Ileal Atresia and Total Colonic Hirschsprung Disease in a 36-week Neonate: A Case Report

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Abstract

Intestinal atresia and Hirschsprung disease are two common causes of bowel obstruction in neonates,simultaneous occurrence is rare. This report delineates a 36-week newborn with ileal atresia and total colonic Hirschsprung who was referred to our unit due to failure of meconium passage during the first 48 hours after birth

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Patient Consent Statement

CONSENT FORM

For a patient's consent to the publication of Information about them in a journal or thesis

Name of a person described in the article or shown in the photograph:**Mahdi Javadpoor**

The subject matter of the photograph or article : Figures related to the treatment steps of **Mahdi Javadpoor** ,

a 36-week male neonate referred to the pediatric surgery service on the second day of life due to Ileal atresia and total Colonic Hirschsprung disease.

Title of article : Ileal Atresia and Total Colonic Hirschsprung Disease in a 36-week Neonate: A Case Report

A medical practitioner or corresponding author**: Khashayar Atqiaee**

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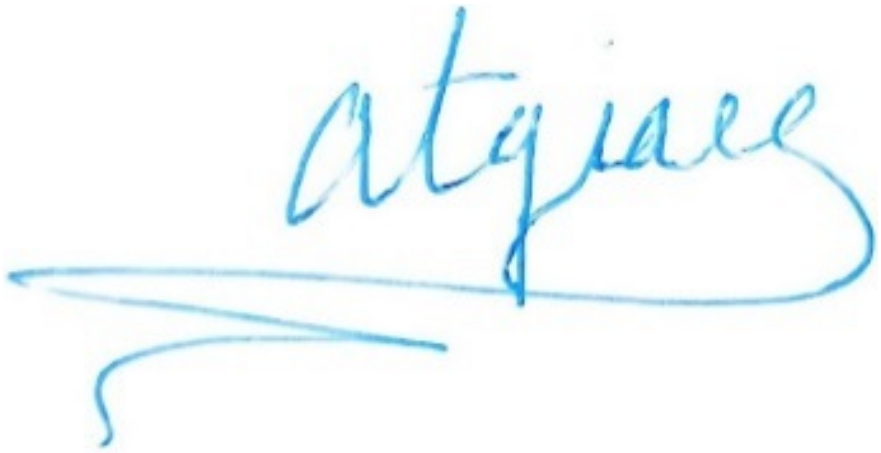

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ABSTRACT

Intestinal atresia and Hirschsprung disease are two common causes of bowel obstruction in neonates, simultaneous occurrence is rare.

This report delineates a 36-week newborn with ileal atresia and total colonic Hirschsprung who was referred to our unit due to failure of meconium passage during the first 48 hours after birth.

Key Clinical Message : Colonic aganglionosis should be in mind in any operated infant with small intestinal atresia repair who continues to exhibit poor bowel function after corrective surgery.

Key words:

Aganglionosis; Hirschsprung disease; Ileum Atresia

1 Introduction

Intestinal atresia is a significant cause of primary bowel obstruction in neonates accounting for 22.4% of all cases [1]. On the other hand, Hirschsprung disease (HD) is the most frequent congenital intestinal motility disorder [2]. HD occurs when bowel segments are not supported by neuronal ganglion formation within the myenteric (Auerbach) and submucosal (Meissner) plexus, which promote gastrointestinal movements.

And in 80% of cases, the absence of ganglion cells is limited to the rectosigmoid area. However, aganglionosis may extend through the colon with the distal ileum (10 %) or more proximal segments [3]. Also, in these cases, a water-soluble enema shows a "question mark" configuration in the foreshortened colon with no transitional zone. [4] The rectal biopsy can always confirm the diagnosis.

Concurrent small intestinal tract atresias associated with Hirschsprung are less than 1%.

In this case report, we simultaneously present an infant born with ileal atresia and Hirschsprung disease.

2 Case Presentation

A 36-week male neonate was referred to the pediatric surgery service on the second day of life due to failure to pass meconium, abdominal distension, and bilious emesis.

The patient was born to a 23-year-old healthy primipara Iranian mother without a history of abortion via normal vaginal delivery. Her birth weight was 2500 grams.

On physical examination, the primary vital signs were in the normal range, bowel sounds were decreased, and the rectum was empty.

Initial abdominopelvic radiography revealed Thumb-sized intestinal loops with no gas in the rectum [Figure 1A]. Small-bowel contrast series confirmed dilated proximal blind end [Figure 1B].

Baseline biochemical and blood gases analyses were in the normal range. On the third day, with a diagnosis of intestinal obstruction, the patient underwent exploratory laparotomy after obtaining informed consent from the parent. We identified Ileal atresia with a proximal dilated, blind-ending1 cm distal bud attached to the ileocecal region (Type IIIa). [Figure 2] The colon appeared as an unused microcolon throughout. After resecting the proximal dilated ileal pouch and ileocecal part, surgery was followed by an end-to-end ileo-ascending colonic anastomosis.

Permanent Hematoxylin & Eosin (H&E) pathological examination revealed the absence of ganglion cells in the cecum, the appendix, and the rectum and the abundance of ganglia in the ileum [Figure 3].

Due to the persistence of abdominal distention, no defecation on postoperative days, and a histopathologic report that was compatible with total segment HD, the patient was a candidate for re-exploration. [Figure 4]

Our findings were a new colon and a functional intestinal obstruction at the anastomosis site with no anastomotic leak.

After taking multiple levels of frozen section biopsies to reaffirm the total colonic aganglionosis, the patient underwent an ileostomy and colonic mucus fistula bypass

The final permanent pathology studies by calretinin test disclosed total aganglionic colon HD.

Swenson pull-through surgery with removal of all colon and ileoanal anastomosis was performed on three month old. The case tolerated oral feeding after five days and was discharged from the hospital in good condition.

3 Discussion

Co-existence of ileal atresia and total colonic HD is a rare event. [5, 6] Conforming to the currently accepted theories, jejunoileal atresia arises due to intrauterine ischemic vascular events in the third trimester, such as intussusception, perforation, volvulus, or thromboembolism; maternal smoking as a hypercoagulation state [7, 8] and cocaine use as vasoconstrictive medications [9]

The migration of ganglion cells was completed through the gastrointestinal tract from proximal to distal by 13 weeks postconception. Therefore, it is suggested that early gestational atresia in the 6th to eighth weeks of gestation would result from an ischemic insult to interrupt the caudal migration of ganglion cells and lead to total colonic HD [10, 11]. Finding a very small micro colon, no fibrotic of the left colon, and no meconium distal to atretic segments strengthen this theory.

The other hypothesis that can justify this concurrency is an increased colonic intraluminal pressure and subsequent perforation ileocecal portion due to a developed HD and the secondary small bowel atresia. However, in our case, there was no evidence of meconium spillage into the peritoneal space during our laparotomy, which weakens the second assumption.

As the common occurrence of microcolon in the cases of distal small intestinal atresia, it is tough to differentiate this colonic appearance during surgery from concomitant total colonic aganglionosis and small bowel atresia.

Therefore, it appears rational to do per-operative colonic biopsies looking for ganglion cells on a frozen section to exclude or confirm the underlying HD in suspicious cases.

A definitive reconstructive operation should be planned once we have established the diagnosis and done a proximal ileostomy.

However, there are controversial questions about the correct timing and the most appropriate treatment options [12, 13].

Several approaches have been described to treat this, such as primary pull-through without ileostomy or total colectomy with standard techniques (Swenson, Duhamel, or Soave). Neither is superior to the others.

Albeit discriminating, the best operative approach should be constructed based on the surgeon's level of expertise [13]; in our case, we did total colectomy with an ileoanal Swenson procedure when the patient status was allowed.

In conclusion, this rare concurrency should be considered in cases of small bowel atresia with poor bowel function after the corrective operation.

Authors' contributions

Mehran Hiradfar designed the work and had the role of directing and supervising.

Khashahyar Atqiaee supervised the project process and did scientific corrections.

Ali Samady Khanghah and Mahdi Parvizi Mashhadi prepared the manuscript and interpreted the data.

All authors read and approved the manuscript

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Consent

The authors confirmed that they had gotten all proper patient written consent formats. The parents

have given their consent for their images and other clinical Information to be reported in the form.

The parents were informed about the confidentiality of the names and initials, and efforts would

be made to hide their identity

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Figures' legends:

Figure 1 : **A** : several dilated gastrointestinal loops with no gas in the rectum. **B:** Small-bowel contrast study reveals dilated proximal blind end.

Figure 2 : **A** : Type (III-a) atresia at the laparotomy**B** : The distal segment of the ileum seen as a bud in the ileocecal region

Figure 3 : The Hematoxylin and Eosin study of the ileal in the left and colonic specimen in the right revealing absence of neurologic ganglia compatible with HD ($\times$ 1000 magnification).

Figure 4 : Contrast barium enema demonstrates an unused colon and a functional intestinal obstruction at the anastomosis site