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CASE REPORT

A newborn with an enteric duplication cyst

Rafael Silva Sampaio[1], Catherine Klein Colombiano[2], Isabela Oliveira Fiorio[3], Mattielli Dias do Carmo[4], Patrícia Leal Pinheiro[5], Juliana Marques Coelho Bastos[5], Gustavo Carreiro Pinasco[4,5], Katia Valéria Manhabusque[4,5]

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Abstract

Enteric duplication cysts are rare congenital anomalies of the gastrointestinal tract. They can be identified from the neonatal period or in the first two years. The pathogenesis is still uncertain opening possibility to several theories correlated to its formation during the embryogenesis. The clinical manifestations are very varied depending on the location and size of the lesion. Diagnosis is initially performed by abdominal ultrasonography and confirmed by histopathological examination. In these patients, the treatment is always surgical. This case report refers to a neonate with ultrasounds in intrauterine period showing abdominal cystic masses and its evolution to the definitive diagnosis of enteric duplication cyst, after surgical treatment and confirmed by histopathological analysis.

[1] Nossa Senhora da Gloria Children's Hospital, Medical Residency Program in Pediatric Intensive Care - Vitória - ES - Brazil.
[2] Hospital Santa Casa de Misericórdia de Vitória, Medical Residency Program in Neonatology - Vitória - ES - Brazil.
[3] Ana Nery Hospital, Medical Residency Program in Pediatric Cardiology - Salvador - BA - Brazil.
[4] Hospital Santa Casa de Misericórdia de Vitória, Pediatrics - Vitória - Espírito Santo - Brazil.
[5] Escola Superior de Ciências da Santa Casa de Misericórdia de Vitória, Scientific Writing Office, Pediatrics - Vitória - ES - Brasil.

Correspondence to:
Rafael Silva Sampaio.
Hospital Infantil Nossa Senhora da Gloria. Rua Alameda Mari Ubirajara, nº 205, Santa Lucia,
Vitória, ES, Brazil. CEP: 29056-030. E-mail: rafaelsampaio_@hotmail.com

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INTRODUCTION

Enteric duplication cysts are rare embryo development anomalies that originate in the gastrointestinal tract[1]. They occur on 0.2% of newborns[1], with a slight preference for male individuals[2].

Diagnosis is usually performed before birth or within the first years of extrauterine life[3]. Morbidity is best prevented when patients are diagnosed early on and treated promptly with surgery[4].

This report describes the clinical evolution of a newborn with two abdominal cysts found in prenatal ultrasound scans. Histopathology examination of surgical specimens confirmed the diagnosis of enteric duplication cysts.

CASE REPORT

A female baby was born from a vaginal delivery after 38 weeks and three days of gestation. She was in cephalic presentation, with meconium in the amniotic fluid and rupture of membranes occurring during delivery. The newborn weighed 2096 g, had a head circumference of 31 cm, a chest circumference of 29cm, and an abdominal circumference of 32 cm. Her length was 43 cm and she had an Apgar score of 7/7. No resuscitation maneuvers were required in the delivery room. Physical examination revealed she had a cyst in the occipital area, signs consistent with loops in the abdomen, a palpable moving cyst in the iliac fossae more evident to the right, and a dislocated right knee.

Ultrasound (US) examination performed in the second trimester found two voluminous cysts in the abdomen, with clear, even contours, extending from the flanks to the pelvis, measuring 5.4 cm and 4.6 cm in their larger axes. US examination in the third trimester showed non-specific thin-wall cysts containing homogeneous fluid showing no color on Doppler US, extending from the pelvis to the flanks.

One hour after birth, the newborn developed respiratory distress and tachypnea, which required referral to a neonatal intensive care unit (NICU) for oxygen therapy with non-invasive mechanical ventilation. Respiratory symptoms worsened and the patient had to be intubated and placed on antibiotics. Chest X-ray scans did not show alterations, while abdominal X-ray images showed a distended abdomen, with a mass effect on the right flank, right iliac fossa, and left iliac fossa. The patient became hemodynamically unstable and was administered vasoactive drugs.

Abdominal US images of the adnexa showed two thin-wall cysts with homogeneous anechoic content without solid contents measuring 4.37 cm (right) and 3.48 cm (left). The medical and surgical teams decided to wait until the patient's condition stabilized.

Physical examination performed after the patient became hemodynamically stable found that her abdomen was swelling gradually and becoming harder to palpation. She did not have elimination dysfunctions. Abdominal X-ray scans showed a pattern consistent with abdominal distension. Abdominal US images revealed voluminous cysts occupying the flanks/iliac fossae, one to the right with fine septa measuring 7.9 cm and another to the left measuring 7.1 cm. In light of the enlargement of the abdominal cysts and the alterations on physical examination, the Pediatric Surgery team prescribed surgery to the patient.

The patient underwent surgery 27 days after birth. The procedure consisted of the removal of two cysts from the mesentery, an enteroanastomosis following enterectomy, and an appendectomy (Figure 1). Histopathology examination of the surgical specimen revealed signs consistent with enteric duplication cysts, without signs of neoplasm. The patient remained in the NICU for another 24 hours to treat potential postoperative complications and stimulate breastfeeding. She was then discharged and referred to outpatient care.

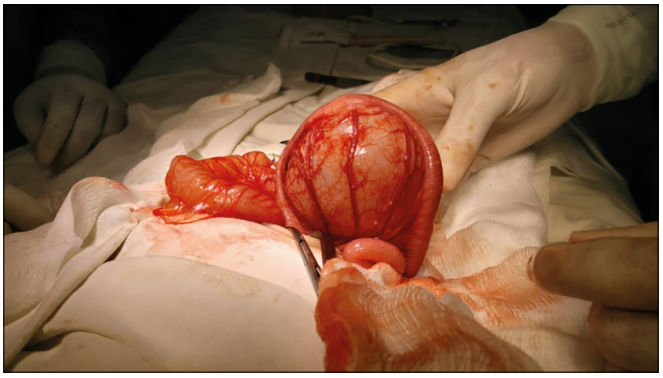


Figure 1. Surgery to remove enteric duplication cysts.

DISCUSSION

Enteric duplication cysts are found mainly in the ileum (33%), followed by the esophagus (20%), colon (13%), jejunum (10%), stomach (7%), and duodenum (5%)[5]. Intestinal duplications encompass a group of lesions that contain a wall of smooth muscle and enteral mucosa found only in the mesenteric border of the bowel[3].

Enteric duplication cysts appear between the fourth and eighth weeks of embryo development[5]. Many are the explanations for its development, including the split notochord theory; another theory that suggests failure in the normal regression of the diverticula in the embryo; median septum formation theory; and a theory that suggests an association between complex colorectal tubular duplications and urogenital anomalies due to partial or abortive gemination[6]. Malformations, particularly vertebral ones, are described in 50% of the patients with enteric duplication cysts[2].

Examination of the abdomen might not find anything in particular, but a moving mass may be found through palpation in 50% of the cases[2], as in the patient described herein. When no alterations are found in physical examination at birth, 70% of the newborns present signs and symptoms such as nausea,

vomiting, distended abdomen, and palpable abdominal masses within a year of birth. Recurrent abdominal pain is one of the most common symptoms and is usually attributed to high pressure inside the duplication cyst due to secretion accumulation[5].

US is the most widely used diagnostic method for enteric duplication cysts. Magnetic resonance imaging (MRI) and computed tomography (CT) are used in esophageal enteric duplication cysts and as an auxiliary tool in cases of difficult surgery[5]. Simple X-ray images are not very specific[3], but are customarily used in emergency situations on account of ease of access, as in this case report. Our patient was decompensated within an hour of birth and the X-ray images taken did not find lung alterations, but signs of abdominal distension with a mass effect on the flanks and iliac fossa, which might explain the patient's hemodynamic instability. US scans taken at a later moment confirmed that the cysts seen in the prenatal scans had considerably increased in size.

The number of cases of enteric duplication cysts identified prenatally has increased, mainly due to the improvements in prenatal ultrasound screening programs and image resolution[5]. However, only 20-30% of the cases are identified before birth[5,7].

Prenatal recognition of enteric duplication cysts permits better planning of the postnatal investigations required to establish a diagnosis and find associated malformations[4]. Diagnoses of enteric duplication cysts are confirmed after histopathology examination of surgical specimens with mucosal tissue from parts of the gastrointestinal tract[8].

Enteric duplication cysts tend to grow gradually and might cause symptoms and potentially fatal complications, including obstruction – volvulus or intussusception, massive bleeding or the development of malignant disease in adult age[5,7]. Therefore, adopting an expectant approach in cases of asymptomatic duplication cysts is not recommended[9]. An expectant approach was initially adopted in our case due to hemodynamic instability. Our patient underwent surgery as soon as possible.

Surgical approaches vary and may involve only the excision of the cyst[5]. Resection of the segment of the bowel that contains the duplication, followed by end-to-end anastomosis, is the most common procedure. More radical approaches, such as segmental or total colectomy, might be needed in cases in which vascularization is shared by the bowel and the duplication cyst[3]. Since it may recur and on account of the chances of development of malignant disease, it is important to make sure that the cysts are fully resected. Minimally invasive surgery has been used more recently and most cysts can be resected via thoracoscopy or laparoscopy[5].

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