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Case Report

Whirl Pattern Complex Gastroschisis

Andrés Felipe Rubio-Duarte[1], Mónica Beltrán-Avendaño[2], Daniella Chacón-Valenzuela[3]

1.Hospital Internacional de Colombia, Colombia
2.Obstetrician-gynecologist, Maternal-Fetal Medicin specialist, Faculty of Health Sciences, Universidad Autónoma de Bucaramanga, Colombia
3.Pediatric surgeon, Faculty of Health Sciences, Universidad Industrial de Santander, Colombia

ABSTRACT

Background: Complex gastroschisis is a rare variant characterized by intrauterine closure of the abdominal defect, which can be accompanied by atresia, necrosis, and, in most cases, lead to short bowel syndrome. This is the first case of gastroschisis presenting as a whirl pattern of loops and raises suspicion of intestinal ischemia and atresia.

Case Report: A newborn at 38 weeks of gestation with a prenatal diagnosis of complex gastroschisis presented with a whirl pattern of gastroschisis and jejunal atresia. The newborn was treated with staged closure of the defect and underwent a second surgical procedure for correction of jejunal atresia, with a successful outcome observed during 2 years of follow-up.

Conclusion: Whirl pattern gastroschisis is a rare presentation of complex gastroschisis, and its diagnosis can be made prenatally, allowing for the preparation of medical and surgical management at birth. It is necessary to actively search for the presence of intestinal atresia or ischemia in this type of presentation.

Keywords: Congenital abnormalities, Gastroschisis, Newborn

Introduction

Gastroschisis is a congenital defect of the abdominal wall, typically occurring on the right side of the umbilical cord, characterized by the evisceration of the intestines and other intra-abdominal organs without the covering of an amniotic membrane (1). It has a prevalence of 3-4 per 10,000 pregnancies in the general population (2,3). There is a higher predisposition among teenage mothers, underweight mothers, and those with malnutrition, a history of consuming acetylsalicylic acid, ibuprofen, and psychoactive substances during pregnancy (4,5). The etiology of gastroschisis is unknown, and its embryological origin is associated with the failure of migration of the ventral lateral fold, primarily on the right side, during early gestational age; other previously proposed hypotheses include abnormal regression of the right umbilical vein and disruption of the vitelline artery (6,7).

There are two types of gastroschisis based on intestinal involvement: Simple and Complex. Complex gastroschisis (CG) is characterized by perforation, necrosis, volvulus, and/or intestinal atresia, accounting for 17% of all cases (8). CG in a whirl pattern is a rare variant characterized by partial in-utero closure of the abdominal defect, which may be accompanied by atresia, necrosis, and, in most cases, lead to short bowel syndrome (8,9). Prenatal diagnosis and postnatal management of closed gastroschisis remain challenging for specialists in maternal-fetal medicine, obstetrics, neonatology, and pediatric surgery. Thanks to advancements in technology, prenatal diagnosis is becoming increasingly common, enabling better medical and surgical

* *Corresponding author*: Andrés Felipe Rubio-Duarte, Hospital Internacional de Colombia, Colombia. Tel: 3213173654, Email: arubio888@unab.edu.co

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approaches at birth (10). The presentation of this case was approved by the Ethics Committee of the University Hospital of Santander.

Case report

An 18-year-old primigravida adolescent from a rural area, with a low educational level, presented with a pregnancy at 38.1 weeks of gestation. She had no previous pregnancies, and during this pregnancy, she had no obstetric complications, did not use tobacco or alcohol, and had inadequate prenatal care, with only one antenatal ultrasound performed. Antenatal ultrasound findings revealed a 4.5cm diameter defect in the left lateral abdominal wall, near the base of the umbilical cord, with protrusion of intestinal loops consistent with gastroschisis. Two ultrasound patterns were observed in the dilated extrabdominal intestinal loops: the first pattern corresponded to a dilated sentinel loop that allowed differentiation of the lumen and wall, and the second pattern was characterized by enlarged intestinal loops with a whirl pattern distribution, occupied by hyperechoic material that made it difficult to define their structure. Additionally, dilated intrabdominal intestinal loops with a larger diameter lumen of 18mm, wall edema, sediment inside without evidence of ascites, and reticulated appearance of the amniotic fluid were observed. These findings were indicative of complex closed gastroschisis (Figure 1). Fetal well-being tests and prenatal laboratory studies remained within normal parameters throughout the pregnancy.

During a cesarean section delivery, a male newborn weighing 2295 grams demonstrated appropriate neonatal adaptation, without the need for hemodynamic or ventilatory support measures. The newborn was transferred to the operating room for surgical intervention. Intraoperative findings included a 14mm diameter defect in the right abdominal wall with a whirl pattern of thin intestinal loops (Figure 2A)., without signs of intestinal necrosis, but with serositis that hindered proper differentiation of the loops, interloop adhesions, and the surrounding skin (Figure 2B). Under general anesthesia, adhesions were released, and the defect was closed in stages using an ABS-SILO system (11).

The newborn was immediately transferred to the neonatal intensive care unit (NICU) and placed on invasive mechanical ventilation. Complete closure of the defect was achieved on the 4th day of life, and ventilation support was successfully discontinued 12 hours after closure. Enteral nutrition was initiated on the 6th day but was not tolerated adequately, and a greenish drainage of 0.6cc/kg/24h was observed through a nasogastric tube. On the 10th day of life, under suspicion of possible atresia, a contrast-enhanced intestinal transit radiography was performed, revealing the absence of contrast passage from the first half of the jejunum. Surgical intervention was scheduled, and a Grosfeld type II jejunal atresia (12) was identified and corrected by resecting a 5-centimeter long atretic segment and performing an end-to-end intestinal anastomosis. The newborn had a satisfactory outcome in the NICU, was discharged at one month of age, and had uncomplicated follow-up for 2 years.

Ethical approval

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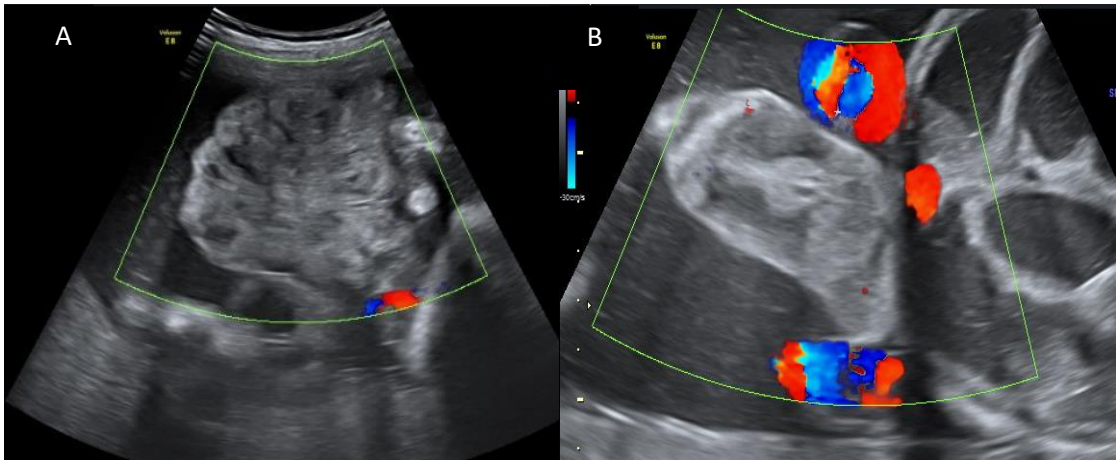

Figure 1. Prenatal ultrasound findings

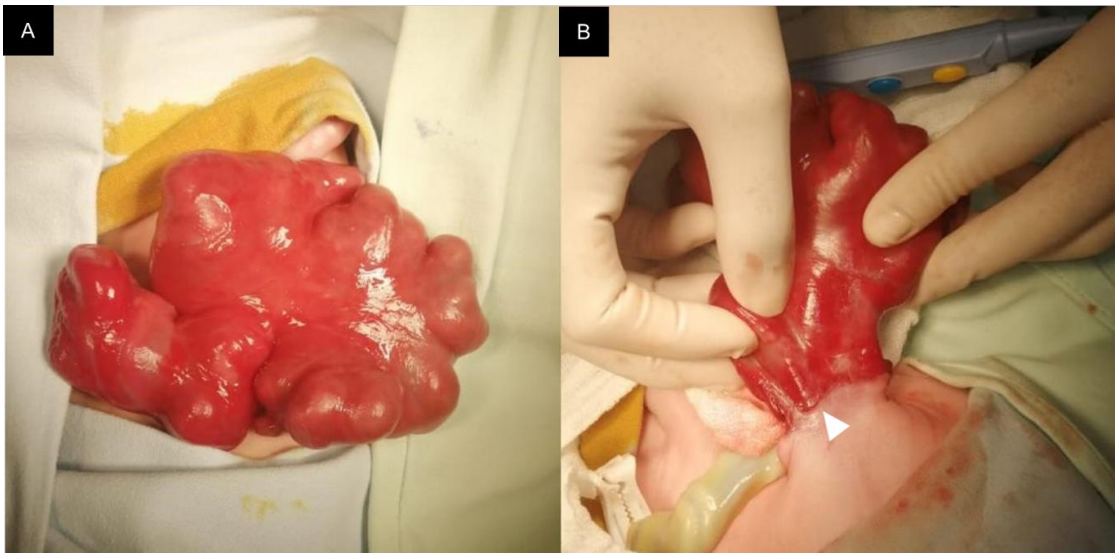

Figure 2. Postnatal intraoperative findings

Discussion

In CG there is premature partial or complete closure of the abdominal defect, leading to ischemia and hypoperfusion of the intestinal loops present in the defect. There is a direct relationship between the severity of ischemia extension and intestinal atresia (10). Evanescent bowel is a rare complication of closed CG, where the ischemic extrabdominal segments are reabsorbed in utero, and the intraabdominal portion of the intestine is reduced, resulting in increased morbidity secondary to short bowel syndrome (13). The ultrasound diagnosis of gastroschisis can be made as early as 12 weeks of gestation. In the follow-up ultrasound evaluation of patients, the possibility of a diagnosis of complex CG should be considered when intrabdominal intestinal loop dilation with hypoechoic appearance is present (14, 15). The severity of sentinel loop edema does not correlate with the presence of complex CG or successful primary correction (16). Short bowel syndrome, present in up to 27% of cases, is a common complication of complex CG (8). The prognosis is related to multiple factors such as prematurity, the presence of other congenital anomalies like cardiac defects, residual intestinal length, and therapeutic interventions such as transplantation and intestinal lengthening procedures (17, 18). Mortality in CG is 7 times higher compared to simple gastroschisis (8). There are no described risk factors for the presence of complex gastroschisis. The mother of our patient was an adolescent, primigravida, and with a low level of education, all of which are previously described risk factors for gastroschisis in general.

The selection of the appropriate timing for pregnancy termination includes several elements such as defect characteristics, severity of edema and dilation of the sentinel loop, reductibility index, and amniotic fluid characteristics (19). The mode of delivery does not change the prognosis; however, scheduled cesarean section allows for interdisciplinary coordination, ensuring timely, efficient, and appropriate care of these cases (20), in the reported case, a term cesarean section was performed, in coordination with the pediatric surgery and neonatology team, for immediate postnatal management.

The postnatal management of newborns with CG includes patient stabilization and subsequent surgical correction of the defect (21). Initial measures include intestinal decompression with orogastric and rectal tubes, coverage of the intestinal loops with plastic material and/or moist dressings, avoiding the use of dry gauze and dressings. Attempts are made to correct the defect by fully introducing the intestinal loops into the abdomen while evaluating intra-abdominal pressure (1, 22).

The presence of signs of intrabdominal hypertension, such as elevated mean airway pressure, poor general condition, and absence of pulses in the lower limbs, are indicators for delayed closure of the defect, using an ABS-SILO system that allows for the slow introduction of intestinal loops into the abdomen in the days following birth, with an average time of 5 days to achieve complete reduction (1, 11, 23), our case, it took 4 days until successful closure was achieved.

Postoperative management of newborns with CG is carried out in the neonatal intensive care unit (NICU) as most cases require mechanical ventilation.

Postoperative care includes orogastric tube drainage, withholding enteral feeding, and continuous monitoring of vital parameters (21, 24). The initiation of enteral nutrition should be individualized, and there is no standard time for initiation. Enteral nutrition should be gradually increased until achieving complete nutrition through this route (21).

When there is persistent and increasing drainage through the orogastric tube, along with oral intolerance, the presence of intestinal atresia should be suspected. Contrast-enhanced hydro-soluble intestinal transit study is the indicated radiological examination to confirm this diagnosis, although it is not routinely indicated in all newborns with gastroschisis (25). In case of confirmed intestinal atresia, as seen in some cases of CG, correction is recommended within the first 6 weeks after birth (26).

Surgical intervention for intestinal atresia is not an urgent procedure. The newborn should undergo surgery in the best nutritional and hemodynamic conditions, with adequate total parenteral nutrition support and minimal hemodynamic support. Adequate preoperative management increases the likelihood of successful primary repair of the defect, avoiding the need for intestinal diversion, which increases morbidity and the need for subsequent surgical interventions (19). The surgical approach depends on the type, location, and extent of the atretic segment, and can be performed through laparotomy or laparoscopy-assisted techniques. In this particular case, surgery was performed at 10 days of life via laparotomy, involving resection and primary anastomosis without the need for intestinal diversion. This did not result in short bowel syndrome, as observed in reported cases of other types of complex gastroschisis, thereby contributing to postoperative recovery. The prognosis of newborns with CG mainly depends on the length of viable intestine within the abdomen (10, 21). Lengthening procedures may be necessary when there is nutritional deficit secondary to short bowel syndrome.

The management of CG requires a multidisciplinary approach involving maternal-fetal medicine, obstetrics, pediatrics, neonatology, and pediatric surgery. Prenatal diagnosis allows for the planning of postnatal management in these cases. The optimal timing for the termination of pregnancy in closed CG cases is still unclear. Prematurity negatively impacts the prognosis. A thorough understanding of the disease spectrum is required to enable individualized management for each case. Follow-up ultrasound evaluations and multidisciplinary teamwork are necessary to achieve the best possible perinatal outcome. As the first reported case in the literature, comparability is not feasible. It is essential to note that this presentation of gastroschisis may occur when prenatal findings include dilated loops with a whirl image. The treatment was successful, and the presence of intestinal atresia should be considered, requiring subsequent surgical management.

Conclusion

Whirl pattern gastroschisis is a rare variant of complex gastroschisis. It should be considered when prenatal ultrasound findings show a cluster of intestinal loops. Intestinal atresia may be present, and therefore, it should be ruled out after the correction of the defect.

Acknowledgments

None.

Conflicts of interest

The authors declared no conflict of interest.

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