



Intrauterine intestinal volvulus prompting emergency delivery and surgery in a 32wk fetus

Kamalesh Pal ^{a,*}, Louay Hassan Zayed ^b, YounisMohamed Morsy Khalid ^c

^a Pediatric Surgery Department, Saudi German Hospital Dubai and Imam Abdulrehman Bin Faisal University, Saudi Arabia and Christian Medical College, India

^b Obstetrics & Gynaecology Department, Saudi German Hospital Dubai; and Alexandria University, Egypt

^c Neonatology division, Pediatrics Department, Saudi German Hospital, Dubai

ARTICLE INFO

Keywords:

Fetal volvulus
Ileal atresia
Mesenteric defect
Vascular accident
Bowel resorption

ABSTRACT

Fetal intestinal volvulus is a medical emergency with potentially fatal consequences thus requires a high index of suspicion and prompt intervention. Echogenic dilated bowels, Whirlpool/Coffee bean signs, polyhydramnios, fetal anemia, fetal ascites with aperistalsis, nonreassuring CTG are few important diagnostic signs. Mostly it has been described without associated malrotation, as segmental volvulus due to malfixation of gut with or without associated anomalies such as meconium ileus, CDH, abdominal wall defects. We describe for the first time a case of 1.9kg female fetus detected to have intestinal volvulus associated with multiple ileal atresias and mesenteric defect. The fetus had classical Whirlpool sign, polyhydramnios and anemia, was delivered by emergency Caesarian Section, post natively detected to have tense abdomen, confirmed Whirlpool sign on post natal USG, underwent emergency laparotomy, resection of necrotic ileal loops & I-C junction and Jejunocolic anastomosis. Child adapted to elemental feeding with 50cm of Jejunum without Ileocecal valve without shortbowel syndrome. Importance of multidisciplinary perinatal care, possible intrauterine vascular accident in pathogenesis of atresia and fetal volvulus has been emphasized in the case report.

1. Introduction

Fetal volvulus is one of the rare catastrophic bowel events affecting the viability of fetal gut and negatively affecting the fetal wellbeing, even causing fetal demise. There has been anecdotal case reports and only few case series [1] in the literature describing this entity. Mostly, segmental volvulus due to malfixation or mesenteric defect with or without associated anomalies such as meconium ileus, cystic fibrosis, duplication/mesenteric cyst, congenital diaphragmatic hernia (CDH), omphalocele/gastroschisis have been described causing limited necrosis of bowel. Rarely, malrotation of gut has been associated with complete fetal volvulus leading to loss of significant length of bowel. Although jejunal atresia has been described, association of multiple ileal atresias and mesenteric defect has been detected for the first time with fetal volvulus in our case. We have also noted an interesting finding of partial resorption of necrotic bowel in the fetus. Fetal mesenteric vascular accident has been purported as the common cause of atresias, volvulus and necrosis of bowel.

1.1. Case report

A 32wks female fetus weighing 1.9kg in a 30years old mother G3P1A1 was detected to have dilated, echogenic bowel with classical Whirlpool sign (Fig. 1), polyhydramnios and fetal anemia as evidenced by Doppler velocimetry of middle cerebral artery (1.6 MoM). No sign of intestinal perforation/meconium peritonitis or any other associated anomalies could be detected at fetal ultrasound. The diagnosis of fetal volvulus was made and parents were counselled for emergency delivery and early postnatal surgical intervention. With the parental consent, preterm fetus was delivered by emergency caesarian section after administering maternal steroid to stimulate fetal lung maturity. Post-delivery neonate was stabilised and confirmed to have tense distended abdomen, Whirlpool sign on postnatal USG as well confirming the fetal volvulus. Emergency laparotomy revealed necrotic bowel loops approximately 50cm distal to D-J junction due to segmental volvulus associated with multiple distal ileal atresias (three in number), mesenteric defect and fibrotic remnant of terminal bowel adjacent to the necrotic segment (Fig. 2–3). There was only 1cm of termi-

* Corresponding author. Saudi German Hospital Dubai, Dubai, PO BOX-391093, United Arab Emirates.

E-mail address: kamalesh_pal@yahoo.com (K. Pal).

<https://doi.org/10.1016/j.epsc.2020.101618>

Received 16 August 2020; Received in revised form 18 August 2020; Accepted 18 August 2020

Available online 21 August 2020

2213-5766/© 2020 The Authors.

Published by Elsevier Inc.

This is an open access article under the CC BY-NC-ND license

(<http://creativecommons.org/licenses/by-nc-nd/4.0/>).



Fig. 1. Whirlpool sign(Thick arrow) in Fetal Ultrasound.



Fig. 2. Segmental Volvulus with distal small bowel necrosis.

nal ileum viable near I-C junction. Resection of approximately 40cm of necrotic bowel, atretic segments, ileo-colic limited resection and ileo-colic anastomosis were done. Post operative recovery was smooth after initial bout of oliguria and acidosis. Child was started on low volume, drip in feed with elemental milk and gradually established breast milk feeding over a month period. Initial stool volume and consistency significantly got normalised with positive weight gain within this timeline and child was discharged on combination of breastmilk and elemental feed weighing 2.1kg. At 2months follow up child was active, playfull, accepting full volume breast feeding, passing 4–5 semiformed bowel motions and weighing 2.7kg.

2. Discussion

Fetal volvulus could be either complete or segmental. Complete fetal volvulus is extremely rare and involves twisting of the bowel from Duodeno-Jejunal junction(D-J) till ascending colon, around superior mesenteric vasculature due to malrotation of gut and narrow mesentery. Whereas, more common segmental fetal volvulus involves lesser extent of bowel and is usually associated with meconium ileus, atresia, mesenteric defect, omphalocele/gastroschisis, mesenteric/duplica-

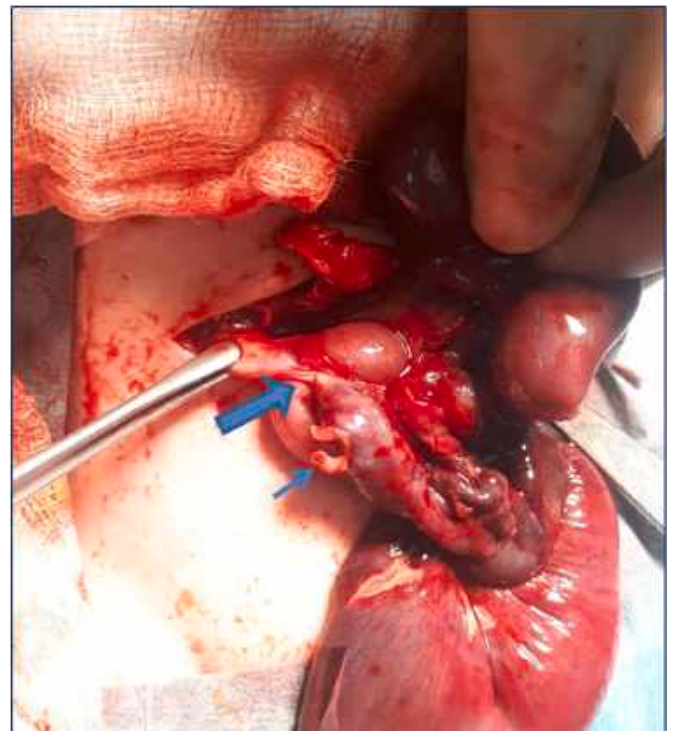


Fig. 3. Multiple ileal atresias (thick arrow) and partial resorption of bowel (thin arrow).

tion cyst or congenital diaphragmatic hernia.[1–3] It may also happen without any discernible bowel or mesenteric defects known as idiopathic fetal volvulus.(2,3). Although classically described dilated echogenic bowel with Whirlpool sign (spiral shaped mass made up of dilated bowel loops seen on ultrasound) or Coffee bean sign (dilatation of the small bowel with a thin outer layer composed of a single bowel wall layer and a thick inner wall due to double wall thickness of opposed bowel loop) [4], polyhydramnios and fetal anemia (due to hemorrhagic ascites/blood sequestration in the necrotic strangulated bowel detected by middle cerebral artery or umbilical artery velocimetry) clinches the diagnosis, there could also be other suggestive findings such as dilated bowel loops, gastric dilatation, fetal ascites, decreased fetal movements, and nonreassuring cardiotocography (CTG). [5]. Fetal volvulus, once suspected, is regarded a surgical

emergency from the obstetric as well as fetal point of view. If neglected or diagnosis is missed fetal volvulus could lead to perforation of bowel, severe hemorrhagic ascites, anemia, hypovolemia, pleural/pericardial effusion, cardiac failure and even fetal demise.[1],[6],[7]. Although the timing of intrauterine volvulus is not certain, however it has been reported to be detected as early as 15th week and as late as near term gestation (35 week).[8].

Most of the case reports have found segmental volvulus of small bowel without malrotation, possibly due to mal fixation of gut, mesenteric defect with or without associated anomalies like CDH, omphalocele, gastroschisis. Although there has been a rare instance of fetal volvulus associated with jejunal atresia in the literature [9], ours is the first case report of multiple distal ileal atresia and mesenteric defect causing fetal volvulus and necrosis of bowel. There was fibrotic strand of remnant of bowel adjacent to the necrotic segment suggesting partial resorption of dead bowel. Mesenteric defect with intrauterine vascular accident could be a plausible cause of these events as has been reported by Black et al. [10]. An inherent musculature deficiency has also been reported in fetal volvulus [11] but resorption of bowel has never been reported hitherto. Extent of bowel loss due to necrosis and/or resorption dictates the prognosis and development of short gut syndrome. Usually segmental volvulus cases do better than complete volvulus associated with malrotation. Our case had approximately 30% of proximal small bowel viable (50cm of Jejunum intact) and the newborn got adapted to elemental feeds in fairly short period of time attaining full breast feed within 45 post-natal days.

3. Conclusion

Fetal intestinal volvulus is a medical emergency which requires high index of suspicion by the antenatal assessment. Classical signs of bowel dilatation, echogenicity, aperistalsis, Whirlpool/Coffee bean signs, polyhydramnios, fetal ascites, fetal anemia, non reassuring CTG, progressive fetal pericardial/pleural effusion should help in detecting the entity. Mostly it is segmental in extent due to mal-fixation of gut, sometimes may be associated with meconium ileus, mesenteric defects, CDH, anterior abdominal wall defects and rarely with malrotation of gut. Multiple ileal atresias, mesenteric defect in our case emphasizes the role of vascular accident in the pathogenesis of this entity. High index of suspicion, prompt referral to tertiary care facility, accelerated delivery and emergency surgical intervention of the newborn is the standard of care to minimise fetal morbidity and potential fetal demise. Extent of bowel necrosis/absence due to atresia or resorption dictates the prognosis and development of short bowel syndrome.

Patient consent

Consent to publish the case report was not obtained. This report does not contain any personal information that could lead to the identification of the patient.

Funding

No funding or grant support obtained from any source.

Authorship.

All authors attest that they meet the current ICMJE criteria for Authorship.

Declaration of competing interest

The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

Acknowledgement

We sincerely thank Dr Naglaa Mehmoud Mohammed, Specialist Radiologist and Dr Talal Ali Mohammad, Consultant Obstetrics and Gynaecology at Saudi German Hospital, Ajman, UAE for initial diagnosis and prompt referral to Saudi German Hospital, Dubai for tertiary care.

References

- [1] Sciarone A, Teruzzi E, Pertusio A, et al. Fetal midgut volvulus: report of eight cases. *J Matern Fetal Neonatal Med* 2016;29:1322–7.
- [2] Durand M, Coste K, Martin A, et al. Fetal midgut volvulus as a sign for cystic fibrosis. *Prenat Diagn* 2008;28:973–4.
- [3] Kargl S, Wagner O, Pumberger W. "Volvulus without malposition—a single-center experience. *J Surg Res* 2015;193:295–9.
- [4] Jakhere SG, Saifi SA, Ranwaka AA. Fetal small bowel volvulus without malrotation: the whirlpool & coffee bean signs. *J Neonatal Perinat Med* 2014;7:143–6.
- [5] Noreldeen SA, Hodgett SG, Venkat-Raman N. Midgut volvulus with hemorrhagic ascites: a rare cause of fetal anemia. *Ultrasound Obstet Gynecol* 2008;31:352–4.
- [6] Tan RMR, Lee J, Biswas A, Amutha C. Ascites, anemia and (intestinal) atresia. *J Perinatol* 2014;34:78–80.
- [7] Steffensen TS, Gilbert-Barnes E, DeStefano KA, Kontopoulos EV. Midgut volvulus causing fetal demise in utero. *Fetal Pediatr Pathol* 2008;27:223–31.
- [8] Crisera CA, Ginsburg HB, Gittes GK. Fetal midgut volvulus presenting at term. *J Pediatr Surg* 1999;34:1280–1.
- [9] Baldassarre ME, Laneve A, Rizzo A, et al. A case of fetal midgut volvulus and jejunal atresia: nutritional support and maintenance of mucosal function and integrity. *Immunopharmacol Immunotoxicol* 2008;30:601–8.
- [10] Black PR, Mueller D, Crow J, Morris RC, Husain AN. Mesenteric defects as a cause of intestinal volvulus without malrotation and as the possible primary etiology of intestinal atresia. *J Pediatr Surg* 1994;29:1339–43.
- [11] Morikawa N, Namba S, Fujii Y, Sato Y, Fukuba K. Intrauterine volvulus without malrotation associated with segmental absence of small intestinal musculature. *J Pediatr Surg* 1999;34:1549–51.