Case Report

Radiological presentation of a case of Type-III ileal atresia with unused microcolon in a rural setup- A case report

Yadav S[1], Rawal G[2], Gupta N[3]

[1]Dr. Sankalp Yadav, General Duty Medical Officer-II, Chest Clinic Moti Nagar, North MCD, New Delhi, India, [2]Dr. Gautam Rawal, Attending Consultant, Critical Care, Rockland Hospital, Qutab Institutional Area, New Delhi, India, [3]Dr. Nishant Gupta, Department of Pediatrics, RML Hospital, President's Estate, New Delhi, India.

Address for correspondence: Dr. Sankalp Yadav, Email: drsankalpyadav@gmail.com

Abstract

Ileal atresia is the result of an in-utero vascular accident, leading to distal small bowel obstruction. Contrast enema is the primary imaging study to diagnose distal bowel obstruction. We present a case of a four day old child (neonate) with a history of bilious vomiting; diagnosed as ileal atresia with unused microcolon on plain film of the abdomen and contrast enema. In operation, the child had Type-III atresia.

Keywords: Ileal atresia, Microcolon, Gastroschisis, In-utero, Mesenteric defect.

Introduction

Neonatal intestinal obstruction occurs either from insults in-utero after the formation of normal bowel or from intrinsic developmental defects, abnormalities of peristalsis or abnormal intestinal contents. One of the common causes of bowel obstruction in newborn is intestinal atresia [1]. In fact, intestinal atresia is one of the three most common causes leading to intestinal obstruction, the other two being Hirschsprung's disease and anorectal malformation [2]. The most common site of intestinal obstruction is in the jejunum and ileum. The incidence of jejuno-ileal atresia varies from 1 in 330 to 1 in 1500 live births [3]. Among all the intestinal atresias, ileal atresia is most common and comprises about 50% of small bowel atresia's [2]. Ileal atresia results from in-utero ischemic insult [4,5]. Microcolon is not common in newborns. It is mainly due to the intestinal obstruction as seen in ileal atresia [6]. There are four types of jejuno-ileal atresia. Type-I is most common, in which there is blind separated intestinal ends with a mesenteric defect. Failure to diagnose neonatal bowel obstruction may result in aspiration of vomit, sepsis, mid-gut infarction or enterocolitis. Diagnosis is done by evaluation of plain radiographs of the abdomen, contrast enema and ultrasound study. We report a rare, unusual presentation of a neonate with Type-III ileal atresia, presenting a history of two episodes of passing meconium and bilious vomiting.

Case Report

A four day old Indian male child, the first product of consanguineous marriage, born of a full term normal vaginal delivery with no perinatal complication, weighing 2.8 kg was brought to our outpatient department with a history of persistent vomiting since birth and two episodes of passage of greenish colored meconium plugs after giving a rectal wash. Family history was non-significant. The child was immediately admitted to the neonatal intensive care unit. He was pink, active and stable. Cardiovascular and respiratory system were normal. However, his upper abdomen was distended without any mass. Later, the child did not pass any stools. Hence, a possibility of meconium ileus was considered clinically. In a plain radiograph study of the abdomen, grossly dilated featureless air filled bowel loops were seen all over the abdomen. (Fig.1)

Manuscript received: 25th Jan 2015
Reviewed: 4th Feb 2015
Author Corrected: 9th Feb 2015
Accepted for Publication: 24th Feb 2015

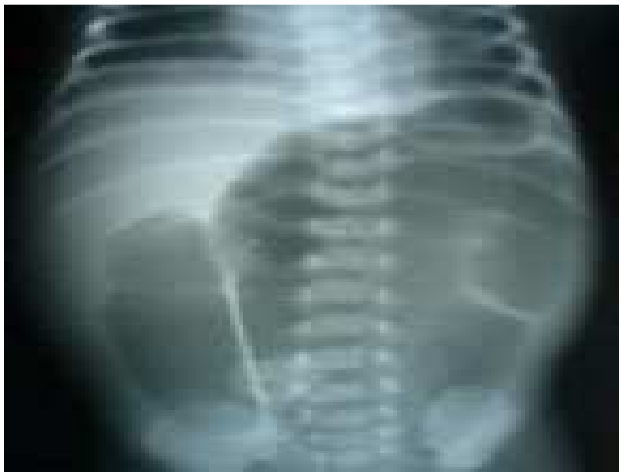
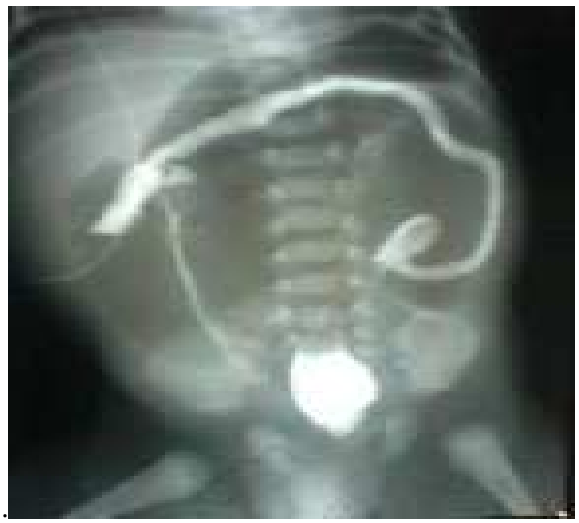


Fig 1: Plain radiograph of the abdomen reveals gas filled dilated featureless bowel loops, suggestive of intestinal obstruction. No calcification is noted in the abdomen.

No abdominal calcification was seen. Renal function test was normal. A provisional diagnosis of intestinal obstruction was given; a possibility of meconium ileus or small bowel atresia was kept in mind. A contrast enema study was done using water-soluble contrast media containing Urografin (1/3rd), Glycerol (1/3rd) and normal saline (1/3rd). This was done considering the possibility of meconium ileus. It revealed a small calibered featureless colon. The appendix was opacified and appeared normal. Caecum was in right iliac fossa. Contrast did not pass beyond the ileocaecal junction. It revealed a small featureless colon suggestive of 'unused microcolon'. (Fig. 2 and Fig. 3).



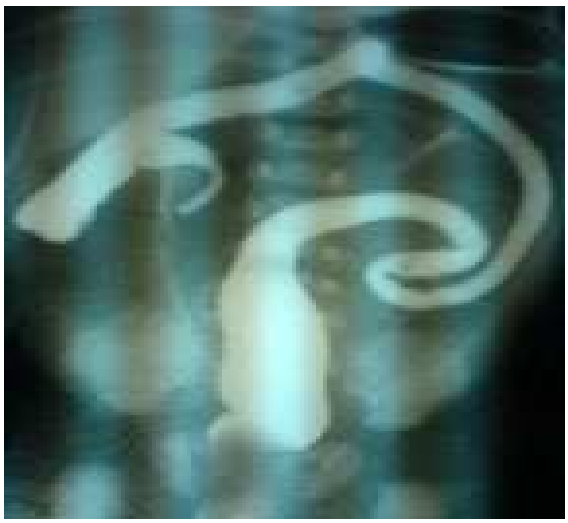
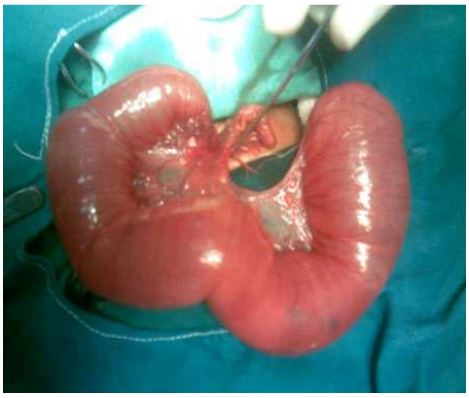


Fig 2 and Fig 3: Contrast enema reveals unused microcolon with obstruction to passage of contrast beyond the ileocaecal valve. In addition, dilated gas filled bowel loops are seen in the background, suggestive of ileal atresia.

There were no filling defects in the colon. A diagnosis of distal ileal atresia was considered in enema study. The child underwent exploratory laparotomy which revealed type-III atresia of ileum just proximal to ileocaecal junction with grossly distended proximal ileal segment, distal ileal bud (distal to atretic segment) and a large mesenteric defect. (Fig. 4 and 5).



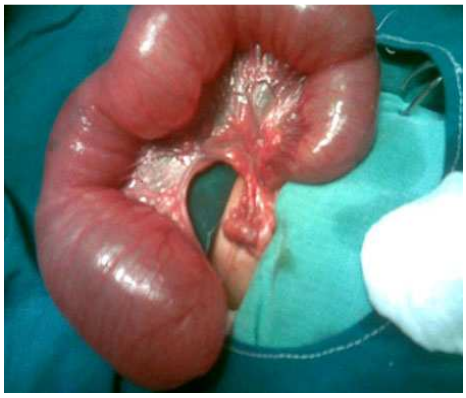


Fig 4 and Fig 5: Intraoperative photograph showing grossly dilated proximal ileum with atretic distal ileum and associated mesenteric defect.

There was no evidence of inflammation. Resection of ileum, 10 cm proximal to atresia as well as distal ileal bud was done and ileocaecal end to side anastomosis was done. Feeding was started using the transgastric jejunostomy tube on the fourth post-operative day. The specimen was sent for histopathological examination. The child fared well and recovered after surgery. There were no postoperative complications. The child was discharged after two weeks.

Histopathological examination of the resected ileum revealed congestion and hemorrhage in the submucosa. There was no evidence of necrotizing enterocolitis.

Discussion

Atresia refers to congenital obstruction with complete occlusion of the intestinal lumen and accounts for 95% of all intestinal obstructions [7]. Stenosis (partial occlusion) accounts for remaining 5% of obstruction [7]. Jejuno-ileal atresia may involve the bowel anywhere from the ligament of Trietz to the ileocaecal valve, with the majority of cases occurring at the extremes of small bowel [8]. Proximal atresia presents with bilious vomiting whereas more distal atresia's present with abdominal distension and failure to pass meconium [4]. Ileal atresia comprises approximately 50% of all small bowel atresia's [2]. In-utero vascular accidents are the major factor leading to intestinal atresia [9-12]. Other factors involved in the etiopathogenesis of ileal atresia are in-utero intussusception, intestinal perforation, segmental volvulus, thromboembolism and gastroschisis [13].

Ileal atresia was first described in 1684 by Goeller [7]. Louw and Barnard in 1955 demonstrated the role of late intrauterine mesenteric vascular accidents as the likely cause of jejunoileal atresia, rather than the popularly accepted theory of inadequate recanalization of the intestinal tract [14].

Prevalence of jejuno-ileal atresia varies in different countries. It is about 2.25 per 10000 live births in France [15]. However, in Spain and Latin America the overall prevalence of small intestinal atresia is 1.3 per 10000 live births [16]. Jejuno-ileal atresia constitutes one third of all causes of neonatal intestinal obstruction [17]. There is no gender specificity and boys and girls are equally affected [17]. The parents are more often consanguineous and vaginal bleeding frequently complicates such pregnancies [13]. Correlation between jejuno-ileal atresia and parental age or disease has been not been proved so far [18-20]. The congenital anomalies and chromosomal abnormalities are less common with jejuno-ileal atresia as opposed to duodenal atresia [13]. The jejunum and ileum are equally affected [13]. Antenatal perforation is common in ileal atresia leading to meconium peritonitis with or without cyst formation [13]. Ileal atresia's have short postoperative course and low mortality [1,7]. A child with ileal atresia present with bilious vomiting, abdominal distention and failure to pass meconium. Passage of meconium does not rule out intestinal atresia as in this case, when two episodes of passing greenish meconium were noted, which was rather unusual. Bilious vomiting in neonates should be considered secondary to mechanical obstruction, until proved otherwise and emergency surgical evaluation is warranted in every such case.

The small bowel atresia is classified into following types viz. 1) Type-I- single membranous atresia with continuity of bowel and intact mesentery with no mesenteric defect, 2) Type-II- consists of a single atresia with discontinuity of bowel wall, 3) Type-IIIA- consists

of atresia without connection by a fibrous cord, with a mesenteric gap, and 4) Type-IIIB- apple peel deformity or Christmas tree deformity, the intestinal segments are separated as in Type-IIIA and the mesenteric defect are large [2]. Atresia is seen in the proximal jejunum near the ligament of Trietz and the pouch is distended and lacks mesentery. The superior mesenteric artery distal to the middle colic branch is absent. The collapsed distal intestinal helically encircles a small marginal artery from ileocolic or right colic arcades or inferior mesenteric artery and its vascularity may be impaired, Type-IV- shows multiple discrete atresia's [13,21]. The incidence of various types of atresia is: Type-I- 32%, Type-II - 26%, Type-III - 26%, and Type-IV - 17% [8].

Intestinal atresia is diagnosed antenatally by ultrasonography. Antenatal ultrasound diagnosis relies on demonstration of multiple interconnecting, overdistended bowel loops, echogenic fetal bowels and polyhydramnios [22-24]. Conventional radiology is the primary imaging modality to diagnose ileal atresia. However, ultrasound may provide diagnostic information, if clinical and plain radiographic findings are atypical. The abdominal radiograph demonstrates multiple air fluid levels with non-visualization of rectum in prone cross table lateral radiograph [21]. Demonstration of a microcolon in contrast enema is diagnostic of a distal small obstruction [21]. It is present in all cases, except those in whom the atresia is caused shortly before birth. If possible, the contrast should be allowed to proceed proximal to the ileocaecal valve to demonstrate the site of atresia [21]. The term 'microcolon' means an abnormal small caliber of the colon. This term is a misnomer, as it implies an intrinsic abnormality of the colon. In fact, the term is synonymous with 'unused colon' [21]. Once the obstruction is relieved, the unused colon becomes of normal caliber [21]. Patients with a high level of obstruction maintains normal caliber of colon by virtue of the passage of the intestinal fluid and desquamated mucosal cells, termed as succus entericus. In patients with low small bowel obstruction, there is insufficient production of succus entericus entering the colon, resulting in an unusual abnormally small colon [21]. In cases of distal obstruction of relatively recent onset, the colon has had time to attain normal caliber and therefore, a microcolon is not present. Thus, the presence of a microcolon is diagnostic of a longstanding distal small bowel obstruction. But, normal colon does not exclude this condition in all cases. There are no filling defects in the microcolon and distended bowel, as in meconium ileus or meconium plug syndrome [8]. Bowel wall calcification in plain x-ray may be seen due to prior ischemia or infarction [8]. Large lucent areas in abdomen represent abnormally dilated distal bowel loops. The contrast enema study should be carefully performed, as the distal blind bowel is prone to perforation. Isotonic non-ionic contrast is used in sick premature infants as it has less adverse effects, if leaked in the peritoneal cavity through perforation [8]. The other differential diagnoses are Hirschsprung's disease, meconium ileus, colonic atresia and megacystis-microcolon syndrome [21]. Especially, a long aganglionic segment may be confused with unused microcolon. Megacystis- microcolon- intestinal hypoperistalsis syndrome is a pseudoatresia with functional small bowel obstruction, microcolon, malrotation and a large unobstructed bladder [8]. Contrast enema also indicates the position of caecum with regards to possible malrotation and shows the level of obstruction in cases of colonic atresias. Differentiation between ileal and colonic obstruction is not possible by plain radiographs, but distinction can be made by the contrast enema study. The unused microcolon is also called 'functional microcolon' [5].

Ultrasound, if performed, reveals multiple loops of dilated small bowel filled with fluid and air, peristalsis may be normal or increased [4]. With associated meconium ileus; there are multiple loops of bowel filled with highly reflective material suggesting the presence of thick, tenacious meconium [4]. The peristalsis has been often poor. Associated meconium peritonitis or meconium pseudocysts, may rarely be seen [4].

Survival of patients with intestinal obstruction has improved within the last twenty years because of an improved understanding of intestinal physiology and the etiologic factors of the condition, refinements in pediatric anesthesia and advances in surgical and perioperative care of newborns [13]. After preoperative stabilization in the form of gastrointestinal decompression, electrolyte disturbances correction, antibiotherapy and normothermia, the treatment consist of exploratory laparotomy, resection of proximal dilated intestine and end-to-oblique anastomosis in distal jejunoileal atresia [6]. After operation, overall survival is 90% in ileal atresia and surgical mortality is less than 1%. Infection related to pneumonia, peritonitis or sepsis is the most common cause of death in infants with jejunoileal atresia [13]. The most important surgical complications are anastomotic leaks and gastroschisis functional anastomotic obstruction [13].

Conclusion

This case of newborn with persistent vomiting had ileal atresia. The plain radiograph of the abdomen and contrast enema helped in clinching the diagnosis and helped in successful management of the child with surgery. The authors stress the importance of conventional radiology in the diagnosis of ileal atresia and also the role of contrast enema in differentiating this condition from other diseases like meconium plug syndrome, meconium ileus and total colonic Hirschsprung's disease. In such a case, conventional methods of diagnosis are still relevant, especially in our rural setup. The case was managed surgically on the basis of contrast enema and plain radiograph findings.

Competing Interests- Authors declare no competing interest.

Authors' Contributions- All the authors have contributed to this case report and have read and approved the final version of the manuscript.

References

1. Shakya VC, Agrawal CS, Shrestha P, Poudel P, Khaniya S, Adhikary S. Management of jejunoileal atresias: an experience at eastern Nepal. BMC Surg. 2010;10:35.

2. Das PC, Rai R, Lobo GJ. Jejunal atresia associated with idiopathic ileal perforation. J Indian Assoc Pediatr Surg. 2008 Apr-Jun; 13(2):88-89.

3. Hasan Z, Gangopadhyay AN, Srivastava P, Hussain MA. Concavo-convex oblique anastomosis technique for jejuno ileal atresia. J Indian Assoc Pediatr Surg. 2009 Oct-Dec; 14(4):207-209.

4. Marilyn J. Siegel. Pediatric general abdomen. In: Hylton B, Meire, Cosgrove DO, editors. Abdominal and General Ultrasound, Churchill Livingstone publication; 2001(Vol 2). p. 1173-74.

5. Touloukian RJ. Intestinal atresia and stenosis. In: Ashcraft KW, Holder TM, editors. Pediatric surgery. 2nd ed. Philadelphia: Saunders; 1993. p. 305-19.

6. Lo WC, Wang CR, Lim KE, Microcolon in Neonates: Clinical and Radiographic Appearance. Journal of Clinical Neonatology 1998;5(1):14-18.

7. O'Neill JA Jr. Duodenal atresia and stenosis. In: O'Neill JA Jr, Rowe MI, Grosfeld JL, et al. eds. Pediatric Surgery. 5th ed. St Louis, Mo: Mosby; 1998. p. 1133-44.

8. Pawa S. Developmental Lesions of Gastrointestinal tract. In: Berry M, editor. Paediatric Radiology, 1st edition. Jaypee publication; 1997. p. 11-12.

9. Louw JH, Barnard CN. Congenital intestinal atresia; observations on its origin. Lancet. 1955;269 (6899) 1065-67.

10. Grosfeld JL. Jejunoileal atresia and stenosis. In: Welth KJ, Randolph JG, Ravitch MM, et al., editors. Pediatric surgery. 4th ed. Chicago: Year Book Medical Publishers, Inc; 1986. p. 838-48.

11. Koga Y, Hayashida Y. Intestinal atresia in fetal dogs produced by localized ligation of mesenteric vessels. J Pediatr Surg. 1975;10(6):949-53.

12. Puri P, Fijimoto T. New observations on the pathogenesis of multiple intestinal atresias. J Pediatr Surg. 1988;23(3):221-5.

13. Shalkow J, "Small intestinal atresia and stenosis treatment & management", emedicine.medscape.com, http://emedicine.medscape.com/article/939258-treatment#a1134; (Accessed on 29 January 2015).

14. Louw JH, Barnard CN. Congenital intestinal atresia; observations on its origin. Lancet. Nov 19, 1955;269(6899):1065-7.

15. Francannet C, Robert E. Epidemiological study of intestinal atresias: central-eastern France Registry 1976-1992 [in French]. J Gynecol Obstet Biol Reprod (Paris). 1996;25(5):485-94.

16. Martinez-Frias ML, Castilla EE, Bermejo E, et al. Isolated small intestinal atresias in Latin America and Spain: epidemiological analysis. Am J Med Genet. Aug 28 2000;93(5):355-9.

17. DeLorimier AA, Fonkalsrud EW, Hays DM. Congenital atresia and stenosis of the jejunum and ileum. Surgery. May 1969;65(5):819-27.

18. Mulholland MW, Lillemore KD, Doherty GM, et al. eds. Intestinal atresias. In: Greenfield's Surgery,

Scientific Principles and Practice. 3rd ed. Philadelphia, Pa: Lippincott Williams & Wilkins; 2002.

19. Francannet C, Robert E. Epidemiological study of intestinal atresias: central-eastern France Registry 1976-1992 [in French]. J Gynecol Obstet Biol Reprod (Paris). 1996;25(5):485-94.

20. Martinez-Frias ML, Castilla EE, Bermejo E, et al. Isolated small intestinal atresias in Latin America and Spain: epidemiological analysis. Am J Med Genet. Aug 28 2000;93(5):355-9.

21. Hernanz- Schulman M, Imaging of neonatal gastrointestinal obstruction, RCNA, Vol 37(6); 1999:1174-1180.

22. Phelps S, Fisher R, Partington A, Dykes E. Prenatal ultrasound diagnosis of gastrointestinal malformations. J Pediatr Surg. Mar 1997;32(3):438-40.

23. Tam PK, Nicholls G. Implications of antenatal diagnosis of small-intestinal atresia in the 1990s. Pediatr Surg Int. 1999;15(7):486-7.

24. Louw JH. Resection and end-to-end anastomosis in the management of atresia and stenosis of the small bowel. Surgery. Nov 1967;62(5):940-50.

.....

How to cite this article?

Yadav S, Rawal G, Gupta N. Radiological presentation of a case of Type-III ileal atresia with unused microcolon in a rural setup- A case report. *Int J Med Res Rev* 2015;3(2):237-242. doi: 10.17511/ijmrr.2015.i2.40.

……..