Case Report

Chronic Intussusception Associated with Malrotation in a Child: A Variation of Waugh’s Syndrome?

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Chronic intussusception is a relatively uncommon disease most commonly observed in older children. Waugh’s syndrome represents a rare entity characterized by intestinal malrotation and acute intussusception. We report a very unusual case of intestinal malrotation associated with chronic intussusception. Clinical presentation, radiological findings, and managing of this association are discussed in the light of the available literature.

1. Introduction

Acute intussusception is a common emergency abdominal condition in children [1]. Common clinical patterns include intermittent abdominal pain, vomiting, and “currant jelly” bloody stool [2]. Intestinal malrotation is a congenital condition caused by abnormal rotation and fixation of the bowel [3]. While bilious vomiting is the most frequent symptom in neonates, in older children intestinal malrotation is most commonly associated with nonspecific symptoms, such as chronic abdominal pain, malabsorption, diarrhea, or constipation, which may delay diagnosis [4]. The association of acute intussusception with intestinal malrotation is known as Waugh’s syndrome (WS) [5]. Chronic intussusception is a distinct clinical entity, characterized by intermittent attacks of abdominal pain lasting more than 14 days; other symptoms of acute intussusception may not present. One impressive clinical feature is significant weight loss due to long-standing anorexia and vomiting [6].

Herein, we describe a rare case of chronic intussusception with intestinal malrotation that could possibly represent a variation of WS. A brief literature review of paediatric cases with WS is presented.

2. Case Report

A previously healthy 4.5-year-old boy was admitted to our department with a 6-week history of intermittent abdominal pain, poor appetite, sporadic nonbilious vomiting, and occasional constipation. A weight loss of five kilograms since the onset of symptoms was reported. On admission, the weight of the patient was 15 kg, as opposed to 20 kg six weeks earlier. Physical examination revealed a soft and mildly distended abdomen. A palpable, tender, round mobile mass was detected at the epigastrium. The white blood cell count was 11,000/μL (normal range, 4,500–9,900/μL), hemoglobin was 12.1 g/dL, and platelets were 420,000/μL. The serum chemistry profile was within normal limits, apart from C-reactive protein of 0.8 mg/mL (normal range, 0–0.5 mg/mL). Abdominal ultrasonography in transverse view revealed alternating hypoechoic and hyperechoic bowel walls suggesting the target sign (Figure 1). Hydrostatic reduction was attempted, without success (Figure 2).

Exploratory laparotomy through a right upper quadrant transverse incision revealed an ileocolic intussusception extending up to the transverse colon (Figure 3(a)). The duodenojejunal junction was found to be on the right of...
the superior mesenteric vessels; the ileocecal junction was freely mobile and the colon was suspended by primitive mesenteric folds. Furthermore, well-defined Ladd’s bands were seen to extend from the ascending colon to the posterior abdominal wall across the duodenum (Figure 3(b)).

The intussusception was manually reduced, and no leading point was found. Ladd’s procedure was also performed including appendicectomy. The child had an uneventful recovery. Six months after surgery, he weighed 22 kg (between 75th and 90th percentile).

3. Discussion
The incidence of chronic intussusception is about 3% of all cases of intussusception in children aged under one year and approximately 10% of children over that age [7]. The true
**Table 1: Published cases of Waugh's syndrome in the literature.**

| Number of studies | Author(s) | Journal | Sex | Age | Clinical onset | Type of intussusception | Type of intestinal malrotation | Outcome |
|-------------------|-----------|---------|-----|-----|----------------|--------------------------|---------------------------------|---------|
| 1                 | Waugh and Lond [8] | Lancet 1911;1:1492 | M (3) | 2.5–3 yr | Acute | Ileocecal (2) Ileocolic (1) | Nonrotation (3) | Successful |
| 2                 | Perrin | Br J Surg 1921;22:9-46 | N/A | N/A | Acute | N/A | N/A | N/A |
| 3                 | van Meurs | Br J Surg 1946;34:91 | M | 5 yr | Acute | Ileocolic | Nonrotation | Successful |
| 4                 | Peck | Surg Gyn Obstetr 1963;16:398 | N/A | N/A | N/A | N/A | N/A | N/A |
| 5                 | Tabibi | J Am Osteopath Assoc 1971;70:686 | M | 8 mo | Acute | Ileocecal | N/A | Successful |
| 6                 | Berry | South Med J 1972;65:1075 | M | 17 yr | Acute | Massive ileocolic | Mobile cecum and RC | Successful |
| 7                 | Stewart | Surgery 1976;7;9:716 | N/A | N/A | Acute | N/A | N/A | Successful |
| 8                 | Filston | J Pediatr Surg 1981;169 (Suppl):684 | M | 4.5 mo | Acute | N/A | Cecum in the RUQ | Successful |
| 9                 | Ornstein | J Pediatr Surg 1981;68:440 | M | 10 mo | Acute | Ileocecal | Volvulus, LB | Successful |
| 10                | Welch | Ann R Coll Surg Engl 1983;65:244 | N/A | N/A | N/A | N/A | N/A | N/A |
| 11                | Burke | Aust N Z J Surg 1985;55:73 | F | 3.5 mo | Acute | Ileocolic | Cecum at the level of duodenum-volvulus | Successful |
| 12                | Brereton et al. [9] | Br J Surg 1986;73:55 | N/A (15) | N/A | Acute | N/A | N/A | Successful |
| 13                | Jain | Arch Surg 1989;124:509 | F | 8 mo | Acute | Ileocolic | DJJ on the right of midline, LB, volvulus | Successful |
| 14                | Ward | Eur J Pediatr Surg 1992;2:239 | M | 3 mo | Acute | Recurrent ileocolic | Mobile cecum, DJJ right of midline, LB (AP) | Successful |
| 15                | Sarin | Indian Pediatr 1995;32:108 | M | 7 mo | Acute | Ileocolic | SHC | Successful |
| 16                | Lobo et al. [11] | Pediatr Radiol 1997;27:606 | N/A (2) | N/A | Acute | Ileocolic (2) | MGV (1), NFAC (1) | Successful |
| 17                | Breckon and Hadley [5] | Pediatr Surg Int 2000;16;370 | M (4), F (2) | 4–9 mo | Acute | Ileocolic (6) | IR | Successful |
| 18                | Luo | Pediatr Surg Int 2003;19:413 | M | 10 mo | Acute | Ileocolic | Small bowel on the RA | Successful |
| 19                | Inan | J Pediatr Surg 2004;39:110 | M | 8 mo | Acute | Ileocolic | Nonrotation | Successful |
| 20                | Rao | Indian J Pediatr 2005;72:221 | N/A | N/A | N/A | N/A | N/A | N/A |
| Number of studies | Author(s) | Journal | Sex | Age | Clinical onset | Type of intussusception | Type of intestinal malrotation | Outcome |
|-------------------|-----------|---------|-----|-----|----------------|--------------------------|-------------------------------|---------|
| 21                | Chirdan and Uba [10] | Nig J Surg Res 2005;7:159 | 5 M 3 F | 13–d–12 mo | Acute | Ileocolic (6) | DJJ to the right of midline (5) | Died (1) |
| 22                | Lukong    | S Afr J Surg 2007; 45:30 | M | 4 mo | Acute | Ileocolic | Successful |         |
| 23                | Rangel    | Med Univers 2007;9:141 | M | 6 mo | Acute | Ileocolic | Malrotation, LB | Successful |
| 24                | Dominguez-Pérez et al. [12] | Acta Pediatr Mex 2008;29:355 | M (5) | 2–6 mo | Acute | Ileocecal (5) | IR (5) | Successful |
| 25                | Al-Jandal | J Pediatr Surg 2009 44:E17 | F | 2.5 mo† | Acute | Colocolic | Nonrotation | Successful |
| 26                | Hardy     | Am Surg 2011;77:78 | M | 3 yr | Acute | Jejunojejunal | Malrotation | Successful |
| 27                | Nwankwo   | J Med Med Sci 2011;2:291 | N/A (2) | N/A | Acute | N/A | N/A | N/A |
| 28                | Baltazar  | J Surg Case Rep 2012;3:22 | F | 3 mo | Acute | Ileocolic | DJJ to the right of SMA, LB | Successful |
| 29                | Behera    | J Clin Diagn Res 2014;8:ND26 | M | 1yr | Acute | Ileocolic | Nonfixed rotation | Successful |
| 30                | Al-Momami | Ann Saudi Med 2014;34:527 | 7 (3 M, 4 F) | 4–11 mo | Acute | Ileocolic (5), ILCA (1), ILCR (1) | Malrotation (6), malrotation and volvulus (1) | Died (1) |
| 31                | Singh AP  | J Case Rep 2014;4:338 | M | 2 yr | Acute | Ileocolic | N/A | Successful |
| 32                | Natesan   | J Evol Med Dent Sci 2015:4:4040 | M | 5 mo | Acute | Ileocolic | MGV | Successful |
| 33                | Gil-Vargas (in press) | Cir Cir 2015 (In press) | M | 7 mo | Acute | Ileocolic | Abnormal fixation of the colon | Successful |
| 34                | Present case | M | 4.5 yr | Chronic | Ileocolic | DJJ to the right of the midline, LB | Successful |

M: male, () number of patients, yr: year, N/A: not applicable, mo: month, RC: right colon, RUQ: right upper quadrant, LB: Ladd’s bands, F: female, DJJ: duodenojejunal junction, IR: incomplete rotation, DS: duodenal stenosis, AP: annular pancreas, SHC: subhepatic cecum, MGV: midgut volvulus, NFAC: nonfixation of the ascending colon, RA: right abdomen, d: days, † corrected age 38 weeks, SMA: superior mesenteric artery, ILCA: ileocoloanal, and ILCR: ileocolorectal.
incidence of WS is not known [5]. A PubMed and Google Scholar search revealed 33 published studies that referred to 76 children with WS (age range, 13 days to 17 years) (Table 1). To our knowledge, the association of chronic intussusception with intestinal malrotation in children has never before been mentioned in the international literature and could represent a variant of WS.

Existing evidence suggests that intestinal malrotation may predispose to acute intussusception. Waugh and Lond originally suggested that an ascending and descending colon relatively unfixed to the posterior wall and freely suspended by its primitive mesenteric folds may provoke an ileocecal intussusception [8]. According to a study by Brereton et al. [9], the principal factor that allows the terminal ileum to pass into the cecum is abnormal fixation and rotation of the ileocecal mesentery, while Breckon and Hadley [5] suggested that a mobile right colon might predispose to intussusception. In our case, one could incriminate the possible role of the freely mobile ileocecal junction as a principal factor of chronic intussusception. Moreover, the primitive mesenteric folds which do not become sufficiently tense to occlude the mesenteric blood vessels may be the cause of longstanding recurrent abdominal symptoms without any further complications such as bowel necrosis.

Ultrasonography (U/S) and contrast enema both constitute reliable imaging tools in the diagnosis of acute intussusception [1]. In the case presented herein, U/S and barium contrast enema confirmed the diagnosis of chronic intussusception. That being said, radiologic evaluation was seen to offer a definite preoperative diagnosis in only seven reports of the reviewed cases of WS (Table 1, studies 12, 16, 18, 23, 25, 29, and 31).

In the majority of listed studies, open surgery (Table 1, studies 1–15, 17–22, 24, and 26–32) was the treatment of choice; a laparoscopic approach was performed in just one case (Table 1, study 25). Notably, in the studies by Breckon and Hadley [5] and Chirdan and Uba [10], intestinal malrotation was not taken into consideration during surgery for acute intussusception; hence, the patients were submitted to reoperation for recurrent symptoms of bilious vomiting. Furthermore, Lobô et al. [11] and Dominguez-Pérez et al. [12] reported one and three cases, respectively, which were managed conservatively. In our case of a possible variant of WS, surgery was necessary to reduce the intussusception, given that the attempted hydrostatic reduction had failed. The operation had a successful outcome. All but two cases of the reviewed studies (Table 1, studies 21 and 29) were seen to have successful results.

4. Conclusion

The association between chronic intussusception and intestinal malrotation has never before been reported in the literature. This coexistence may represent a possible variant of WS. In cases of chronic intussusception, a high degree of suspicion is warranted in order to guide towards the proper diagnosis.

Consent

Informed and written consent for publication of this case report was obtained from the patient.

Competing Interests

The authors declare that there is no conflict of interests regarding the publication of this paper.

References

[1] B. T. Bucher, B. L. Hall, B. W. Warner, and M. S. Keller, “Intussusception in children: cost-effectiveness of ultrasound vs diagnostic contrast enema,” Journal of Pediatric Surgery, vol. 46, no. 6, pp. 1099–1105, 2011.
[2] V. K. Pepper, A. B. Stanfill, and R. H. Pearl, “Diagnosis and management of pediatric appendicitis, intussusception, and Meckel diverticulum,” Surgical Clinics of North America, vol. 92, no. 3, pp. 505–526, 2012.
[3] M. S. Shalaby, K. Kuti, and G. Walker, “Intestinal malrotation and volvulus in infants and children,” British Medical Journal, vol. 347, Article ID f6949, 2013.
[4] N. Spigland, M. L. Brandt, and S. Yazbeck, “Malrotation presenting beyond the neonatal period,” Journal of Pediatric Surgery, vol. 25, no. 11, pp. 1139–1142, 1990.
[5] V. M. Breckon and G. P. Hadley, “Waugh’s syndrome: a report of six patients,” Pediatric Surgery International, vol. 16, no. 5–6, pp. 370–373, 2000.
[6] B. I. Rees and J. Lari, “Chronic intussusception in children,” British Journal of Surgery, vol. 63, no. 1, pp. 33–35, 1976.
[7] D. Macauley and T. Moore, “Subacute and chronic intussusception in infants and children,” Archives Disease in Childhood, vol. 30, no. 150, pp. 180–183, 1955.
[8] G. E. Waugh and B. S. Lond, “Referred penile pain in intussusception with notes of three cases,” The Lancet, vol. 177, no. 4579, pp. 1492–1494, 1911.
[9] R. J. Brereton, B. Taylor, and C. M. Hall, “Intussusception and intestinal malrotation in infants: Waugh’s syndrome,” British Journal of Surgery, vol. 73, no. 1, pp. 55–57, 1986.
[10] L. B. Chirdan and A. F. Uba, “Association of midgut malrotation with intussusception,” Nigerian Journal of Surgical Research, vol. 7, no. 1-2, pp. 159–161, 2005.
[11] E. Lobô, A. Daneman, J. M. Fields, M. S. Keller, D. J. Alton, and B. Shandling, “The diagnosis of malrotation during air enema procedure,” Pediatric Radiology, vol. 27, no. 7, pp. 606–608, 1997.
[12] S. T. Dominguez-Pérez, C. Baéza-Herrera, G. Jaimez, M. L. Martínez-Rivera, and T. González-Mateos, “Síndrome de Waugh. Promerors informes en México,” Acta Pediatr Mexicana, vol. 29, no. 6, pp. 355–358, 2008.