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Closing Mirror Image Left-sided Gastroschisis with Vanishing Splenogonadal Fusion and Left Testis

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Abstract

Gastroschisis is a para umbilical defect of the anterior abdominal wall without any peritoneal sac, which is nearly always located to the right of the umbilicus. We present a baby boy with a closing mirror image of left-sided gastroschisis, which had a very rare and interesting association with vanishing splenogonadal fusion and left testis with blind ending epididymis and the ductus deferens. This was successfully managed by primary closure without any anesthesia, sutures, or formal silastic silo as the bedside procedure and had an uneventful recovery.

Keywords: Abdominal wall defect, antenatal diagnosis, birth defects, closing gastroschisis, gastrointestinal malformation, left-sided gastroschisis, premature ganglion cells, splenogonadal fusion, testicular atrophy, vanishing testis, vas deferens.

Introduction

Our team has extensive experience in prenatal diagnosis **[1]**, fetal surgery **[2,3]**, and various forms of gastroschisis **[4,5,6]**, but we have never come across such an exciting case with several issues and implications with regard to the diagnosis and subsequent therapeutic interventions. Gastroschisis is a common congenital central abdominal-wall paramedian defect, which typically occurs to the right of the umbilicus. It is generally small, without a membrane, an isolated defect through which the bowel eviscerates. Although splenogonadal fusion has been reported as an isolated anomaly, its association with gastroschisis has not been reported **[7]**. There are only a few cases of left-sided gastroschisis that have been reported in the world literature. We wish to add one more case of left-sided gastroschisis that had associated abnormalities of the vanishing splenogonadal fusion and loss of the left testis with blind-ending epididymis and vas deferens.

Case Report

A baby boy weighing 2600g was delivered at 36+5/40-weeks of gestation by emergency cesarean section because of increasing fetal distress. Mother was primigravida, 16 years of age. Antenatal scans detected gastroschisis at anomaly scan and were monitored throughout the pregnancy at scheduled intervals till had fetal distress was experienced. The patient was born in good condition with normal APGAR scores and had stable vital signs. The patient was born at the Maternity and Child Health (MCH) hospital, which is part of our university teaching hospitals in Rajkot, Gujarat. India (the original name of ‘Rasul Khanji Zanana Hospital’ established by the erstwhile Nawab of Junagadh, Rasul Khanji, in 1897 — has now been rebuilt at a cost of Rs 100 crore and the biggest government facility with 700 beds for treatment). The patient was transferred to the neonatal intensive care unit at the K T Children Government University Teaching Hospital next door. The clinical examination showed a small-sized bowel evisceration through the defect situated on the left of the umbilicus with adhesion on the supralateral and inferolateral aspect of the defect **(Fig.1)**

Fig.1 At presentation. A Right side is clear and normal. B. Left-sided full-thickness defect without sac-note adhesions on the supralateral and inferolateral aspects of the defect.

The left scrotum was hypoplastic, empty with no palpable testis with no palpable cord structures in the left groin. The heart sounds were on the left chest, and there was no evidence of situs inversus. The abdominal cavity was relatively well developed with minimal viscero-abdominal disproportion.

All laboratory investigations, including the complete blood count, biomedical profile, and coagulation screen, were normal. Babygram showed normal chest and stomach gas shadow in the upper abdomen with a nasogastric tube in place. A 25 ml warm normal saline rectal washout yielded a meconium plug decompressing rectosigmoid, and a paracetamol suppository was placed for analgesia.

After resuscitation and stabilization of the baby, as the defect was small and the evisceration was reasonable and without too much of the peel, an initial trial of reduction without anesthesia, silo, or primary surgical closure under anesthesia was attempted in the procedure room of our neonatal intensive care unit under strict aseptic conditions. The eviscerated organs included small and large bowel loops, which were neither thickened nor oedematous. The adhesions on both lateral areas of the defect were gently broken. To our surprise, the inferolateral adhesions showed vas and epididymis parts seen to be blind ending, going down towards the iliac vessels, which were excised and sent for histological analysis. The supralateral adhesion on the inner peritoneal side attached bids on a string appearance of lymph nodes-like structures - scattered small spenenculi getting attached to the lower pole of the spleen, which was excised and sent for histopathology.

The eviscerated bowel could be reduced into the abdominal cavity successfully with ease and no tension at all. The defect was 4 cm long and located to the left of the umbilicus. A decision to perform sutureless primary closure similar to the silastic silo final stage closure procedure using the umbilical cord stump and the fixing dressing from the silastic silo kit, leaving a natural umbilical scar. A percutaneous long central line was inserted through the elbow under aseptic conditions and total parental nutrition was started.

Histological findings confirmed a vas deferens with epididymis caudally and a splenogonadal fusion of the intermittent beads type cranially. Postoperatively, the patient had constipation and delayed progression of feeds, and a rectal suction biopsy showed premature ganglion cells. The patient received total parenteral nutrition for twenty-nine days and was discharged home after thirty-seven days in hospital. At 1 year follow-up, the patient remained asymptomatic and is thriving well with normal gastrointestinal function and an almost invisible scar **(Fig. 2)**.

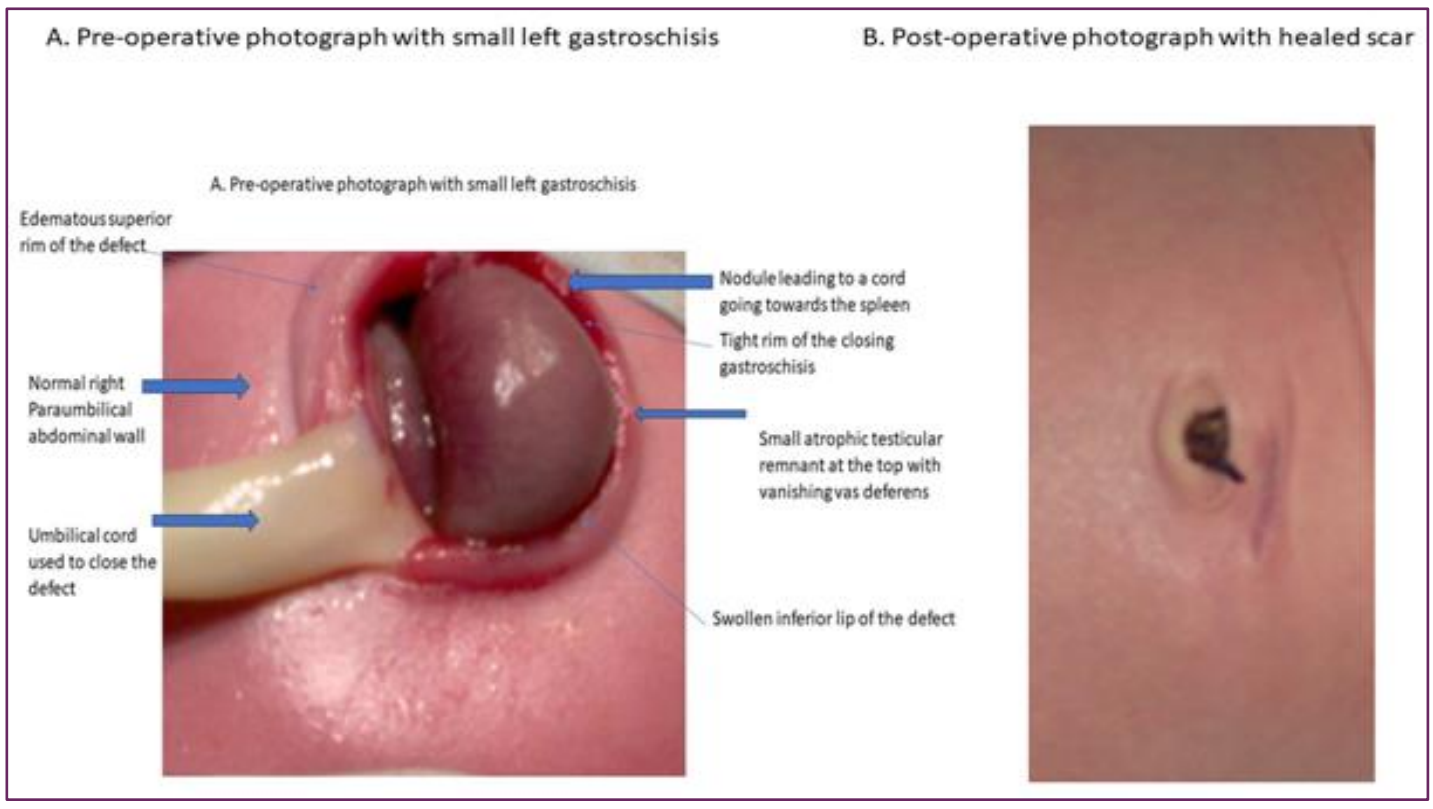

Fig. 2 A. Pre-operative status B. Follow-up photograph-note nicely healed suture, less scar, and slightly asymmetrical umbilicus due to wound contraction following healing on the left side.

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Discussion

In the past, animal models of gastroschisis have been associated with high mortality and morbidity with variable degrees of intestinal evisceration. We, therefore, developed a modern, innovative fetal surgical technique, an improved model of gastroschisis in fetal rabbits was developed by our team **[2]**. Widely held belief or surgical dogma is that any weakness or defect in the abdominal wall is responsible for any herniation of abdominal contents and viscera, which was our hypothesis. To our surprise, both partial thickness and full thickness abdominal wall defects did not bring the herniation of the bowel, negating our hypothesis on one hand and showing the fetal healing capacity of scarless healing instead. When the bowels were eviscerated, it mimicked the clinical model with classic clinical changes to the bowel. This allowed us to start lateral thinking outside the usual box, which brings raised intraabdominal and intraluminal pressure, ultimately leading to the evisceration of intestines through the most vulnerable weak point in the right paramedian region of the adnominal wall **[3]**.

In most patients with gastroschisis, the abdominal wall defect occurs on the right side of the umbilicus. Occasionally, a mirror image variation on the left side is reported, and recently, a case of combined left-sided gastroschisis and exomphalos has been reported **[8]**. Left-sided gastroschisis is more common in females and associated with a higher incidence of extraintestinal anomalies compared with right-sided lesions. Although the etiology remains unknown, it may differ from that of right-sided gastroschisis.

There are two types of left-sided gastroschisis: the mirror image variety, which is similar to right-sided lesions, and the left-sided abdominal wall defect in the left upper quadrant or flank distant from an intact umbilicus, which is a more severe defect often associated with more congenital anomalies. Approximately half of the 31 total cases of both varieties of left-sided gastroschisis reported in the literature describe other associated anomalies **[8]**. This left-sided condition is more associated with other congenital anomalies than its counterpart on the right side, with a higher occurrence of facial clefting **[9]**. Recently, more cases of gastroschisis have been reported in a pathology museum, in association with the persistent right vitelline vein, cecal agenesis, multiple intestinal atresia, and Meckel's diverticulum **[10, 11, 12, 13, 14]**.

The first case of a mirror image or discordant left and right gastroschisis in monochorionic twins has been reported in the literature, which may shed further light on the pathogenesis of gastroschisis **[15]**. The embryoetiogenesis of gastroschisis remains controversial and largely unproven. Multiple theories have been proposed, but none have ever been supported by a thorough embryological study.

A recent comprehensive developmental mechanism of development has been studied in embryos and fetuses by anatomical and microscopic observations of the developing abdominal wall and cord of embryos and fetuses, along with clinical features of gastroschisis. Gastroschisis could be the result of amniotic damage, possibly from some unidentified toxin. Further bowel damage can be explained by the subsequent mesenteric injury. Section of placental membranes revealed vacuolization of the amnion **[16]**. Without increased macrophage infiltration of the chorion in left-sided gastroschisis **[17]**.

The evisceration of the stomach and the small and large intestines is common, and evisceration of the gall bladder, urinary bladder, uterus, ovary, and testis have all been reported. All cases could be closed primarily, but one case required staged closure. Situs inversus, pubic diastasis, bifid clitoris, double vagina, anteriorly placed anus, atrial/ventricular septal defect, patent ductus arteriosus, choledochal cyst, cerebral arterio-venous malformation, macrocephaly, scoliosis and stenosis of superior vena cava have all been variously reported **[18,19,20,21]**.

Although multiple hypotheses have been proposed, the embryogenesis of gastroschisis is controversial and largely unproven. Our cases with small, left-sided defects associated with vanishing splenogonadal fusion and multiple spenenculi, in addition to vas deferens and testicular pathology, certainly add to the spectrum of gastroschisis, but at the same time, they challenge the existing theories.

The management of left-sided gastroschisis is similar to the right-sided lesions, and the option is a wide spectrum ranging from bedside closure, silastic silo delayed primary closure, surgical primary closure, or delayed operative silo closure in most cases of simply isolated gastroschisis. In closing, gastroschisis with significant bowel loss may need corrective bowel strengthening procedures if a midgut volvulus with complete loss of small bowel may need even transplantation. In newborns with isolated gastroschisis, the overall prognosis is very good. In rare cases, gastroschisis is associated with other congenital malformations, making the prognosis worse such as associated with meromelia of the limbs **[22]**.

Conclusion

In conclusion, we believe that this hitherto unreported observation alerts us to the need to examine the baby and all the adhesions with the surrounding structures like a string of multiple spenenculi and any abnormal findings, especially with absent testis and findings of blind-ending epididymis and vas deferens in male neonates undergoing repair of left-sided gastroschisis. Findings in our case certainly shed new light on the possible pathogenesis of this congenital anomaly and its natural course of the disease The case has value in extending the observations that even a very small or minimal eviscerated gastroschisis with minimal or no peel on these structures, which can be closed without any anesthesia and surgical procedure, there is no correlation or any good prospects of recovery of the bowel function quickly as it takes longer. We believe that the premature ganglion cells, in association with injury to the bowel by amniotic fluid irritation and the closing gastroschisis reducing the blood supply, have led to the delay in functional bowel recovery. Identifying and reporting these unusual cases is essential to further expand our understanding of these conditions and their potential associations. These cases illustrate the anatomic spectrum of gastroschisis as well as support intrauterine spontaneous closure of the abdominal wall defect as an etiology for midgut atresia.

Compliance with ethical standards:

Acknowledgments

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Conflict of interest:

The authors have no conflict of interest to declare. No funding source was involved in this study.

Ethical approval:

All procedures performed on human participants were in accordance with the ethical standards of the institutional and national research committee and with the 1964 Declaration of Helsinki and its later amendments or comparable ethical standards.

Informed consent:

Informed consent was obtained from the parents and all the relatives involved prior to all the procedures. Parents and all involved parties were informed about the procedure**.**

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