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Case Report / Olgu Sunumu

Multiple isolated enteric duplication cysts in newborn: An unusual cause of sepsis

Yenidoğanda multipl izole enterik duplikasyon kistleri: Sepsisin nadir bir nedeni

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Abstract

Gastrointestinal duplication cysts are uncommon congenital anomaly that may occur anywhere throughout the gastrointestinal system from the mouth to the anus. While cysts occur usually in the small bowel, the stomach ranks third after the small intestine and esophagus. As these cysts may be complicated by infection and other complications such as bleeding or obstruction, they should be treated with complete excision before the symptoms start. Although duplication cysts more often cause symptoms related to signs of obstruction, they may also present with clinical sepsis. In this article, we present a newborn case with a large duplication cyst adjacent to the ileum and the greater curvature of the stomach and a second simple cyst under the liver.

Keywords: Duplication cyst, gastrointestinal, newborn, sepsis, simple cyst, stomach.

Öz

Gastrointestinal duplikasyon kistleri, ağızdan anüse kadar gastrointestinal sistemin herhangi bir yerinde görülebilen, konjenital anomalilerdir. Kistler genellikle ince bağırsakta meydana gelirken mide, ince bağırsak ve yemek borusundan sonra üçüncü sırada yer alır. Bu kistler enfeksiyon, kanama veya tıkanma gibi diğer komplikasyonlara neden olabileceğinden, semptomlar başlamadan eksize edilmelidir. Duplikasyon kistleri daha sıklıkla tıkanma bulgularına bağlı semptomlar oluştursa da, klinik sepsis ile de birlikte seyredebilir. Bu yazıda ileuma ve midenin büyük kurvatürüne yapışık olan büyük bir duplikasyon kisti ve karaciğerin altında ikinci bir basit kisti olan yenidoğan bir olgu sunuldu.

Anahtar sözcükler: Duplikasyon kisti, gastrointestinal, yenidoğan, sepsis, basit kist, mide.

Duplication cyst, also known as enteric duplication cyst, is a rare congenital abnormality that occurs during fetal development. Duplication cysts can occur anywhere along the gastrointestinal tract, from the mouth to the anus, but are most commonly found in the small intestine or colon.[1] Gastrointestinal duplications have also historically been called, enterogenous cysts, ileum duplex, inclusion and cysts (giant diverticula).[2] Ladd[3] recognized the importance of accurate diagnosis and surgical intervention for duplication cysts and developed a surgical technique for their removal. He explained the term 'duplications of the alimentary tract' in 1937 and he also described triple pathological diagnostic criteria for the diagnosis: the presence of smooth muscle and mucosa that are similar to the adjacent gastrointestinal segment; epithelial lining that represents some part of the alimentary tract; and an adjacent to part of the digestive system.[4] These duplications are relatively rare congenital anomalies and incidence reported to be 1 in 4,500 births.[5] Patients usually present with abdominal distension or serious symptoms related to the location and size of the cyst. Duplication cysts may cause a leading point for intussusception and a localized volvulus of the small intestine. Although

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intestinal obstruction is the most common clinical presentation, newborns may also present sepsis clinic.[6,7]

Small bowel duplications account for about half of all gastrointestinal duplications, mostly in the ileocecal region. The esophagus is the second most common site, accounting for approximately 20% of digestive tract duplications. Gastric duplications account for 9% of gastrointestinal duplications.[8] Gastric duplications usually become symptomatic early in life and often present with pain, vomiting, or melena. Most are cystic, originates from the greater curvature and do not have connection with the lumen.[9] The operative treatment of small bowel duplications would vary depending on the type and size.

In this article, we present a newborn patient with a large duplication cyst adjacent to the ileum and the greater curvature of the stomach and a second simple cyst under the liver. The patient was admitted to the intensive care unit (ICU) with signs of clinical sepsis. Abdominal distension was noticed after hospitalization.

CASE REPORT

A 3,580-g, the full-term female infant was born at 39 weeks of gestation by normal spontaneous vaginal delivery. Pregnancy was uneventful. Prenatal ultrasound (US) and neonatal physical examination findings at birth were normal. She tolerated oral feeding and had daily defecation. The infant was asymptomatic until 22 days of life. She was admitted to our hospital with complaints of difficulty in sucking, weakness, and fever. Her family reported that there was no abdominal distension at birth and general condition worsened with abdominal distension in the last two days.

On clinical examination, the patient had a fever (38.3°C), icteric skin, severe abdominal distension and hypotonic muscle tone. There was no respiratory distress; however, she had a mild tachycardia (heart rate up to 170/min). Cardiovascular examination and chest radiography findings were normal. The limb pulse oximeter was around 95%.

Laboratory tests were performed. Accordingly, there was leukocytosis (25.6×10^9/L) and thrombocytopenia (97×10^9/L) detected in the complete blood count (CBC). Total bilirubin and direct bilirubin (5/4 mg/dL, respectively) were high, and C-reactive protein (CRP) (45 mg/L, normal range: 0-5) was high. The patient was hospitalized in the neonatal ICU with a preliminary diagnosis of clinical sepsis. Ultrasound and computed tomography (CT) were performed for abdominal distension. Ultrasound showed two cystic masses, the first one was adjacent to the stomach and ileum and the other was under the liver (Figure 1a). On CT, thin-walled cystic mass lesions measuring 10×8.5 cm to the left of the midline, 6×4 cm to the right of the midline, and 3×7 cm adjacent to the anterior abdominal wall were observed (Figure 1b, c).

Blood cultures were taken and intravenous broad-spectrum antibiotics were started (ampicillin amikacin and ornidazole). Oral feeding was stopped. Gastric drainage was provided with a nasogastric tube. On the seventh day of the hospitalization, abdominal distension and CRP (43 mg/dL) values persisted.

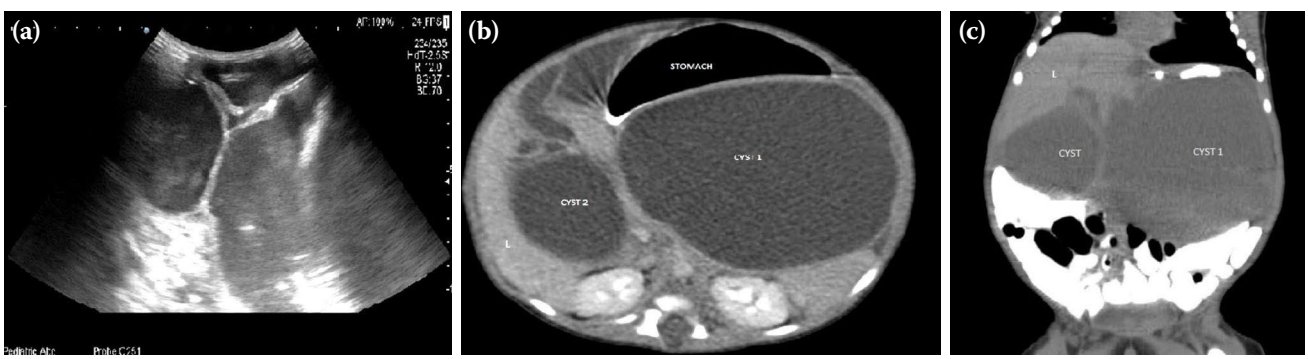

Figure 1. Radiological images of cysts. **(a)** Two cysts on ultrasound. **(b, c)** Computed tomography. Both cysts are seen in the coronal and transverse section, respectively.

We decided to remove the cyst to prevent further deterioration of the patient's clinical condition.

During surgery, the giant cyst was observed between the stomach and ileum. The small intestine could not be explored, as the cyst was too large. Giant cystic mass was approximately 20 cm in diameter. The giant cyst which was the near the stomach was opened and were aspirated. Approximately 500 cc cyst fluid was aspirated. After the aspiration, the small intestines were explored (Figure 2). The abdomen was scanned and the second cyst was seen between the liver and the cecum. The second cyst was approximately 10 cm in diameter and extending from liver to the caecum. It was adherent to the gallbladder as well. First, the cyst that adhered to the liver and gallbladder was totally removed from the superior and inferior to the caecum.

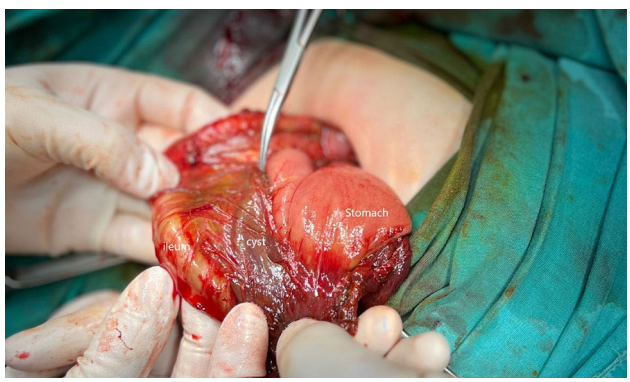

Figure 2. The image of giant cyst adjacent to the greater curvature of the stomach and ileum.

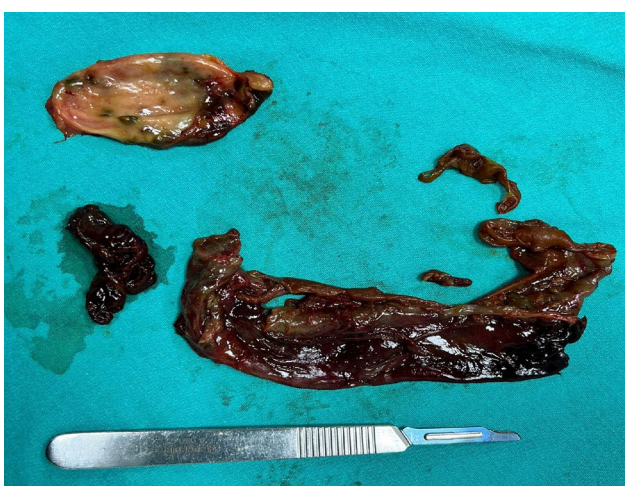

Figure 3. The image of the cysts after they are taken out of the abdomen in operation.

Afterwards, the huge cyst adjacent to the greater curvature of the stomach and jejunum was removed (Figure 3). As there was no wall in the ileum resection, anastomosis was not needed. The pathology result of the specimens were gastric duplication cyst and enteric duplication cyst, respectively.

DISCUSSION

Gastrointestinal duplication cysts are very uncommon and potentially dangerous congenital anomalies that may occur anywhere along the gastrointestinal tract from the mouth to the anus. Before the widespread use of prenatal US screening, pediatric surgeons typically encountered duplications when they resulted in intestinal complications.[9] The major abdominal presentation is bowel obstruction, which may occur by a variety of mechanisms. Gastrointestinal hemorrhage is a less frequent manifestation of duplications. This may occur due to the presence of heterotopic gastric mucosa, which is present in about 40% of esophageal duplications, one-third of small bowel duplications, and less than 10% of colorectal duplications.[10] In this case, although there was a simple cyst adjacent to the stomach, no melena or intestinal obstruction was observed.

Most small bowel duplications are cystic and can be completely excised with a segmental bowel resection. Long, tubular small bowel duplications are rare but present a challenge, since total excision may result in short bowel syndrome.[11] In this case we presented, excision was not difficult, as the cysts were not tubular and had a simpler structure. If it was attached to the intestine and had a long tubular structure, resection would be required. As such duplication cysts may be complicated by infection, bleeding, intestinal obstruction or volvulus, and may lead to malignant transformation, many surgeons believe that they should be treated by early complete excision when possible.[12] Ideally, the goal for the management of duplication cysts should be to make a diagnosis and to provide treatment before the onset of symptoms or complications. Development of adenocarcinoma of the mucosal lining is rare, with only a few cases been described in the literature.[13]It is, therefore, acceptable to manage asymptomatic duplications that are not amenable to resection expectantly. Antenatal diagnosis is often possible and can be

followed closely in the postpartum period after appropriate evaluation. In our patient's antenatal follow-up, no cyst was diagnosed on US screen and an uneventful pregnancy process was observed. Even after delivery, no abdominal distension was noticed.[14] Interestingly, the patient developed abdominal distension with clinical sepsis. However, we could not detect infectious bacterial agent in the culture scans. Gastric duplication cyst should be kept in mind during evaluation of an infant with an abdominal mass and vomiting. The newborn did not have vomiting. We decided to perform surgery not based on the clinical condition of the patient, but based on radiological appearance.[15]

Radiological imaging is very useful in diagnosis, it should be kept in mind that surgery is compulsory for definitive diagnosis as well as treatment. Excision of gastric duplication cyst with gastric preservation is usually safe. The most common initial screening examination for these patients should be US scan. Computed tomography should be performed, when the diagnosis is unclear on US.[15]

In conclusion, gastrointestinal duplication cysts have life-threatening complications. If they are detected in the antenatal period, further imaging should be performed after birth. Almost all surgeons recommend removing the cysts so that they do not cause complications.

Patient Consent for Publication: A written informed consent was obtained from the parent of the patient.

Data Sharing Statement: The data that support the findings of this study are available from the corresponding author upon reasonable request.

Author Contributions: Conception, Literature review, conceived and designed the analysis, collected the data, Wrote the paper: S.K.; Collected the data, Critical review: S.T., M.F.Ş.

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