

Are Intussusceptions More Common and Severe in Malrotation? - Waugh Syndrome

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Abstract

Waugh syndrome-The association of intussusception and malrotation is often overlooked and underdiagnosed. Malrotation need to be diagnosed preoperatively (reversal of SMA and SMV axis on doppler) or atleast looked for intraoperatively. Here is reported a case of 8 month old male child brought with complains of pain abdomen associated with fever since 4 days and passing red current like stools since last 2 days. No history of preceding diarrhoea. Parents gave history of something coming out of rectum while crying in pain since one day. Physical examination revealed mild pallor, tachycardia with distended abdomen and decreased bowel sounds. Per rectal examination showed prolapsed congested mass. Child was taken for emergency right supraumbilical transverse laparotomy and revealed mobile ascending colon and duodeno-jejunal flexure on right side of midline, with a long intussusception of terminal ileum, appendix and cecum into transverse colon, descending colon and rectum. Standard reduction was tried and it was possible to reduce it till transverse colon. Enterotomy was made into mid transverse colon to facilitate reduction of small bowel. On reduction, terminal ileum, appendix and ascending colon were not viable and was resected. In view of sepsis, ileostomy and distal stoma were made. Child had uneventful post-operative course and stoma was reversed at 6 weeks.

Keywords: Surgery; Pediatric surgery; Gastrointestinal surgery; Waugh syndrome; Malrotation; Intussusception; Enterotomy

Introduction

Waugh syndrome is the name given to association of malrotation and intussusception. The incidence of malrotation is 1 in 500 live births, [1] with bilious vomiting and sometimes, acute abdomen as prime mode of clinical presentation. Although literature reports of intussusception having associated lack of normal rotation and fixation of the intestine (malrotation), in as high as 40% of cases, [2] this combination is not often reported [3]. We want to report this often overlooked clinical combination, more so in era of conservative management of intussusception, which can have profound effect on mortality and morbidity of child.

Case Presentation

An eight month old male child brought to pediatric surgery emergency with complaint of pain abdomen associated with fever since last three days and passing blood in stool since last two days. There was no history of preceding diarrhea. Parents gave history of something coming out of rectum while crying in pain since one day. Physical examination revealed mild pallor, tachycardia with distended abdomen and decreased bowel sounds. Per rectal examination showed prolapsed congested mass, and it was not possible to reposition it back (Figure 1). X-ray showed multiple air fluid levels (Figure 2). Hemoglobin was 9.2 g/dL and total leucocyte counts were mildly raised. Child was taken for emergency laparotomy which revealed mobile ascending colon and duodeno-jejunal flexure on right side of midline, with a long intussusception of terminal ileum, appendix and cecum into transverse colon, descending colon and rectum (Figure 3a).

Standard reduction was tried which was possible till transverse colon. Enterotomy was made into mid transverse colon, which facilitated reduction after resection of gangrenous intestine through enterotomy (Figure 3b). In view of multiple pus flakes over intestine, infected peritoneal fluid and sepsis in child, ileostomy and distal stoma were made along with primary repair of iatrogenic enterotomy. Mesentery was widened. Stoma functioned on day two and child had symptomatic improvement. Distal stoma washes were started from POD-14. At 6 weeks post-surgery, child underwent ileo-colic anastomosis and doing well in follow up.



Figure 1: Per rectal examination showing congested mass.

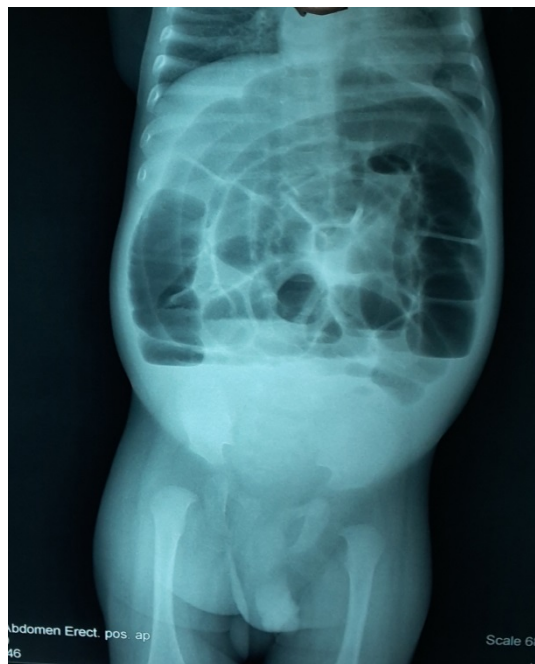


Figure 2: X-ray image examination showing multiple air-fluid interfaces.

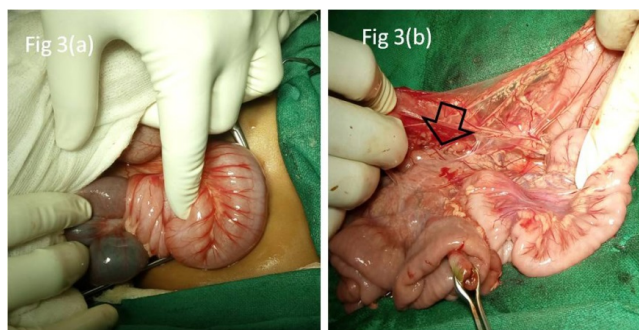


Figure 3: Intraoperative view.

Discussion

The association of malrotation and intussusception is referred to as Waugh syndrome by Brereton et al. [2] after George E Waugh, who first described the association in a report in 1911 [4]. It has been postulated that the presence of mobile ascending colon in cases of malrotation, predisposes for prolapse of terminal ileum and proximal colon into non fixed distal colon [5].

Malrotation has various clinical presentations, ranging from chronic abdominal pain to acute midgut volvulus with ischemic bowel injury. Child may present with bilious vomiting, diarrhea, failure to thrive and malabsorption. Our child was asymptomatic till this age for common clinical features of malrotation but intussusception lead to clinical presentation. The intussusception as a mode of clinical presentation in

malrotation is rare and in a recent series, it is reported that only 54 cases had been reported in literature till date [3].

Intussusception presenting as rectal prolapse has already been described in literature, [6,7] thus signifying the need to differentiate two entities. The features by which two can be differentiated are that the anal crypts are everted with rectal prolapse and not with intussusceptions. Also an examining finger can be passed between the prolapse and the anus in patients with intussusception but not in patients with rectal prolapse.

The reported incidence of Intussusception presenting as anal protrusion is 16% [8] to 29% [9]. However, how many of these with Intussusception and anal protrusion has associated malrotation is not known. On the other hand, in a series of eight cases of malrotation with Intussusception, two of them had anal protrusion of Intussusception [10].

Obiora et al. reported 10 cases of anal protrusion in 62 patients of intussusceptions [8]. The average time of presentation was 5.9 days in children who presented with anal protrusion, whereas our patient with anal protrusion has short history of three days only. It can be hypothesized that children with malrotation can have rapid progression of Intussusception and anal protrusion can be very early.

Upper gastro-intestinal dye study and CECT abdomen has been advised in literature to confirm the diagnosis of malrotation, but it is not possible in acute presentations as our case. Although we prescribed a ultrasound abdomen, but opinion was not possible due to excessive bowel gases.

Management of intussusception has evolved with more and more surgeons opting for non-operative means of reduction. USG and X-rays are used for imaging during this reduction, either by pneumatic or hydrostatic means. However, in acute presentations like of anal protrusion of intussusceptions, conservative managements by non-operative reductions are not likely to be effective and surgery is the best possible management.

Although, both USG and X ray with contrast can diagnose lack of normal rotation and fixation of the intestine during non-operative means of reduction, it is often overlooked and only few series have reported its incidence [2]. This can have disastrous consequences and child can present later with complications of malrotation [10]. It is suggested that whenever non operative reductions are done, malrotation must be looked into (reversal of SMA and SMV axis on Doppler) and if present, Ladd's procedure must be done.

Laparotomy and manual reduction of intussusceptions is the standard when non operative reduction is not possible. In our child, operative manual reduction was possible initially till splenic flexure but most part of transverse colon seemed salvageable. To avoid long bowel resection, we made an enterotomy in mid transverse colon and facilitated further reduction using fingers via enterotomy site, which allowed us to save length of transverse colon till hepatic flexure. Literature reveals cases of facilitated reduction via already perforated sites and allowing salvageability [11].

Conclusion

A suspicion of malrotation should always be kept in mind while dealing with infant acute abdomen. Waugh syndrome may be missed on hydrostatic or pneumatic reduction of intussusception but child can present later with features of malrotation or recurrent intussusceptions,

signifying the need of carefully looking for features of malrotation in all cases of intussusception. Intussusception appears to have rapid progression in children with malrotation. A proximal iatrogenic enterotomy can facilitate difficult reductions and allowing bowel length preservation.

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