American Journal of Food Science and Nutrition
ISSN: 2232-8232
Volume 01, Issue 03, March, 2020

Intrauterine intussusception: a rare cause of ileal atresia and Hirschsprung's disease

Maha LANDOLSI[1], Aida DAIB[2], Youssef HELLAL[3]

ABSTRACT— Intrauterine intussusception is an extremely rare cause of intestinal atresia. It may occur at late stage of pregnancy and cause impairment of blood supply to a segment of intestine leading to its resorption and atresia. Of same, the aganglionosis caused by an intrauterine vascular accident has been shown. The coexistence of Hirschsprung's disease, intrauterine intussusception and ileal atresia has yet to be described in the literature. We reported a rare case of 2-day old newborn presented with acute bowel obstruction. An abdominal radiograph identified distended loops of bowel with no air in the rectum. An emergency surgery was performed. At laparotomy, an ileal atresia associated to ileocecal intussusception were found. Distally, there was an unused colon. A terminally anastomosing resection were performed. The child had developed a postoperative functional obstruction requiring a surgical revision. A corrective anastomsis was found with a dilated proximal small bowel and an unused colon. An ileostomy and multi-level biopsy were performed. Histology confirmed the diagnosis of total colonic Hirschsprung disease. The last surgery was performed at the age of two months. It consists of a total colonic resection and Sovae ileorectal pull-through.

KEYWORDS: Hirschsprung's disease intrauterine intussusception

1. INTRODUCTION

Ileal atresia caused by intrauterine intussusception is very rare and few cases were reported in literature [1]. The coexistence of Hirschsprung's disease, intrauterine intussusception and ileal atresia has yet to be described in the literature. The etiology of intrauterine intussusception remains unknown [2]. It may cause vascular disruption in a segment of intestine leading to its resorption and atresia [3]. Of same, the aganglionosis caused by an intrauterine vascular accident has been shown [2], [4]. we present our case of an ileal atresia associated to colonic aganglionosis caused by an intrauterine intussusception.

2. CASE REPORT

A 3400-g full term male was delivered by cesarean section because of failure to progress. On his second day of life, he was presented an abdominal distension, bilious vomiting without meconium emission. On physical examination, his general state was preserved. His abdomen was moderately distended and it was diffusely tender. An abdominal radiograph showed dilated loops of small bowel without gas in the rectum. Transrectal probe test was negative. An emergency surgery was performed. At laparotomy, an ileal atresia associated to ileocecal intussusception were noted with an unused colon (fig1, fig2). A terminally resction anastomosis ileo colic with modeling was performed. Five days later, the child has developed a postoperative functional obstruction. Surgical revision was performed. A corrective anastomsis was found with a dilated proximal smal bowel and an unused colon that was relieved with ileostomy. Suspecting Hirschsprung's disease, multi-level biopsy was performed. Final histoligical examination reported no ganglion cells in the cecum whereas biopsies of the ileum demonstrated the presence of ganglion cells. A complement of a rectal suction biopsy detected the absence of ganglion cells in the colonic submucosa, confirming the diagnosis. The final surgery was performed at the age of two months. It consisted of a total colectomy with soavae ileo-endomuscular pull-through. The post- operative clinical course was satisfying (fig3).

3. DISCUSSION

The association of Hirschsprung's disease and ileal atresia is extremely rare and has an incidence of 0.25 percent to 0.8 percent [5]. The theory of the origin of intestinal atresia is an acute intrauterine vascular accident caused by disruption of mesenteric flow leading to polypoid protrusion and bowel necrosis and resorption. [2], [6]. Therefore, intestinal atresia caused by intrauterine intussusception is a rare clinical entity. Their incidence has been reported to range from 0.6 percent to 13 percent [7], [8]. It is characterized by lower and single intestinal atresia seen in full-term babies with near-normal caliber colon, suggesting late event during pregnancy [3]. On the other hand, Hirschsprung's disease associated to intestinal atresia is explained by a possible common vascular accident with loss of bowel which consequently causes a failure of the caudal migration of the myenteric plexus [9], [10]. In this combination we often find an intestinal atresia with a small-caliber <microcolon> [11]. Although the finding of abnormal blood supply to the bowel would possibly support this hypothesis. The early development of ganglion cells between the 5th and 12th week of gestational life and the later delay of the development of the intrauterine vascular accident and consequently the ileal atresia in the most cases would seem to be against this explanation. In addition, the lack of histological evidence of ischemia in the colon in most cases concluded that the hypothesis of a vascular cause for coexistence of Hirschsprung's disease and ileal atresia is based on circumstantial evidence and few experimental data [12]. Therefore, this theory is challenged by several others authors. The possible association to chromosomal abnormalities has suggested but the corroboration of this theory is lacking in the literature [12]. Another theory has suggested that the bowel proximal to an aganglionic segment filed dilated intrauterine volvulus, infarction, and ischemia and caused an atresia [13]. This hypothesis would seem to be the most plausible and logical explanation for the coexistence for these abnormalities. Thereby, our observation holds this hypothesis and we suggest that the intrauterine intussusception was secondary to the large disparity size between the proximal and the distal aganglionic bowel, occur in the late stages of pregnancy, causing ischemia and a single ileal atresia. As ileal atresia caused by intrauterine intussusception occurs very late in pregnancy, a normal to near- normal size colon is revealed [6] and the surgical procedure usually consists to resection of the dilated proximal and terminally primary anastomosis to the distal portion. Unlike the combination of colonic aganglionosis, where there is an unused colon or microcolon [11], [13]. Considering the low frequency of bowel atresia associated to Hirschsprung's disease, the therapeutic and the diagnostic approach are not well described in the literature [13]. In that event, the operation should include a biopsy of terminal ileum, the cecum, or the appendix for ganglions cells examination and ileostomy confection. In our case, we don't have suspected the Hirschsprung's disease during the initial operation and the diagnostic of ilelal atresia caused by intrauterine intussusception was retained. Therefore, we practiced resection of the dilated proximal ileum and cecum, a modeling and terminally primary anastomosis to the distal portion. Despite a corrective surgery, the child has keeping an occlusion. In conclusion, the coexistence of Hirschsprung's disease should be suspected in any child who had small bowel atresia repair but continues to exhibit poor bowel function after corrective surgery [11].

4. CONCLUSION

The intrauterine intussusception as a cause of intestinal atresia confirmed the vascular theory. It is a clinical rare entity whose early surgical intervention is required for good outcome. In that event Hirschsprung's disease is identified as a rare cause of prolonged distal bowel obstruction after a corrective ileal atresia repair. Under these circumstances, multi-level biopsy and research of ganglion cells before further surgery should be practiced.

5. REFERENCE

[1] Sue L. Hall. (1987). Neonatal intussusception associated with neonatal small left colon. Clinical

pediatrics, 191-193.

[2] Hiroaki Komuro, Tetsuo Hori. (2004). The etiologic role of intrauterine volvulus and intussusception in Jejunoileal atresia. Journal of Pediatric Surgery, 39: 1812-1814.

[3] Leuw JH and Barnard CN. (1955). Congenital atresia. Lancet 2, 1065- 1067.

[4] Williams MD and Burrington JD. (1993). Hirschsprung's Disease Complicating Colon Atresia. Journal of Pediatric Surgery, 28, 637-639.

[5] Ikeda K and Goto S. (1986). Additional anomalies in Hirschsprung's disease: an analysis based on the nationwide survey in Japan. Zeitschrift für Kinderchirurgie, 41, 279-281.

[6] Carlos Pueyo. (2009). Intrauterine intussusception: a rare cause of intestinal atresia. Journal of Pediatric Surgery, 44, 2028–2030.

[7] Todani T, Tabuchi K, Tanaka S. (1975). Intestinal atresia due to intrauterine intussusception: analysis of 24 cases in Japan. J Pediatr Surg, 10, 445–51.

[8] Luca U, Esposito C, Porreca A. (1995). Intrauterine intussusception as a cause of ileal atresia occurring late during pregnancy. Eur J Pediatr Surg, 5, 116-8.

[9] Okamoto E, Ueda T. (1967). Embryogenesis of intramural ganglia of the gut and its relationship to Hirschsprung's disease. J Pediatr Surg ,2, 437-443

[10] Janik JP. (1997). Ileal atresia with total colonic aganglionosis. J Pediatr Surg, 32, 1502- 3

[11] Madhurima Guptaa. (2005) Custer ileal atresia associated with Hirschsprung disease (total colonic aganglionosis). Journal of Pediatric Surgery, 40, E5–E7.

[12] SW Moore. (1990). Intestinal atresia and Hirschsprung's disease. Pediatr Surg Int, 5, 182-184.

[13] JP Janik. (1997). Ileal atresia with total colonic aganglionosis. J Pediatr Surg, 32, 1502-7503

FIGURES:

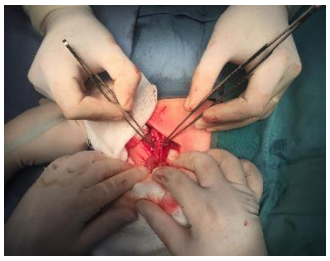

Fig1: intrauterine intussusception with the appendix

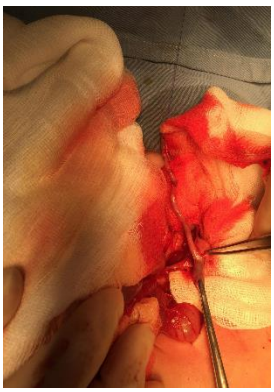

Fig2: ileal atresia

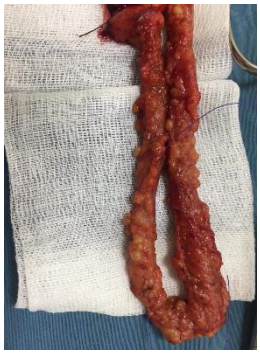

Fig 3: total colectomy