Case Report

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A Rare Case of Jejunal Atresia Due to Intrauterine Intussusception

Surgery Section

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ABSTRACT

Intestinal atresia is generally caused by intrauterine vascular obstructions involving mesenteric vessels. Intrauterine intussusceptions (IUI) are one of these disruptive events. Intestinal intussusceptions affects children commonly between 3 months and 3 years of age, but it rarely affects in intrauterine life. The relationship between intrauterine intussusception and intestinal atresia has been demonstrated by few cases in literature, suggesting intrauterine intussusception as a rare cause of intestinal atresia. We report a 7-day-old full term neonate presenting with intrauterine intussusceptions (jejuno-jejunal) resulting in jejunal atresia.

Keywords: Intestinal atresia, Neonatal intestinal obstruction

CASE REPORT

A 2600 gm full term female neonate was born at VIMS Medical College Hospital Bellary, in the year 2015 by emergency lower segment caesarean section (indication was foetal distress) to a 16-year-old primigravida, out of non consanguinous marriage. The neonate presented to the paediatric emergency with respiratory distress and not passing meconium even after 24 hours of birth, except mucous per-rectally. Anus was normal, permeable without meconium stain on introduction of rectal tube.

On admission the baby was hypothermic, pale, with intercostal and subcostal retractions, S3 Gallop, without any abdominal distension. Birth asphyxia with respiratory distress was suspected.

On third day of life, grade-2 short systolic murmur was detected along with above mentioned features. Hence cardiac cause (congenital heart disease) was suspected. Due to non accessibility of paediatric cardiologist, echocardiography was deferred. On fifth day of life the baby developed bilious vomiting 12-15 episodes/24 hours in the absence of abdominal distension. On the sixth day baby developed abdominal distension along with persistent bilious aspirate. Abdominal radiography and ultrasonography showed dilated small bowel loops [Table/Fig-1].

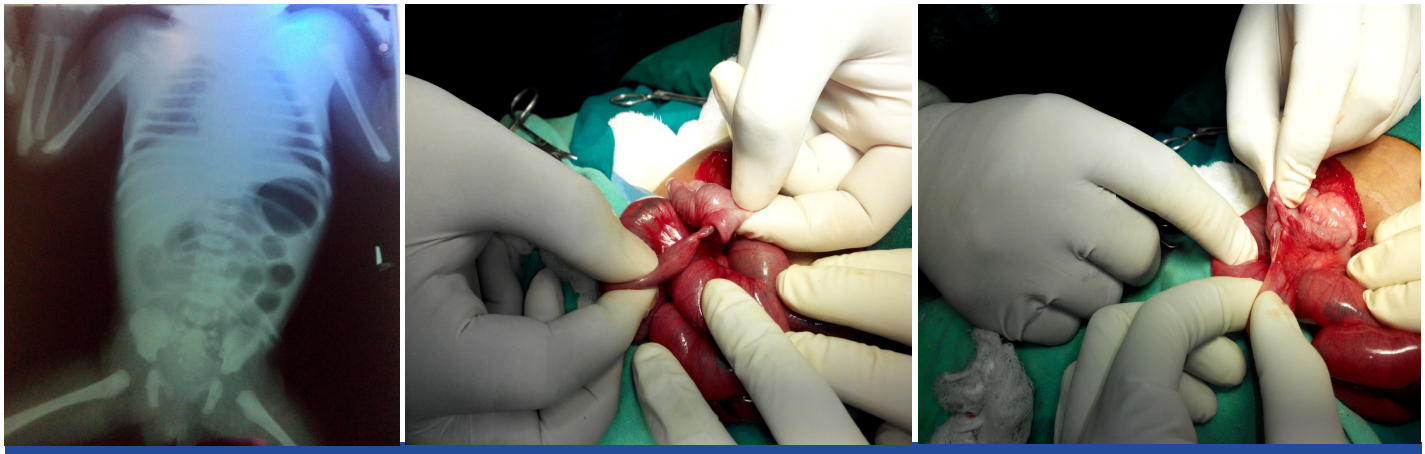
With the diagnosis of intestinal obstruction the parents were explained about the need for emergency surgery, and consent for the same was taken from the parents. The baby was explored after adequate resuscitation. On laparotomy dilated stomach and small bowel was noted involving duodenum and proximal jejunum with collapsed distal small bowel and large bowel. On careful examination of bowel, a 3 cm jejuno-jejunal intussusception was noted, about 15 cm distal to duodeno-jejunal junction [Table/Fig-2]. A Meckel's diverticulum was also noted, which was untouched. The jejunal intussusception was reduced, [Table/Fig-3] the proximal part shows atretic changes in the jejunum and had a small diverticulum with gangrenous changes at its tip [Table/Fig-4,5]. A 5 cm of small bowel from proximal part and 3 cm from distal part was resected and, end to back anastomosis was done in jejunum. The resected segment was sent for histopathological examination (HPE). HPE report showed, proximal segment had features of diverticulum with ischemic changes and distal segment had features of ischemic changes. Intraoperative and postoperative period was uneventful.

On the eighth postoperative day echocardiography was done and it revealed as moderate PDA (patent ductus arteriosis) along with large secondary ASD (atrial septal defect) left to right, with pulmonary arterial hypertension. The baby was put on oral digoxin and frusemide. The baby was discharged on 14th postoperative day and was advised for regular follow-up with cardiologist.

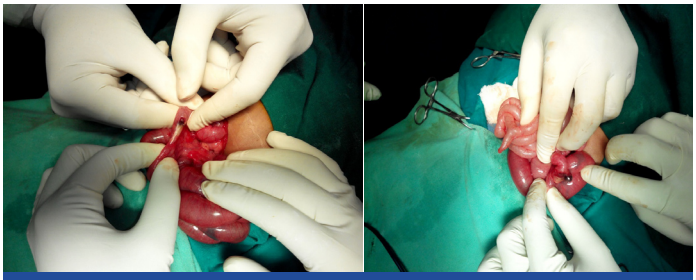
DISCUSSION

The aetiology of intestinal atresias is explained by two theories,

1. Luow-Bernard theory: Late intrauterine mesenteric vascular catastrophe leading to ischemic bowel damage resulting in



[Table/Fig-1]: X-ray abdomen showing dilated bowel loops
[Table/Fig-2]: Jejuno-jejunal intussescetion
[Table/Fig-3]: Intussesception being reduced



[Table/Fig-4]: Atretic segment **[Table/Fig-5]:** Atretic segment with gangrenous diverticulum with distal meckel's diverticulum

necrosis of intestinal segment with consequent resorption and creation of an atretic pouch This theory seems more likely than alternative theory [1]. These intrauterine vascular events may include volvulus, internal hernia, constriction or bowel incarceration in omphalocele, a tight gastroschisis defect, thrombosis of mesenteric vessels and rarely intussusception.

2. Tandler theory: Lack of re-vacuolisation of the solid cord stage of intestinal development.

In 1949 Laufman [2] demonstrated that isolated sterile devascularized loops of intestine got converted to fibrous bands when left in the peritoneal cavity. Hence, an intrauterine intussusception may lead to vascular insufficiency and an atresia.

Intrauterine intussusception is a rare entity and, it has been described as a rare cause of intestinal atresia. The commonest site of the atresia is usually ileum. In 1888 Chiari was first to recognize that IUI is a rare cause of intestinal atresia, whereas Davis and Poynter were the first to report it in English literature in 1922. In a review of 1500 cases of intestinal atresias by Evans, only 9 (0.6%) cases were due to IUI. Till date less than 100 cases of this association have been noted in the review of literature worldwide, only 2 cases reported from India [3,4].

In a study of 277 cases of intestinal atresias over 25 years, conducted by Laura K et al., intrauterine intussesception was noted only in 2 cases i.e. 1.6% of cases [5]. According to pubmed data base in 30 different publications of intrauterine intussusceptions (between 1975 and 2012) [6,7], reported a total of 79 patients with IUI causing atresia.

Antenatal ultrasonography (USG) rarely detects IUI during pregnancy. Fetal ascites is a reliable finding on antenatal USG suggesting IUI. A "target-like" lesion i.e., a round hyperechoic area of fetal bowel surrounded by hypoechoic ring of bowel wall may be seen on USG. Intestinal dilation, meconium pseudocyst and intra abdominal calcifications are the other non-specific sonographic findings [8]. An abnormal prenatal ultrasound finding was reported in 11% of the reported cases. As per Ming Kwang Shyu persistent ascites, pseudocyst or dilated bowel loops on antenatal USG are highly suggestive of IUI and predict need for postnatal surgery (92%) [9]. Therefore, a pregnant women with suspected fetal anomalies should deliver in a tertiary care centre where emergency neonatal and paediatric surgical facilities are available for better outcome.

The surgical management includes a small bowel resection and end-to-end (or back) anastomosis or a primary intestinal diversion with a delayed anastomosis. The prognosis of this entity is good, and follow up in most of the time is uneventful.

Intestinal atresia due to IUI is usually single and of types II and IIIa involving jejunum or ileum. In IUI the lesion is identical to classic intussusceptions as seen in infants and older children [10].

In a study done by Imai Y et al., it has been explained how an intrauterine intussusception could arise secondary to atresia, resulting in a separated polypoid intussesceptum, with relatively good preservation of the structure of the intestine, observed at the obstructed end on the distal side [11]. Whereas in our case there was no separated polypoid intussesceptum hence it shows that atresia developed secondary to intussusception, so a cause-effect relationship could be established.

In our case along with intrauterine intussusception and jejunal atresia there was associated congenital heart disease (PDA with ASD). Among intestinal atresias associated congenital malformations are more common with duodenal atresia followed by jejunal and rarely ileal. More proximal the atresia is, more the chances of associated anomalies [12].

CONCLUSION

Intrauterine intussusception is a rare cause of intestinal atresia. It follows the theory of intrauterine disruption of mesenteric blood flow leading to intestinal atresia. Prenatal ultrasonography or radiograph rarely establishes the diagnosis. Definitive diagnosis is established only by intraoperative and histopathological features. This entity has a good prognosis provided intervention is on time. Associated congenital malformations like congenital heart disease are not uncommon.

REFERENCES

[1] Giuseppe B, Rodolfo B, Weindling M. Neonatology: A practical approach to neonatal disease. 2012;690.
[2] Laufman H, Martin WB, Method H. Observations in strangulation obstruction, the fate of sterile devascularized intestine in the peritoneal cavity. *Arch Surg* 1949;59:550-64.
[3] Evans CH. Atresias of the gastrointestinal tract. *Int Abstr Surg*. 1951;92:1-8.
[4] Pavri DR, Marshall DG, Armstrong RF, Gorodzinsky FP. Intrauterine intussusception: case report and literature review. *Can J Surg*. 1983;26:376-78.
[5] Laura K, Vecchia D, Grosfeld JL, West KW, Rescorla FJ, Scherer LR, et al. Intestinal atresia and stenosis. a 25 year experience with 277 cases. *Arch Surg*. 1998;133:490-97.
[6] Deshmukh SD, Bavikar R, Naik AM. Jejunal atresia in a neonate due to intra-uterine intussusception. *Indian Pediatr.* 2012;49:149-50.
[7] Kong FT, Liu WY, Tang YM, Zhong L, Wang XJ, Yang G, et al. Intussusception in infants younger than 3 months: a single center's experience. *World J Pediatr*. 2010;6:55-59.
[8] Shimotake T, Go S, Tsuda T, Iwai N. Ultrasonographic detection of intrauterine intussusception resulting in ileal atresia complicated by meconium peritonitis. *Pediatr Surg Int*. 2000;16:43-44.
[9] Shyu MK, Shih JC, Lee HN, et al. Correlation of prenatal ultrasound and postnatal outcome in meconium peritonitis. *Fetal Diagn Ther*. 2003;18:255-61.
[10] Rattan KN, Singh Y, Sharma A, Pandit SK, Malik V. Intrauterine intussusception--a cause for ileal atresia. *Indian J Pediatr*. 2000;67:851-52.
[11] Imai Y, Nishijima E, Muraji T, Hashimoto K, Hayashi Y, Itoh H. Fusion of intussusceptum and intussuscipiens in intrauterine intussusception: a rare type of intestinal atresia. *Pathol Int*. 1999;49:962-67.
[12] Sweeney B, Surana R, Puri P. Jejuno ileal atresia and associated malformations; correlation with timing of in utero insult. *J Pediatr Surg*. 2001;36(5):774-76.

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