

A rare case of intrauterine intussusception causing ileal atresia

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SUMMARY

Intrauterine intussusception is a rare but evident cause of intestinal atresia and is usually detected intraoperatively. We report on a term neonate who presented to the Department of Paediatric Surgery, Sabah Women and Children's Hospital, Malaysia with delayed passage of meconium and intestinal obstruction, wherein the lower contrast showed a claw sign. This was a good clue that this neonate had intrauterine intussusception and this suspicion was confirmed upon laparotomy. We found an ileo-ileal intussusception causing ileal atresia, requiring resection and primary anastomosis.

INTRODUCTION

Intussusception is a condition where one segment of the intestine invaginates into a segment of intestine distal to it. It is a well-known cause of acute intestinal obstruction in infants and young children and has rates reported to be between 0.5 and 4.3 per 1000 live births. Intrauterine intussusception (II) on the other hand, is a rare occurrence, that was first recognised by Chiari in 1888 and then reported for the first time in the published reports in English in 1922 by Davis and Poynter. A review of 1500 cases of intestinal atresia by Evans, noted that II was the cause in only 0.6% of the cases,¹ and Grosfeld J.L reported in detail regarding a case of II causing ileal atresia in 1970. Todani et al reported on 24 cases of II causing intestinal atresia in Japan, and in that report, all the cases involved the small intestine, 87.5% were full-term infants and mostly without any associated anomaly (only 1 premature infant had patent ductus arteriosus).²

CASE REPORT

We report here a case that was referred to the Department of Paediatric Surgery, Sabah Women and Children's Hospital, Malaysia for delayed passage of meconium and vomiting. The baby boy was delivered by a 20-year-old lady who was uneventful antenatally, and prenatal ultrasounds in the third trimester did not reveal any significant abnormalities. He was born term at 38 weeks gestation with a birth weight of 2.8kg and allowed breastfeeding on demand immediately after birth. However, he did not pass meconium in the first 24 hours of life, and only had bowel opening at 40 hours of life and the stool was of normal dark greenish colour. He also developed bilious vomiting within the first 24 hours, after which he was kept nil by mouth with ryles tube inserted into the stomach. On examination, he was non-syndromic, there was no heart murmur, his abdomen was distended, without

any palpable mass or abdominal discoloration, his anus was patent, and per rectum there was no gush of air/explosive stools. Abdominal x-ray showed dilated bowels centrally with sudden paucity of bowel gas. We proceeded with lower GI contrast study, which showed patent but small calibre colon, with contrast refluxing beyond the ileocaecal valve and abruptly stops with a claw sign. The small bowel on the background appeared dilated proximally (Figure 1). With a working diagnosis of small bowel atresia, we proceeded with laparotomy via a right upper transverse incision and found that there was a type IIIA ileal atresia 123cm from the duodeno-jejunal junction and 28cm from ileocaecal valve. Closer inspection of the distal atretic segment revealed bowel intussusceptum with apex around 5cm from the atretic end (Figure 2). The atretic ends were resected followed by primary end to end anastomosis. On post operation day five, tube feeding was initiated and breastfeeding on demand the next day. He was discharged home well on post op day 10.

DISCUSSION

In this case of intrauterine intussusception, our patient was a term neonate without any associated anomalies, consistent with previous reviews.² Intussusception that happens during fetal life can result in intestinal atresia if the time elapsed is long enough for gangrene and resorption of the affected bowel to occur. It was shown by Tsujimoto et al that intestinal atresia could develop in 4-5 days after an intussusception in rabbit fetal models.³ In some cases, the II was complicated with meconium peritonitis, which fortunately did not happen in our case. Despite the passage of normal meconium after 40 hours of life, we could not then rule out intestinal atresia. In a review by Todani et al, 45% of the cases of II causing intestinal atresia passed normal meconium as well,² consistent with the assumption that the II may have occurred in late fetal life after the intestinal lumen had formed. Looking at the lower GI contrast study, the claw sign was a good clue that this neonate had intrauterine intussusception. In the presence of significant bowel dilatation, a lower contrast study can provide useful information such as the claw sign stated, distal atresias, and transitional zones. The review by Todani suggested that the presence of occult blood in the meconium could also be a guide to the diagnosis of II causing intestinal atresia preoperatively.² Jejunal and ileal atresia, unlike duodenal atresia (theory of non-recanalization of gut) happens due to vascular disruption of mesenteric flow leading to intestinal ischemia, necrosis and subsequently resorption which creates an atretic pouch. These vascular accidents can be more

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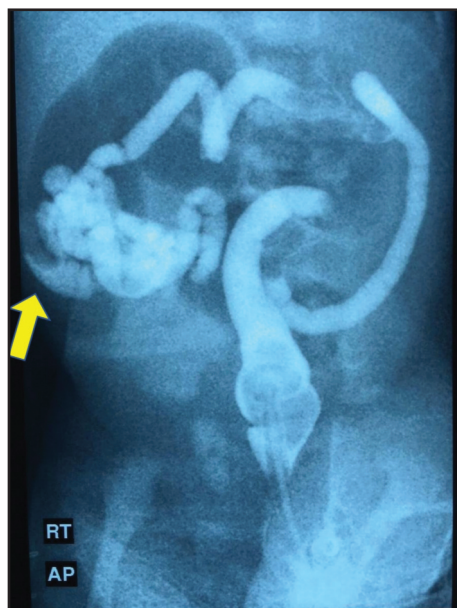


Fig. 1: Contrast enema showing patent colon, with contrast refluxing beyond the ileocaecal valve and abruptly stops with a claw sign (arrow). Small bowel appears dilated proximally.

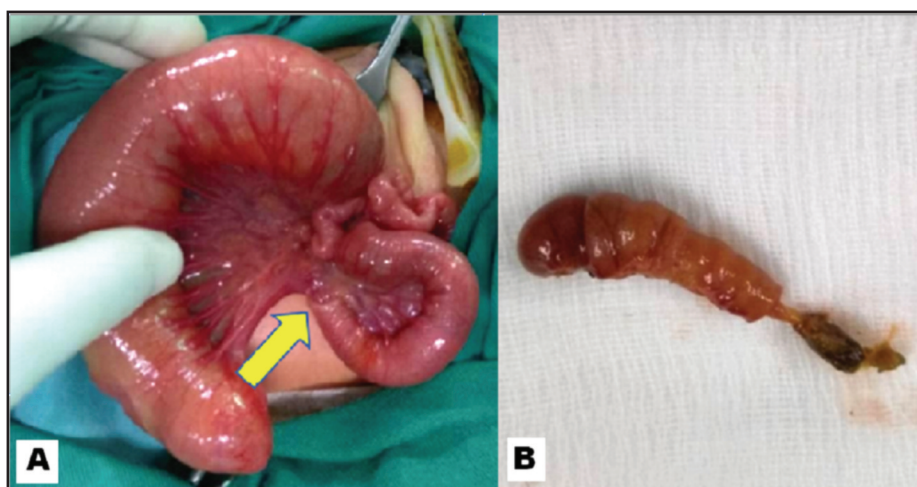


Fig. 2: Intraoperative finding. (A) Ileal atresia with mesenteric defect 123cm from DJ, dilated proximal pouch. Bowel intussusception (arrow) noted at the distal atretic segment (28cm from ICV), with the apex around 5cm from the atretic end. (B) Resected distal atretic segment showing the gangrenous intussusceptum bowel protruding out of the intussuscipten bowel.

commonly caused by intrauterine volvulus, internal herniation, incarceration in an omphalocele, a tight gastroschisis defect, or thrombosis of mesenteric vessels. II causing small bowel atresia remains an uncommonly reported entity. It is postulated that II impedes the blood flow to the affected bowel segment, and results in gangrene and resorption as mentioned above. However, the cause as to why II occurs, remains a mystery. The pathogenesis of intussusception in children has been described as being secondary to uncoordinated peristalsis of the gut, lymphoid hyperplasia or pathologic lead points. Can the same be said for intrauterine intussusception? There have been isolated reports of Meckel's diverticulum playing a role as the lead point to II, but in this case, it was not demonstrable. More studies are needed to better understand this pathology.

CONCLUSION

Intrauterine intussusception is a rare but evident cause of intestinal atresia. It has a good prognosis if treated with surgical intervention promptly.

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CONFLICT OF INTEREST

The authors declare that there are no conflicts of interest.

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