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Case report

Intestinal obstruction caused by a duplication cyst of the cecum in a neonate

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= Abstract =

Duplication cysts are rare congenital malformations, that may be detected anywhere along the alimentary tract, and they may communicate with the intestinal tract. Cystic duplication of the cecum is especially rare. About 80% of these cases are detected in the first 2 years of life as a result of an acute intestinal obstruction, which manifests as vomiting, recurrent abdominal pain, recurrent gastrointestinal bleeding and constipation. We report a case of intestinal obstruction secondary to a duplication cyst of the cecum in a neonate. The patient underwent surgery and was diagnosed subsequently, and is presently healthy. (Korean J Pediatr 2009;52:261-264)

Key Words : Duplication cyst, Cecum, Intestinal Obstruction, Neonate

Introduction

Duplication cysts are rare congenital malformations that are characterized by their attachment to the alimentary tract, presence of a well-developed coat of smooth muscle, and an epithelial lining resembling some part of the alimentary tract[1]. Furthemore, cystic duplication of the cecum is especially rare, with only 18 reported cases in the English literature till date[1]. Different authors have used terms such as enterogenous cyst, ileum duplex, giant verticula, and unusual Meckel's diverticulum to describe the congenital cystic abnormalities of the gastrointestinal tract. In 1937, Ladd recommended the use of the term "alimentary tract duplications" for such congenital malformations[2-4]. Duplications of the alimentary tract may be detected at any age and they may occur anywhere along the course of the gastrointestinal tract. We report our experience with a full-term infant born to a mother who had no notable drug or clinical history with the except of polyhydramnios. The infant was brought to the emergency with symptoms of vomiting immediately after feeding, and abdominal distensions; upon further examination and operation, he was diagnosed as a case of duplication cyst.

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Case report

A full-term male infant weighing 3,480 g was born by cesarean section, and passed meconium. On Day 2 after birth, the patient was transferred to our hospital because he began vomiting bile-stained fluid immediately after feeding and abdominal distention. Maternal history was non-specific except for the presence of polyhydramnios. Physical examination revealed diffused abdominal distension, and a small-sized firm mass measuring approximately 2×2.5 cm that was palpable in the Right Lower Quadrant (RLQ) area. Other examinations were normal. Hematologic analysis revealed the following: hemoglobin, 17.1 g/dL; hematocrit, 48.9%; white blood cell count, 16,980/μL; platelet count, 217,000/μL; and total bilirubin, 10.17 mg/dL. Venous blood gas analysis revealed the following: pH, 7.406; PCO_2, 35.4 mmHg; PO_2, 30.2 mmHg; base excess, -2.1; and HCO_3^-, 21.7 mM/L. Other findings were non-specific. A plain X-ray of the abdomen was suggestive of an intestinal obstruction; ultrasound examination revealed a cystic lesion measuring approximately 4×3 cm-sized cystic lesion in the RLQ area.

On the second day after admission, vomiting continued to

occur immediately upon feeding. He was in the hypokalemic state of 3.09 mEq/L, so we administrated potassium chloride intravenously. We examined the abdominal CT for the possibility of a duplication cyst. Abdominal CT revealed compression of the ascending colon and a cystic lesion in the retroperitoneal area, which was suggestive of duplication or peritoneal cysts (Fig. 1).

Other anomalies were not observed. Due to the obstruction of the ascending colon by the cystic lesion, we also examined the colon series. During the colon series, we could observe the passage of barium from the colon to the small bowel, but only a small amount. He had a consultation with General Surgery and an operation was performed. Afterwards, biopsy demonstrated a diagnosis of a duplication cyst of the cecum. Since the operation (Fig 2, 3), he had any complications.

On the postoperative 7th day, he could be fed completely. The baby was discharged on the 9th day after admission.

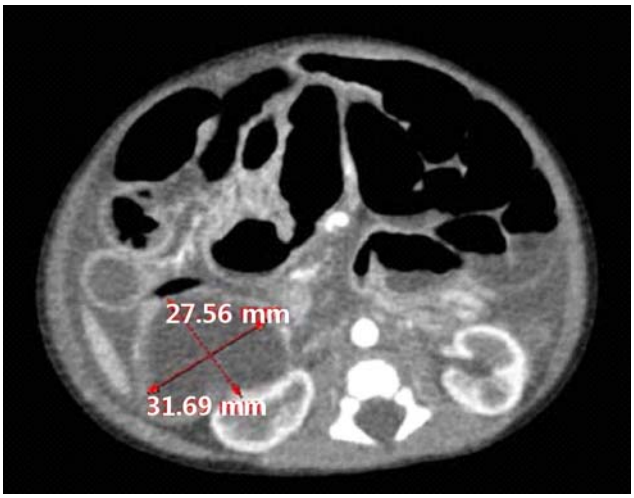

Fig. 1. Abdominal CT shows a 2.7×3.1 cm sized retroperitoneal cystic mass with compression of the ascending colon and cecum.

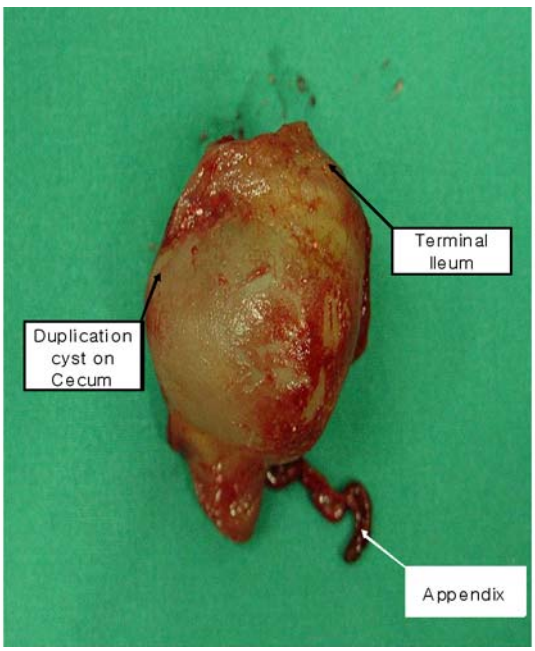

Fig. 2. The cystic mass which was attached to the ileocecal area and cecum was removed.

Discussion

Duplication cysts are defined as spherical or tubular structures that are firmly attached to at least one point in the alimentary tract, possess well-developed coats of smooth muscle, and have an epithelial lining resembling some part of alimentary tract[5, 6]. In Japan, 49 cases of duodenal duplication cysts have been reported to date[7].

Commonly, duplication cysts are communicated with the intestinal lumen, involve the mesenteric border of the associated alimentary tract, and share a blood supply with the native bowel. There are 3 categories for classification: (i) localized duplications (ii) duplications associated with spinal cord defects and vertebral malformations and (iii) duplications of the colon. Multiple duplication cysts are found in 10-15% of cases[5].

The etiology of intestinal duplications remains unclear. The persistence of embryonic diverticulae during the development of the gastrointestinal tract, intrauterine vascular accidents, and recanalization and fusion of embryonic longi-

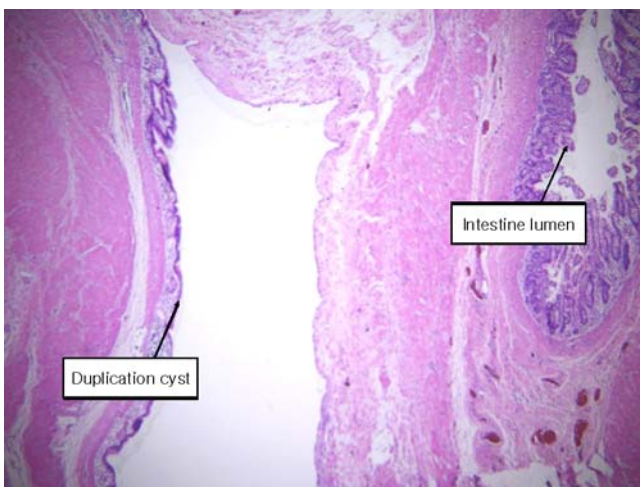

Fig. 3. Biopsy shows the cystic mass is similar to the luminal layer of the small bowel.

tudinal folds are some of the etiological hypotheses that have been proposed to explain the formation of duplication cysts in the intestines[1, 8, 9].

Morphologically, they are divided into spherical and tubular types, the tubular type being less commonly found in the small intestine than the spherical type[1].

Duplications of the alimentary tract may present at any age, but 80% present in first 2 years of life and only 13% of all alimentary duplications are colonic. Among abdominal duplications, the ileum is the most common site of involvement[10, 11]. Most cases develop within the first 3 months of life and a palpable mass is the most frequent mode of presentation, seen in 50% of these infants. Conversely, duplication cysts of the cecum are very rare. In an 18-year review, Grosfeld et al.[11] could find only one case of cecal duplication in a neonate; barium study revealed a palpable abdominal mass with a filling defect. Oudshoorn and Heij[1] have reviewed 362 cases of duplication cysts reported in the literature and found only 16 cases of cecal duplications.

Symptoms of intestinal duplication cysts vary according to the size, morphology and location of the cysts. Common symptoms include acute intestinal obstruction, vomiting, recurrent abdominal pain, recurrent gastrointestinal bleeding, constipation or an incidental detection. A communication between the gastrointestinal tract and the duplication is found in only 20% of the cases. Thirty percent of the patients have ectopic gastrointestinal mucosa[1, 3, 12]. Muscular atrophy secondary to vascular insufficiency, primary aplasia of an intestinal segment, and congenital or acquired damage to the nerves of the myenteric plexus can all result in segmental dilatation of ileum[8, 13]. Although ultrasound, CT scan and MRI have been useful, a correct preoperative diagnosis is seldom possible as the symptoms are so varied and the entity so rare. Ultrasonography and barium studies are helpful imaging modalities for the diagnosis of duplication cysts [14-16]. Ultrasound shows an echogenic inner mucosal layer with a hyperechogenic outer muscle layer. The CT scan shows a well-defined cystic structure with low attenuation and contrast enhancement of the outer rim[15, 16]. Some enteric duplication cysts contain gastric mucosa and may be demonstrated with a 99Tcm-pertechnetate scintigraphy[15-17]. Although rare, duplication cysts are an important differential diagnosis to consider in children, especially in neonates who present with a palpable abdominal mass such as mesenteric cyst, ovarian cyst, pancreatic cyst or choledochal cyst. Malignant changes of a duplication cyst have been reported in adults, but it is thought that these lesions are relatively benign in childhood[9]. End-to-end anastomosis after segmental resection of bowel with duplication cysts is the preferred choice of treatment in these cases[9, 12].

In this report we describe a fullterm infant who experienced vomiting immediately upon feeding and who presented with abdominal distension; the infant was diagnosed postoperatively as having a duplication cyst.

한 글 요 약

신생아에서 맹장의 장 중복낭종에 의해 발생한 장 폐쇄 1예

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금승운 · 황민우 · 나종인 · 유승택 · 강동백* · 오연균

장 중복낭종은 드문 선천성 기형 질환으로 구강에서 항문까지 어느 곳에서나 발견할 수 있고 장관과 교통하기도 한다. 특히 맹장의 장 중복낭종은 더욱 드문 질환이다. 이들은 구토나 반복적인 복통과 위장관 출혈 및 변비 등의 증상으로 급성 장폐색을 발생시키며 생후 2년내 80%에서 발견된다. 저자들은 신생아에서 맹장의 장 중복낭종에 의해 발생한 장 폐색 1예를 경험하였기에 보고하고자 한다.

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