Left-Sided Gastroschisis: A Rare Congenital Abdominal Wall Defect

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ABSTRACT

Gastroschisis is a congenital abdominal wall defect that occurs at the immediate right of the umbilicus. Left-sided gastroschisis, which is located in the left periumbilical region, is extremely rare, as only 22 cases have been reported worldwide. Although most cases of gastroschisis are not complicated by other anomalies, extra-intestinal anomalies are more commonly associated with left-sided gastroschisis than classic right-sided gastroschisis. In this report, we describe a case of left-sided gastroschisis complicated by cardiac anomalies. We also discuss the suggested theories of its pathogenesis and present a literature review. For patients with left-sided gastroschisis, we recommend evaluation of the associated anomalies because extraintestinal anomalies in the genitourinary, cardiovascular, or central nervous systems are more common in left-sided gastroschisis than in classic right-sided gastroschisis.

Key Words: Abdominal hernia, Gastroschisis, Omphalocele

INTRODUCTION

Gastroschisis is a congenital abdominal wall defect characterized by a full-thickness defect to the right side of the umbilical cord¹⁾. The incidence rate of this condition was originally 0.3–1 case per 10,000 births¹⁾, but it has now increased to 5 cases per 10,000 births²⁾. Most infants with gastroschisis do not have any other anomalies. However, left-sided gastroschisis is an extremely rare anomaly and is located in the left periumbilical region. Only 22 cases of left-sided gastroschisis have been reported to date^{3,4)}. It is interesting that most patients with left-sided gastroschisis were found to have extraintestinal anomalies⁵⁾. In this report, we describe the case of a male infant with left-sided gastroschisis who was successfully treated with staged operation. We also include the suggested theories of its pathogenesis and a review of medical literatures that describes the presentation and associated anomalies of left-sided gastroschisis. To the best of our knowledge, this is the first reported case of left-sided gastroschisis in South Korea.

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CASE REPORT

A male infant was born after 35 weeks 6 days of gestation through cesarean delivery. The surgery was performed electively, as an abdominal wall defect was diagnosed by using prenatal ultrasonography. The infant's weight was 1,970 g (10–25th percentile), height was 44 cm (10–25th percentile), and head circumference was 32 cm (25–50th percentile). His mother's age was 24 years and father's age was 27 years. The mother's obstetric history was gravida 3, para 1. At 19 weeks of gestation, a prenatal ultrasonography detected the development of gastroschisis in the infant, which had eviscerated both the small and large bowels. The mother had been smoking during the course of the pregnancy. In the physical examination of the infant after birth, he was found with left-sided gastroschisis. The abdominal wall defect was located immediately left of the umbilical cord (Figure 1).

The defect was circular and approximately 3×3 cm in size. The eviscerated bowel was thickened and edematous, and not covered with a membrane. The general appearance of the patient was otherwise normal. His respiration was regular and respiratory rate was 35 breaths per minute. His pulse rate was 150 beats per minute. The umbilical cord vessels were normal, as they contained two arteries and a vein. The eviscerated bowel could not be reduced to fit into the abdomen, so primary closure of the defect was impossible. Therefore, we constructed an artificial silo immediately after delivery in the operating room. With the baby under general anesthesia, the eviscerated bowel was placed inside the silo so that it can undergo spontaneous reduction (Figure 2).

Echocardiography revealed the following anomalies: patent ductus arteriosus, atrial septal defect shunting flow from the left to the right, and peripheral pulmonary stenosis. Chromosomal analysis detected 46, XY, normal chromosomes without any

Figure 1. A circular defect approximately 3×3 cm in size. The eviscerated bowel is thickened and edematous, and not covered with a membrane.

Figure 2. An artificially constructed silo was applied immediately after delivery, under general anesthesia. Then, the eviscerated bowel was placed inside the silo.

Figure 3. As the size of the eviscerated bowel had decreased, we inserted it in the abdominal cavity and repaired the abdominal wall defect at 18th postnatal day.

structural aberrations. The patient did not have brain or renal anomalies. He could breathe spontaneously without any help of ventilators or oxygen. Fourteen days after the operation, the size of the eviscerated bowel had decreased (Figure 3). Thus, we inserted the eviscerated bowel into the abdominal cavity and repaired the abdominal wall defect. Enteral feeding was initiated at postoperative day 1. Full enteral feeding was achieved by the 18th postnatal day, and the duration of parenteral nutrition was 15 days.

DISCUSSION

The sex ratio in cases of gastroschisis is 1.0:1.4 (male to female)^{6,7)}. Most cases of gastroschisis are not complicated by other anomalies. The incidence of associated anomalies is about 10%⁵⁾, most of which are gastrointestinal anomalies such as jejunal atresia or ileal atresia, which are caused by vascular injury to the bowel. Furthermore, left-sided gastroschisis occurs

Table 1. Summary of Cases of Left-sided Gastroschisis from the Literature

Case	Year	References	Sex	Gestational age (wks)	Birth weight (g)	Associated anomalies
1	1988	Blair et al.	M	-	-	None
2	1989	Hirthler et al.	F	27	900	Hyaline membrane disease
3	1989	Hirthler et al.	M	Term	3,800	None
4	1993	Toth et al.	F	35	1,540	None
5	2000	Thepcharoennirund et al.	F	36	1,700	None
6	2000	Thepcharoennirund et al.	F	40	2,450	None
7	2001	Pringle KC	M	34	2,065	Left testis herniating through defect
8	2002	Fraser et al.	M	28	880	None
9	2002	Ashburn et al.	F	37	2,800	None
10	2004	Ameht et al.	M	Term	-	None
11	2004	Orpen et al.	F	Term	2,750	Pseudo-exstrophy, atrial septal defect, patent ductus arteriosus, and ureteral reflux
12	2004	Wang et al.		-	-	Situs inversus
13	2004	Yoshioka et al.	F	38	2,604	None
14	2004	Yoshioka et al.	F	34	1,700	Necrosis of the herniated bowel
15	2006	Gow et al.	F	39	2,815	None
16	2007	Prasun et al.	M	24 (terminated)		Multicystic renal dysplasia
17	2008	Suver et al.	F	34	3,100	Jejunal atresia, microcolon, absent corpus callosum, optic dysplasia, panhypopituitarism, and intestinal atresia
18	2008	Suver et al.	F	35	2,200	Cerebral arteriovenous malformations
19	2008	Suver et al.	F	34	2,200	Atrial septal defect and pulmonary valve stenosis
20	2010	Patel et al.	F	34	2,160	Small left colon syndrome
21	2013	Mandelia et al.	M	Term	2,260	Patent ductus arteriosus
22	2015	Hombalkar et al.	M	-	2,400	Cecal agenesis, short gut, malrotation

Data obtained from Mandelia A, et al. J Clin Diagn Res 2013;7:2300-2[4], Suver D, et al. Am J Surg 2008;195:663-6[5].

more frequently in female infants. Associated extraintestinal anomalies more commonly occur in the left-sided gastroschisis than in the classic right-sided gastroschisis⁵. The associated extraintestinal anomalies in patients with left-sided gastroschisis include genitourinary anomalies such as pseudo-exstrophy, multicystic renal dysplasia, and vesicoureteral reflux. The associated cardiac anomalies in these patients include patent ductus arteriosus, atrial septal defect, and pulmonary stenosis, which were also detected in our patient. Other anomalies include situs inversus, agenesis of corpus callosum, cerebral arteriovenous malformations, optic dysplasia, small left colon syndrome, microcolon, and intestinal atresia (Table 1).

The pathogenesis of gastroschisis remains controversial. The most favored theory is the premature or delayed involution of the right umbilical vein, which normally occurs during the first 6 weeks of gestation^{8,9}. The abnormalities in the involution of the right umbilical vein cause ischemia at the junction of the abdominal wall and umbilical cord. Therefore, gastroschisis generally occurs at the immediate right of the umbilical cord. Other theories that describe the pathogenesis of gastroschisis are as follows: i) delayed closure of the umbilical ring at the junction of the abdominal wall and umbilical cord causes the rupturing of the amniotic membrane at that junction¹⁰, and ii) breakdown of the omphalomesenteric artery that leads to the evisceration of bowels outside of the abdominal wall through that junction¹¹.

The pathogenesis of left-sided gastroschisis remains unclear to date. The suggested theories of its pathogenesis are as follows: i) premature regression of the left umbilical vein results in mesenchymal ischemia and necrosis on the left side of the umbilicus⁴; and ii) the yolk stalk is normally positioned to the right of midline, so the malpositioning of the yolk stalk to the left of midline can be due to the wall fold failure leading to left-sided defects⁵. Both genetic and environmental factors such as maternal exposure to smoking, cocaine, pseudoephedrine, analgesics, and irradiation are considered etiological factors^{12,13}. Feldkamp et al. reported that young mothers are more likely to deliver babies with gastroschisis¹⁴. In animal models, an increased incidence of ventral body wall closure defects was observed in offspring when their mothers were fed with a diet containing low protein and zinc, and were exposed to carbon monoxide and radiation. Various teratogens and certain genetic traits were also found to cause these defects¹³. Scientists do not yet know the genes responsible for the closure of ventral body wall in humans. Nevertheless, Beckwith-Wiedemann syndrome

is related to insulin-like growth factor 2 (IGF-II)¹³. Furthermore, Rieger syndrome is related to the *PITX2* gene¹⁵. Thoracoabdominal syndrome is related to the X chromosome¹⁶. However, we need to further evaluate the genes responsible for anterior abdominal wall defects.

In this report, we describe an infant born with a left-sided gastroschisis. This case was complicated by cardiac anomalies. We discussed the suggested pathogenesis of left-sided gastroschisis and presented a review of literatures related to the same. We recommend evaluation for the possible presence of associated anomalies in patients with left-sided gastroschisis because extraintestinal anomalies in the genitourinary, cardiovascular, or central nervous system are more common in left-sided gastroschisis than in the classic right-sided gastroschisis.

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