Case Series

Submitted: 08-10-2020
Accepted: 23-12-2020

DOI: https://doi.org/10.47338/jns.v10.932

Vanishing gastroschisis with jejunal atresia and extreme short bowel syndrome: A case series

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KEYWORDS

Vanishing gastroschisis, Extreme short bowel syndrome, Jejunal atresia

ABSTRACT

Background: Vanishing gastroschisis may occur due to spontaneous partial or complete closure of anterior abdominal wall defect around the viscera, leading to small bowel ischemia and resultant entry/exit level atresia and extremely short length of the remaining bowel. The prognosis is very poor, even after aggressive surgery, and requires prolonged total parenteral nutrition.

Case Series: We report two female neonates, one with closed and another with closing vanishing gastroschisis, associated with jejunal atresia and extreme short bowel syndrome. In both patients, the antenatal scans showed gastroschisis without the evidence of vanishing gastroschisis. In both neonates, palliative surgeries were done. Both patients died after a few days due to short bowel syndrome and sepsis.

Conclusion: When antenatally detected gastroschisis presents with closed or closing anterior abdominal wall defect, (vanishing gastroschisis), the parents/caregivers must be counseled about the poor prognosis of this condition. A tailored approach to either palliation or aggressive therapy is essential in this rare condition.

INTRODUCTION

Gastroschisis is characterized by the herniation of the midgut and occasionally other abdominal viscera through an anterior abdominal wall defect (AAWD), on the right of the umbilicus insertion, into the amniotic sac.[1,2] There has been an increase in the incidence of gastroschisis from 2.5 to 4.4 per 10000 live births over the past few decades.[3] Several hypotheses of etiology have been put forward, but the recent dual vascular/thrombotic model better explained it.[4] Herein, we present two variants of vanishing gastroschisis in neonates detected postnatally, both being associated with jejunal atresia and extreme short bowel syndrome.

CASE SERIES

Case 1: A 2-day-old female neonate was brought to our institute with bilious vomiting. On clinical examination, there was abdominal distension with visible palpable few loops, and a normal umbilicus. She was born to a 20-year-old primigravida. There was a history of antenatal diagnosis of gastroschisis. On analysis of her antenatal scans, the first obstetric scan at 16 weeks of gestation showed bowel herniation through an abdominal wall defect without any covering sac (Fig.1A,1B). Fetal anomaly scan at 20 weeks gestation was indifferent, but the 26-week scan showed additional findings of early-onset fetal growth restriction, normal amniotic fluid volume, and three vessels in the cord. Mother had labor pains at 32 weeks, and spontaneously delivered a female premature baby of 1.95kg at home. At birth, there was no visible abdominal wall defect, but the baby developed bilious vomiting and feeding intolerance on day one and referred to us by the pediatrician on day 2. Plain X-ray abdomen showed markedly dilated small bowel loops with air-fluid levels suggestive of obstruction (Fig.1C). The baby was resuscitated and optimized. On exploratory laparotomy, there was about 25 cm of remaining markedly dilated small bowel from gastroduodenal junction to jejunal atresia (Type 1) and colon was about 15 cm. Most of the midgut structures including part of the jejunum, ileum, cecum, appen-

dix, and most of the colon vanished (Fig.1D). A jejunostomy and distal colonic mucus fistula were formed after discussion of findings with the parents. On postoperative day 2, the stoma started functioning and feeding was started. After counseling, the parents choose palliative care at home. The patient died on the 25th day after surgery at home without any hospital visit.

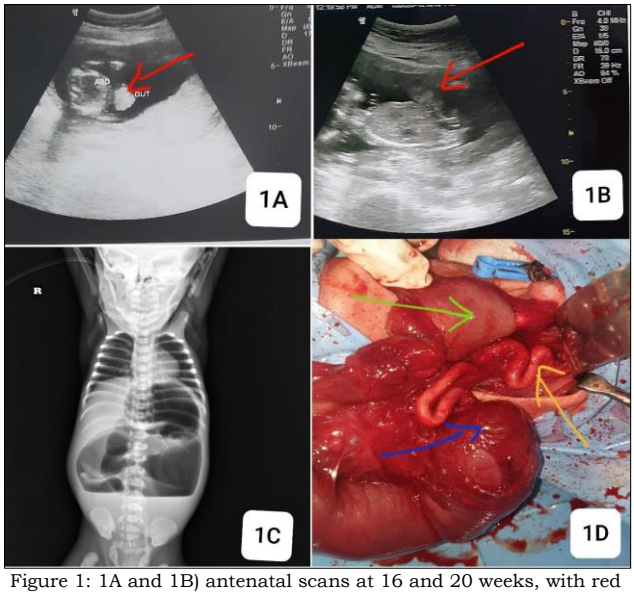

Figure 1: 1A and 1B) antenatal scans at 16 and 20 weeks, with red arrow showing gastroschisis. 1C) plain abdominal erect X-ray with dilated small bowel and 3 air-fluid levels. 1D) intraoperative finding of dilated stomach (green arrow), jejunal atresia (blue arrow), and short length atretic colon (yellow arrow).

Case 2: A 38-year-old primigravida mother delivered a female premature baby at a hospital setting by spontaneous vaginal delivery (weighing 1.5 kg at 33 weeks of gestation). After delivery, the baby was shifted to the Pediatric Surgical unit with bilious vomiting and a blackish mass visible at the central abdomen on day 1 of life. Antenatal scan at 18 weeks of gestation, showed bowel herniation through an abdominal wall defect without any covering sac (Fig.2A,2B). The karyotyping report was 46, XX. Antenatal scans were done on the 20th and 22nd weeks were indifferent. But a 26 weeks scan showed fetal growth restriction (FGR) below 1st centile with normal amniotic fluid volume, and three vessels in the cord. On clinical examination, there were visible gangrenous bowel loops protruding through a very small abdominal wall defect a cm right and above the umbilicus insertion with a gelatinous layer at the base (Fig 2C). Abdominal ultrasound and X-ray showed markedly dilated small bowel loops and very narrow abdominal wall defect of 7mm suggestive of obstruction with gastroschisis (Fig.2D). On exploratory laparotomy, there was dense adhesion between the peritoneum and protruding ends of the small bowel and colon at the narrow defect. The distal end of the small bowel and proximal end of the colon were atretic (Fig.2E). There was only about 20 cm remaining small bowel from the gastroduodenal junction to complete jejunal atresia and the colon was nearly 12 cm. Most of the midgut structures including part of jejunum, ileum, cecum, appendix, and most of the colon were absent (Fig.2F). A jejuno-colic end to back anastomosis was performed and the abdomen was closed in layers. Histology of the specimen revealed ischemic bowel necrosis. On postoperative day 4, the patient died of septicaemic shock.

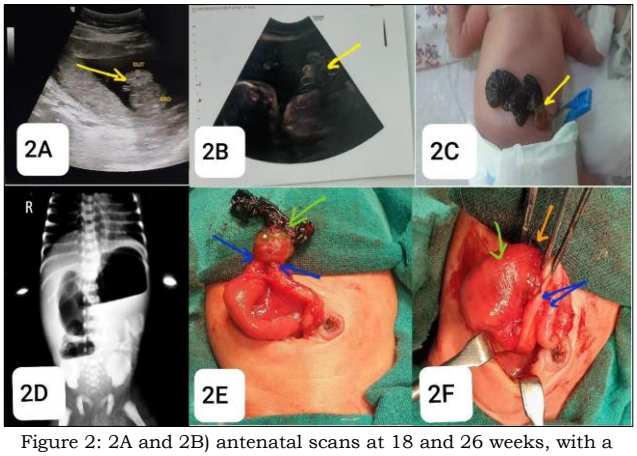

Figure 2: 2A and 2B) antenatal scans at 18 and 26 weeks, with a yellow arrow showing gastroschisis. 2C) gangrenous VGS after birth. 2D) plain abdominal X-ray with dilated small bowel. 2E) intraoperative finding of jejunal atresia and colonic atresia at entry/exit of GS (blue arrow) and vanished midgut (green arrow). 2F) the dilated jejunum (green arrow), short atretic colon (blue arrow), and an end to back anastomosis (orange arrow)

DISCUSSION

The hypotheses which could describe the finding of our patients are 1) Spontaneous partial or complete closure of anterior abdominal wall defect around the intestines and superior mesenteric artery, resulting in strangulation necrosis of the midgut; 2) incarceration and atrophy of protruding bowel at entry/exit with subsequent closure of the defect; 3) midgut volvulus at the narrow defect and resulting in gangrene.[5]

The incidence of vanishing gastroschisis is 4.5 to 6% of all gastroschisis.[6,7] Regarding various possible phenotypic presentation of closing/closed gastroschisis, at one end, there is a closed abdominal ring with viable viscera; on the other extreme, it would be intestinal atresia at abdominal ring or infarction of midgut, intestinal resorption (matted thick fibrotic)m and normal-appearing abdominal wall termed as vanishing midgut.[6,7] Kumar et al. classified phenotypically vanishing gastroschisis as Type I (vanishing gut with lumen), Type II (vanishing gut without lumen or nubbin of tissue), and Type III (antenatal evidence of gastroschisis and at the birth total absence of midgut).[8] Type III was the finding in case 1 neonate (closed VGS) whereas in case 2 it was of type II variety (Closing VGS).

In the present series, case 1 had spontaneous closure of antenatally diagnosed gastroschisis abdominal wall defect, whereas in case 2 there was closing gas-

troschisis with very narrow 7mm anterior abdominal wall defect, on the right side of the umbilicus. Spontaneous closure of gastroschisis defect in-utero was also observed by Barsoom et al.[9] Our cases had VGS with jejunal atresia and short atretic distal colon (extreme short bowel syndrome) which is extremely rare and rarely reported.[10,11]

Table 1: Survivors of vanishing gastroschisis

Author	Case	Year	AGIR	Organ transplant	Age at time of publication	Feeding	GA	BW	SBL
Kimble [12]	1	1999	Bianchi (14W)		12Y	oral	36	2.42	40
Barsoom MJ [9]	2	2000	Bianchi (5M)		8M	TPN	34	1.76	10
Ogunyemi [13]	3	2001	AGIR	Liver and Bowel (53m)	53M	oral	32	1.85	15
Winter [14]	4	2005	Bianchi	Bowel	32M	oral	35	ns	17
Sandy [15]	5	2006	STEP (30M)		40M	weaning TPN	35.5	2.5	<30
Vogler [6]	6	2008	AGIR		NS	oral	32-34	NS	53
	7		AGIR		NS	oral			62
	8		AGIR		NS	oral			170
	9		STEP		NS	oral			40
	10		AGIR		NS	oral			180
	11		AGIR		NS	oral			60
	12		STEP		NS	weaning TPN			23.5
Buluggiu [16]	13	2009	Bianchi (5M)		25M	oral	38	2.86	45
Houben [7]	14	2009	Bianchi 5W & 12W)	Liver 12m	11Y	oral	36	NS	NS
	15		Bianchi (6M)		15M	supplemental PN	35	NS	18
	16		AGIR		6Y	oral	35	NS	NS
Khalil [17]	17	2010	LILT 6M		2Y	oral	36	2.5	30
Lawther [18]	18	2010	STEP 9M		9M	supplemental PN	33	1.9	14.5
Dahl [19]	19	2011	AGIR		21M	oral	38	3.28	120
Kumar [8]	20	2013	STEP	Bowel 2Y	2Y	supplemental PN	37	3	30
	21		STEP	Transfer to a transplant center	NS	NS	33	2.12	20
	22		STEP	Transfer to a transplant center	NS	NS	35	2.23	8
Wood [20]	23	2013	Bianchi		3Y	oral	36	2.53	30
	24		Bianchi			supplemental PN	35	2.73	20
	25		Bianchi		8M	oral	37	2.56	20
	26		Bianchi			TPN	33	1.9	20
Abdel-Latif [21]	26	2017	AGIR		1M	ORAL	38	3	70
Sergi [4]	27	2018	AGIR		10Y	ORAL	34	2.17	76
Singh [22]	28	2018	AGIR		48M	Oral	36	2.2	50
Ponce [23]	29	2018	LILT 7M		7Y	supplemental PN	32	2.15	27
Abi Rached [24]	30		AGIR		3Y	supplemental PN	35	2.56	50

AGIR: autologous gastrointestinal reconstruction, GA: gestational age (weeks), BW: birth weight (KG), SBL: small bowel length distal to the duodenojejunal junction(cm), STEP: serial transverse enteroplasty Procedure, LILT: longitudinal intestinal lengthening and tapering, NS: not specified, W: weeks, M: months, Y: years.

In neonates with gastroschisis, survival has approached up to 90% due to advancements in surgical techniques and neonatal care. But the prognosis of vanishing gastroschisis is still very poor due to short bowel syndrome. Mortality is due to short bowel syndrome, central-line related sepsis, and in the long-run total parenteral nutrition-related liver failure.[6,7]

Vanishing gastroschisis usually needs an aggressive approach (Fig.3). The only surgical options available for patients with some residual bowel are bowel lengthening procedures such as the Bianchi procedure, serial transverse enteroplasty procedure (STEP), and longitudinal intestinal lengthening and tapering (LILT). For patients with very minimal bowel, the only surgical option can be a small bowel transplant.

All patients postoperatively require prolonged total parenteral nutrition (TPN), which can cause liver failure and may even require a liver transplant. Because of this, most of the parents usually choose palliative care plans and avoid aggressive surgical plans especially in developing countries.[11] Moreover, the outcomes of children after Bianchi surgery can be better when adequate bowel length is achieved. The 30 VGS survivors are summarized in Table 1. In this series also the parents choose palliative procedures, jejunostomy in case 1 and end to back jejuno-colic anastomosis in case 2 & refused bowel lengthening procedures. In this series, both patients succumbed to their illness.

In conclusion, the prognosis of complicated vanishing gastroschisis is still very poor with high morbidity and mortality. In-utero vascular accidents in gastroschisis lead to jejunal atresia or short bowel syndrome. Advancements in neonatal bowel transplants in the future may improve the survival of patients with short bowel syndrome.

Acknowledgements: Nil

Conflict of Interest: None

Source of Support: Nil

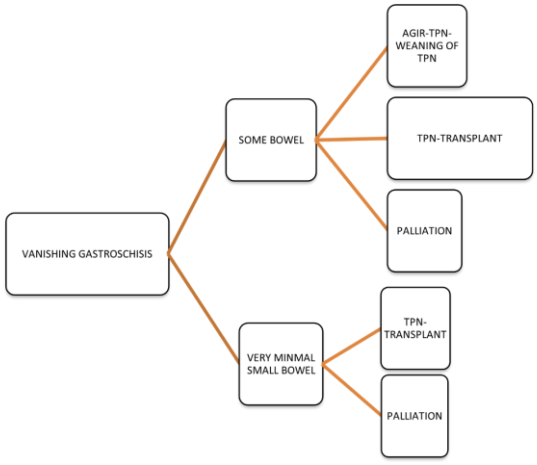

Figure 3: Approach for neonates with Vanishing Gastroschisis. (AGIR: autologous gastrointestinal reconstruction)

Consent to Publication: Author(s) declared taking informed written consent for the publication of clinical photographs/material (if any used), from the legal guardian of the patient with an understanding that every effort will be made to conceal the identity of the patient, however it cannot be guaranteed.

Author Contributions: Author(s) declared to fulfil authorship criteria as devised by ICMJE and approved the final version.

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