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Congenital intestinal atresia associated with a mesenteric cystic lymphangioma in a low birth weight neonate: A case report

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ABSTRACT

INTRODUCTION: Congenital intestinal atresia requires emergency surgery soon after birth. Lymphangioma, a benign tumor, is caused by an anomalous lymphatic system. We report a case of congenital intestinal atresia associated with a mesenteric cystic lymphangioma in a low birth weight neonate.
PRESENTATION OF CASE: At 21 weeks of pregnancy, ultrasonography revealed a cystic lesion in the fetal abdominal cavity. At 31 weeks, magnetic resonance imaging showed dilatation of the small intestine. This low birth weight (1752 g) male infant was born by vaginal delivery at 32 weeks 3 days' gestation. Laparotomy on day 2 of life revealed jejunal atresia and a mesenteric cyst. The cyst was removed and intestinal anastomosis was performed. Histologically, the cyst proved to be a mesenteric lymphangioma.
DISCUSSION: The most popular theories regarding the mechanism of congenital intestinal atresia include reperfusion injury and intestinal tract blood flow disturbance. In this fetus, intestinal torsion had occurred around the mesenteric cystic lymphangioma, which apparently disturbed the mesenteric blood flow and caused intestinal atresia.
CONCLUSION: There have been few reports of the combination of a mesenteric cystic lymphangioma and congenital intestinal atresia. This case supports the theory that small bowel atresia and stenosis are caused by accidental blood flow disturbance.

1. Introduction

Congenital intestinal atresia requires emergency surgery during the neonatal period. The exact mechanism of atresia is controversial, although various theories have been proposed. The most popular theories are that congenital intestinal atresia is caused by reperfusion injury or blood flow disturbance in the intestinal tract [1,2].

Lymphangioma is a benign tumor caused by anomalous development of the lymphatic system [3]. Most pediatric lymphangiomas are located in the neck or axilla. Lymphangioma of the small bowel mesentery is rare and accounts for only 1% of all lymphangiomas [3,4]. Congenital intestinal atresia associated with mesenteric cystic lymphangioma has rarely been reported. We therefore report a surgical case of congenital intestinal atresia associated with mesenteric cystic lymphangioma in a low birth weight infant. The work has been reported in line with the SCARE criteria [5].

1.1. Presentation of case

A male newborn was diagnosed prenatally with intestinal atresia or duplication. Ultrasonography at 21 weeks' gestation revealed a cystic lesion in the fetal abdominal cavity. At 28 weeks' gestation, fetal intestinal dilation and polyhydramnios were apparent, which suggested small intestinal atresia. The fetus was again evaluated at 31 weeks' gestation using magnetic resonance imaging. This showed dilation of the small intestine and the cystic lesion (Fig. 1), which suggested intestinal atresia or duplication.

The male infant was born vaginally to a 27-year-old mother with gravidity 0 and parity 0 at 32 weeks 3 days' gestation. His birth weight was 1752 g. There was no remarkable medical history in his parents. They had no surgical history, allergies, oral medications, or history of alcohol consumption or smoking. Physical examination showed significant abdominal distension with diminished bowel sounds and no palpable abdominal mass. Blood biochemistry and other laboratory results were normal. No metabolic alkalosis was observed. Plain abdominal radiography showed a dilated stomach and upper jejunal dilation (Fig. 2a). Contrast-enhanced radiography

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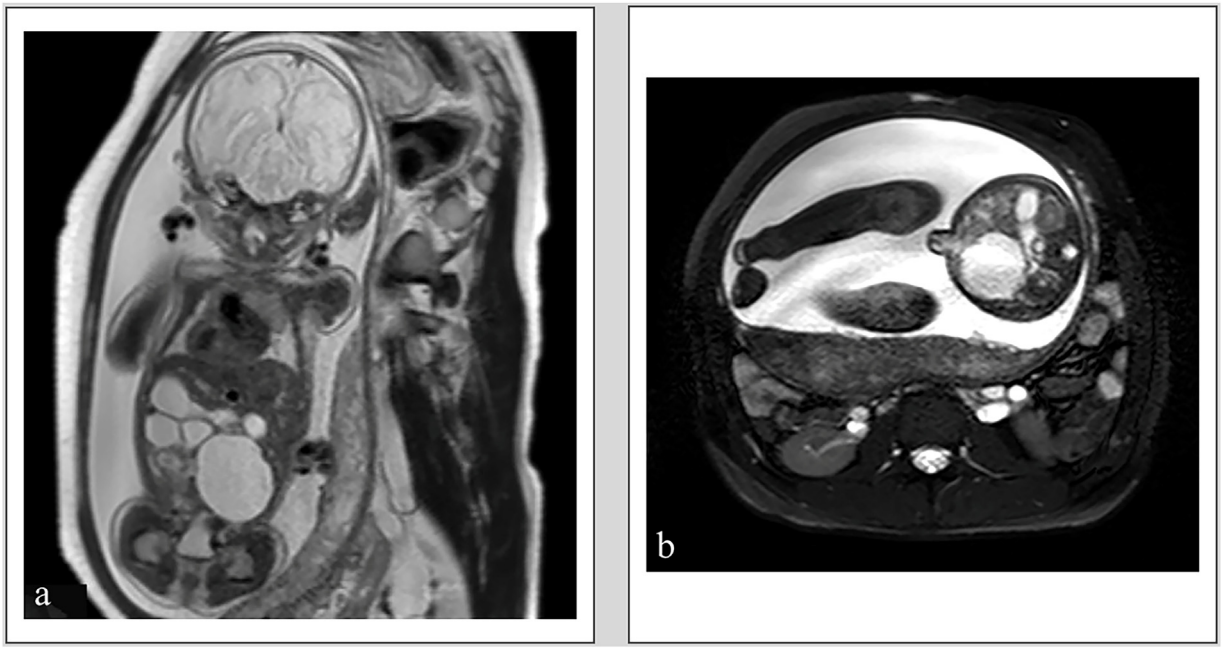

Fig. 1. Magnetic resonance imaging shows the dilated small intestine (a) and the cystic lesion (b).

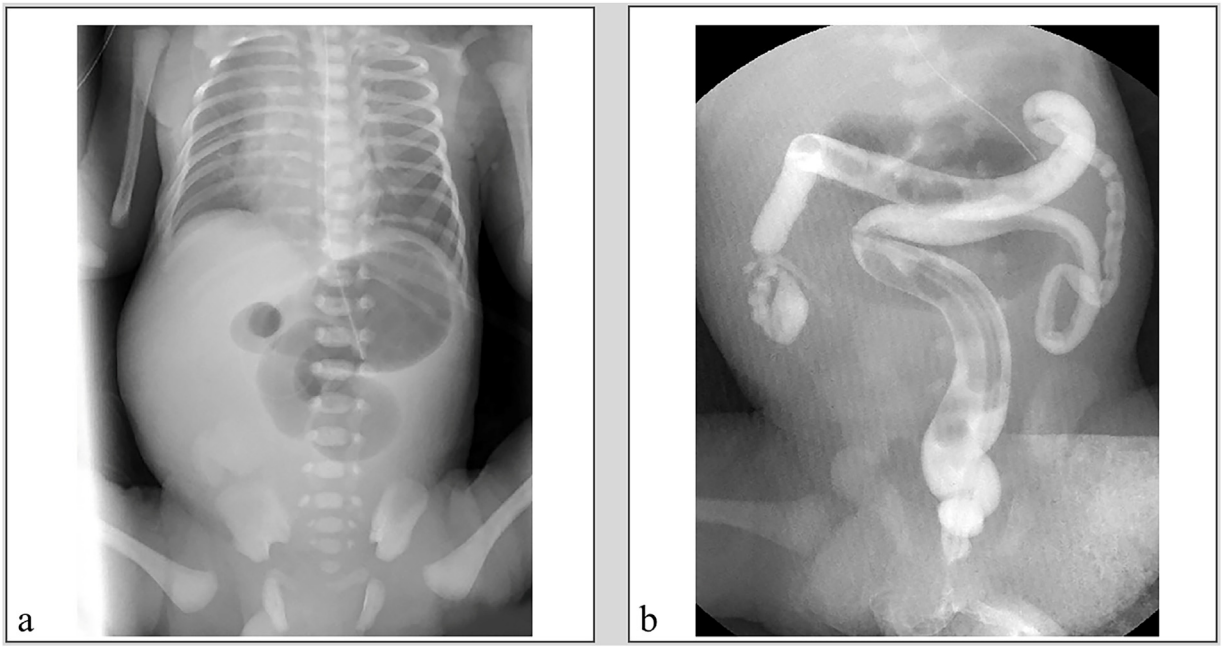

Fig. 2. a Abdominal plain radiography shows the dilated upper jejunum. b Contrast-enhanced radiography shows the microcolon.

showed that it was possible to obtain images as far as the terminal ileum, but the colon appeared as a microcolon (Fig. 2b).

On the second day of life, laparotomy was performed with a right transverse incision above the level of the umbilicus. It revealed a 45-mm cystic mass at the jejunal mesentery and 360° torsion of the jejunum, along with the cyst (Fig. 3a). When the torsion was released, we observed jejunal atresia 14 cm distal to the ligament of Treitz and a cystic mass that had developed in the mesentery of the proximal jejunal segment of the small intestine. The ratio of the diameters of the proximal and distal jejunal segments was 3:1 (Fig. 3b). An end-to-back anastomosis was performed. The cystic mass was excised together with the atretic jejunal segment (Fig. 3c). There was no malrotation, appendectomy was not required, and an intraabdominal drain was not needed. Histological examination of the specimens showed intestinal atresia and a mesenteric cystic lymphangioma (Fig. 4a, b).

Oral feeding began on postoperative day 7. During the postoperative course, the infant developed premature apnea attacks and gastrointestinal allergies that required conservative treatment. He was discharged 3 months after the surgery and had a good postoperative course.

2. Discussion

Lymphangioma is a benign tumor that arises because of anomalous development of the lymphatic system [3]. Cystic lym-

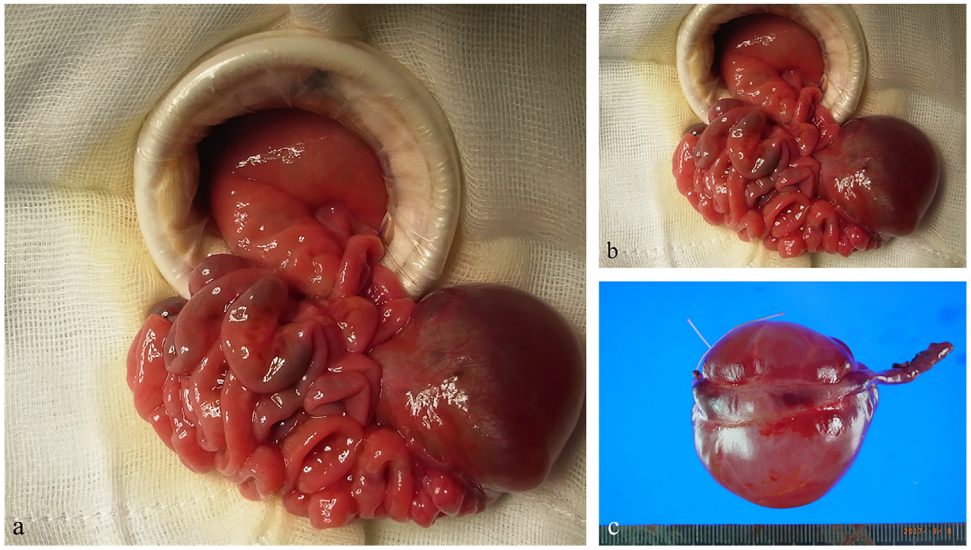

Fig. 3. a The cystic mass at the jejunal mesentery and the 360° torsion of the jejunum with the cyst are apparent. b Jejunal atresia occurred 14 cm distal to the ligament of Treitz. The difference in the diameters of the proximal and distal jejunal segments was 3:1. c Resected cystic mass and atretic jejunal segment.

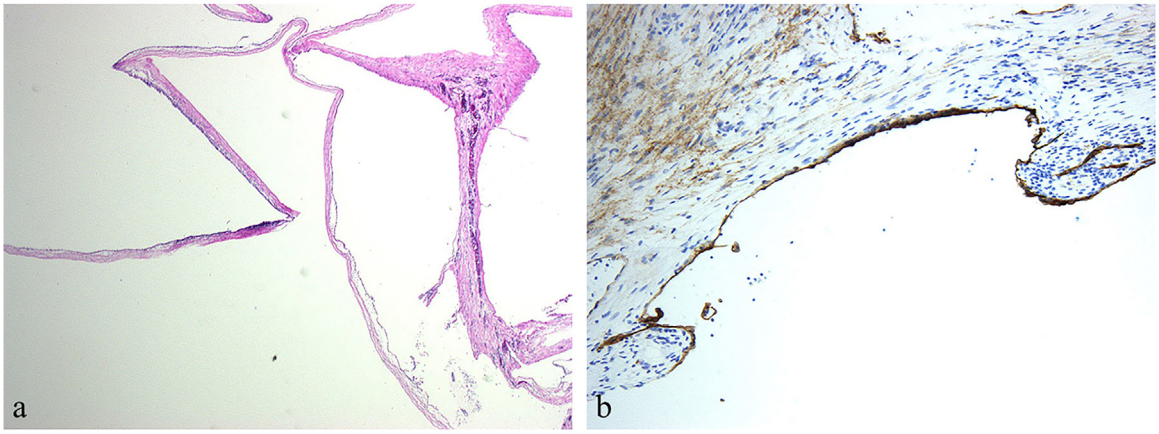

Fig. 4. a Cystic spaces are apparent (HE, ×100). b Immunoreaction for D2-40 shows positivity of the flattened endothelial cells lining the lymphatic spaces (anti-D2-40 antibody immunolabeling) (×100).

phangioma is histopathologically characterized by a thin, irregular wall covered by endothelium that contains lymphatic tissue [5]. Abdominal cystic lymphangiomas are rare, with a reported incidence of 1/20,000–250,000 [6,7]. Among all intraabdominal lymphangiomas, 10% involve the mesocolon and 5% the retroperitoneum. The small bowel mesentery is affected more frequently [8,9]. In all, 65% of lymphangiomas occur at birth and 90% during the first 2 years of life [10]. They can remain asymptomatic for a lifetime or can produce such complications as secondary infection, rupture with hemorrhage, and volvulus or intestinal obstruction, depending on the increasing size of the lesion [11,12]. As recent findings, Luks et al. reported the PIK3CA mutation in lymphatic malformation [13]. The mechanism by which the PIK3CA mutation that occurs during embryogenesis causes malformation is not fully understood, but it is thought to be due to the role of PI3K in signaling pathways such as the vascular endothelial growth factor pathway [14,15].

Congenital intestinal atresia requires emergency surgery during the neonatal period. Intestinal atresia is one of the most common causes of intestinal obstruction in neonates, with an incidence of 1/5000 newborns [16]. A retrospective review of 277 neonates reported that the levels of obstruction were 49.8% (138 cases) in the duodenum, 46.6% (128 cases) in the jejunoileal portion, and 7.6% (21 cases) in the colon. Obstruction was seen in multiple locations in 3.6% (10 cases) of the neonates [17]. In another report, the proximal jejunum was affected in 31% of cases, the distal jejunum in 20%, the proximal ileum in 13%, and the distal ileum in 36%. Multiple areas of atresia were recorded in 6%–20% of cases [18]. The exact mechanism of atresia is controversial, with various theories having been proposed. The most popular theories for the development of congenital small intestinal atresia are reperfusion injury or blood flow disturbance in the intestinal tract [1,2]. Louw and Barnard, who conducted animal studies reported that small intestinal atresia is caused by damage to the mesenteric blood vessels [2]. Their theory that intestinal atresia and stenosis are caused by accidental blood flow disturbance has been supported.

There have been few reports of the association of a mesenteric cystic lymphangioma and congenital intestinal atresia [19]. In addition, most reports are postnatal diagnosis cases. Kosir et al. reported 13 postnatal cases of abdominal lymphangioma during a 16-year period. Four of the patients had volvulus, and two had intestinal gangrene [20]. In our case, ultrasonography at 21 weeks' gestation showed a cystic lesion in the fetal abdominal cavity, and at 28 weeks' it involved the fetal intestine. It appears that the intestinal torsion occurred around the mesenteric cystic lymphangioma dur-

ing this prenatal period. Thus, the mesenteric blood flow, which had been disturbed by torsion, caused the congenital intestinal atresia. This case suggests reconsidering the pathogenesis of congenital intestinal atresia.

Recently, the early and accurate prenatal diagnosis has enabled because of developments in diagnostic imaging technology. However, the reports including clinical progress in image findings from the prenatal period regarding the congenital intestinal atresia due to torsion by the mesenteric cystic lymphangioma have not been carried out. This is an informative report for prenatal diagnosis of intestinal atresia and the mesenteric cystic lymphangioma.

3. Conclusions

We report a rare surgical case of congenital intestinal atresia associated with mesenteric cystic lymphangioma in a low birth weight infant. The case supports the theory that small intestinal atresia and stenosis are caused by accidental blood flow disturbance. This report could be useful for prenatal diagnosis in similar cases.

Declaration of Competing Interest

The authors declare no conflicts of interest in association with the present study.

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Ethical approval

The study is exempt from ethical approval in our institution as it is a “Case report” and not a research study.

Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Author contribution

AF and TI treated the patient and designed the study. AF wrote the manuscript. NK, TK and TM critically reviewed the manuscript. All authors read and approved the final manuscript.

Registration of research studies

None.

Guarantor

AF and TM accept full responsibility for the study and guarantee its accuracy.

Provenance and peer review

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