenal Obstruction; Intestinal Volvulus; Meconium; Peritonitis; Infant, Premature; Congenital Abnormalities; Neonatology; Surgical Procedures, Operative; Prenatal Diagnosis

Introduction

Congenital duodenal atresia is the most common small bowel anomaly and it occurs in 1 in 3,000 - 10,000 live births [1, 2]. This anomaly affects male infants more frequently than females [3]. In theory, it represents an obliteration of the duodenal lumen and it is usually described as a result of failure of recanalization between the 8th and 10th week of gestation. Another possible cause of duodenal atresia is annular pancreas, which occurs as a result of extraluminal duodenal obstruction by the surrounding pancreatic tissue [4, 5]. Congenital duodenal atresia can be observed in newborns with Down syndrome, VACTERL (vertebral defects, anal atresia, cardiac defects, trachea-esophageal fistula, renal anomalies and limb abnormalities) anomalies, intestinal malrotation, biliary tract anomalies, mandibulofacial anomalies, but also as an isolated malformation [2, 6].

In newborns with clinical signs of high intestinal obstruction, midgut volvulus must be considered as well. Although it is known that the incidence of symptomatic intestinal malrotation is 1 in 6,000 births, it is not precisely published how many of these present with neonatal intestinal volvulus [2]. Intrauterine intestinal volvulus represents a pathologic intestinal and/or proximal colon rotation around superior mesenteric artery, with subsequent...
intestinal venous congestion, followed by bowel ischemia and finally necrosis. The most frequent type of intestinal volvulus (also called “classic”) is a result of intestinal malrotation, that is, clockwise rotation of the small bowel and ascending colon around the superior mesenteric artery. Another type of intestinal volvulus, called segmental, occurs when intestinal loops are twisted due to presence of meconium ileus, mesenteric or intestinal cysts, mesenteric defects, intestinal atresia, or it may be idiopathic. The rarest type of intestinal volvulus is without malrotation and malposition, which is usually found in premature and/or low birth weight newborns [7]. Unlike intestinal atresia, this condition is rarely discovered before birth, but must be considered in differential diagnosis of acute abdominal distension in a newborn [2].

In infants with bowel perforation due to intestinal atresia, volvulus, hemochromatosis, cystic fibrosis or viral infection, a condition known as meconium peritonitis may occur. This condition represents a sterile peritoneal inflammation, which usually has poor prognosis, with mortality of 80% [3, 4].

The presence of all the three above-mentioned pathologic conditions is quite rare, and just a few cases have been described in the literature so far [8–10]. They are considered to be a consequence of the regulatory factor X6 gene mutation, and may also be associated with neonatal diabetes, gallbladder agenesis and anomalies of the pancreas. Long-term prognosis in these patients is poor, with high mortality in the first months of life [8–10].

Case Report

We present a preterm newborn in his very first hours of life. He was born at 34 weeks of gestation weighing 1970 g. The intrauterine ultrasound showed polyhydramnios, so intestinal obstruction was suspected.

The infant presented with vomiting and mildly distented meteoristic abdomen in the upper quadrants. A double bubble sign was seen on an abdominal X-ray (Figure 1), while abdominal ultrasound revealed distended stomach with no detectable passage into the small intestine.

After hydro-mineral and acid-base regulation, nasogastric probe was placed, and the patient was prepared for surgery. Under general anesthesia, right transverse supraumbilical laparotomy was made. Intestinal loops adherent to each other and to the liver in the form of conglomerate of intestinal loops were visible immediately after entering the peritoneal cavity (Figure 2). After conglomerates were carefully separated, we found out that they were formed as a consequence of intrauterine intestinal volvulus (Figure 3) associated with meconium peritonitis. Careful intestinal and mesenteric detorsion was performed, bowel loops were wrapped in warm gauze and abdominal cavity was properly cleaned. We initially expected to find a duodenal atresia, with a typically dilated proximal and obliterated distal portion, i.e., type II duodenal atresia (Figure 4). Considering that part of the jejunum (not longer than 10 cm) was ischemic and nonviable, it had to be resected, while anastomosis was created between the duodenum and the rest of the viable jejunal loops. Nasojejunal probe was placed as a stent through the site of anastomosis.

The postoperative recovery included antibiotic therapy, blood transfusions and internal homeostasis maintenance was continued in surgical intensive care.
Two weeks after the surgery, the baby suddenly deteriorated due to a cardiac tamponade. After cardiopulmonary resuscitation, pericardiocentesis was performed and clear fluid evacuated. Six days later, the baby was extubated, with sufficient oxygen saturation. After several weeks, he developed sepsis, which was successfully treated with antibiotic therapy. After 77 days of hospitalization, with full enteral feeding, the patient was discharged. At this moment, he is a one year old boy, still regularly controlled.

**Discussion**

Advances in prenatal diagnosis have facilitated early recognition of congenital anomalies. Prenatal ultrasound differentiates approximately one third of congenital intestinal obstructions. Accounting for 54% of prenatally detected cases, duodenal atresia is the most common anomaly of this type. Ultrasound characteristic of congenital duodenal atresia includes the double bubble sign, which is a presentation of distended stomach and duodenal pouch separated by the pylorus, with or without polyhydramnios [6]. Postnatal diagnosis of duodenal atresia, besides clinical signs and symptoms such as bilious vomiting sometimes with upper abdominal quadrants distension, relies on native abdominal X-ray as well. These findings are generally similar to the prenatal ones [11]. Similar clinical signs and symptoms (vomiting and mildly distended, meteoristic abdomen in its upper quadrants) as well as radiological finding of double bubble sign and distended stomach with no detectable passage into the small intestine, were present in our patient.

Reviewing the literature, we found several articles describing cases where radiological (double bubble sign) and even clinical findings suggested duodenal atresia, but patients have subsequently been found to have midgut volvulus [12, 13]. That is why in a newborn with acute abdominal distension, intestinal volvulus must be suspected as one of the possible etiologic causes. In patients with midgut volvulus, prenatal ultrasonography may detect dilated bowel loops, as well as polyhydramnios. Postnatal radiological findings include lack of intestinal air in the right abdomen, with all the loops displaced to the left [14]. In cases where consequent intestinal ischemia occurs, distended, "tubularized" bowel loops can be seen, sometimes with digitiform impressions as a result of wall edema [15]. As a result of intestinal volvulus, meconium peritonitis can also be seen. This condition, found in our newborn as well, despite its high mortality, may not always be an indication for surgical treatment. Approximately one third of these patients can be treated conservatively. Numerous studies proved that radiological findings, such as intestinal obstruction, volvulus or pneumoperitoneum may point to the need for surgical intervention [16, 17].

Treatment of duodenal atresia, after initial nasogastric tube placement, fluid and mineral balance regulation, relies on operative treatment via laparoscopic or open approach. Depending on the affected part of the duodenum, as well as intestinal lumen disproportion, intestinal continuity is established by creating duodeno-duodenal or duodeno-jejunal, end-to-side or side-to-side anastomosis [2].

Surgical strategies for treating intestinal volvulus include complete intestinal and mesenteric detorsion by counterclockwise rotation, wrapping the affected bowels with warm gauze and checking for viability after several minutes, followed by placing loops of small intestine to the right and colon to the left, as well as performing appendectomy (Ladd procedure). In cases where intestinal viability is unclear, second-look surgery can be performed within 24 – 48 hours. If a nonviable bowel is still present, it can be resected, followed by creating end-to-end anastomosis, or intestinal stoma [18]. In our patient, due to nonviable bowel loops, resection of this part of the intestine followed by a creation of anastomosis between proximal part of the duodenum and the rest of the jejunum seemed to be the best possible option.
Any congenital anomaly in the newborn period is a diagnostic-therapeutic challenge, even for an experienced surgeon. Association of two or more anomalies represents a specific form of challenge, which is commonly associated by numerous complications in affected organs or other organ systems.

Considering that radiological findings in our patient revealed a certain double bubble sign, we expected an isolated duodenal atresia. After intraoperative finding of conglomerate of intestinal loops, intestinal volvulus and meconium peritonitis, our decision to perform intestinal resection with primary anastomosis seemed as an appropriate solution for managing both, duodenal atresia and volvulus. However, meconium peritonitis was probably an important factor complicating the surgical course and postoperative recovery.

**Conclusion**

Treating congenital anomalies of the newborn is a challenge, even for an experienced surgeon, especially when associated with premature birth and low birth-weight. Despite adequate surgical techniques and intensive treatment, complex congenital anomalies are associated with high morbidity and mortality rates.

**References**

1. Alnosair AA, Naga MI, Abdulla MR, Al-Salem AH. Congenital duodenal atresia with ‘apple-peel configuration’ of the small intestines and absent superior mesenteric artery: a case report and review of literature. J Pediatr Surg Case Rep. 2014;2(5):215-8.

2. Morris G, Kennedy A Jr, Cochran W. Small bowel congenital anomalies: a review and update. Curr Gastroenterol Rep. 2016;18(4):16.

3. Miscia ME, Lauriti G, Lelli Chiesa P, Zani A. Duodenal atresia and associated intestinal atresia: a cohort study and review of the literature. Pediatr Surg Int. 2019;35(1):151-7.

4. Holcomb GW, Murphy JD, Ostlie DJ. Ashcraft’s pediatric surgery. 6th ed. Philadelphia: Elsevier; 2014.

5. Coran AG, editor. Pediatric surgery. 7th ed. Philadelphia: Elsevier; 2012.

6. Tonni G, Grisolia G, Granece R, Giacobbe A, Napolitano M, Passos JP, et al. Prenatal diagnosis of gastric and small bowel atresia: a case series and review of the literature. J Matern Fetal Neonatal Med. 2016;29(17):2753-61.

7. Best EJ, O’Brien CM, Carseldine W, Deshpande A, Glover R, Park F. Fetal midgut volvulus with meconium peritonitis detected on prenatal ultrasound. Case Rep Obstet Gynecol. 2018;2018:5312179.

8. Mitchell J, Punthakee Z, Lo B, Bernard C, Chong K, Newman C, et al. Neonatal diabetes with hypoplastic pancreas, intestinal atresia and gall bladder hypoplasia: search for the aetiology of a new autosomal recessive syndrome. Diabetologia. 2004;47(12):2160-7.

9. Galan-Gomez E, Sanchez EB, Arias-Castro S, Cardesa-Garcia JJ. Intrauterine growth retardation, duodenal and extrahepatic biliary atresia, hypoplastic pancreas and other intestinal anomalies: further evidence of the Martínez-Frias syndrome. Eur J Med Genet. 2007;50(2):144-8.

10. Concepcion J, Reh CS, Daniels M, Liu X, Paz VP, Ye H, et al. Neonatal diabetes, gallbladder agenesis, duodenal atresia, and intestinal malrotation caused by a novel homozygous mutation in RFX6. Pediatr Diabetes. 2013;15(1):67-72.

11. Satapara J, Bahri N. Antenatal diagnosis of duodenal atresia: USG and MRI findings. Eurorad. 2019. Doi: 10.1594/eurorad/case.16488

12. Gilbertson-Dahdal DL, Dutta S, Varich LJ, Barth RA. Neonatal malrotation with midgut volvulus mimicking duodenal atresia. AJR Am J Roentgenol. 2009;192(5):1269-71.

13. Tahjian DB, Moriarty KP. Duodenal atresia with an anomalous common bile duct masquerading as a midgut volvulus. J Pediatr Surg. 2001;36(6):956-7.

14. Canizo Vasques D, Izquierdo Renau M, Inarejos Clemente EJ, Bejarano Serrano M, Sunol Capella M, Morillo Palomo A. Intrauterine intestinal volvulus. J Pediatr. 2020;219:278-9.

15. Ballesteros Gómiz E, Torremadé Ayats A, Durán Felibidaló C, Martín Martinez C, Caro Tarragó A. Intestinal malrotation – volvulus: imaging findings. Radiologia. 2015;57(1):21-23.

16. Chandrasekaran N, Benardete D, Cariello L, Meraz D. Prenatal sonographic diagnosis of meconium peritonitis from duodenal atresia. BMJ Case Rep. 2017:2017: bcr2017219208.

17. Caro-Dominguez P, Zani A, Chitayat D, Daneman A. Meconium peritonitis: the role of postnatal radiographic and sonographic findings in predicting the need for surgery. Pediatr Radiol. 2018;48(12):1755-62.

18. Langer JC. Intestinal rotation abnormalities and midgut volvulus. Surg Clin North Am. 2017;97(1):147-59.

Bukarica S, et al. Duodenal Atresia and Intestinal Volvulus and Meconium Peritonitis