

CASE REPORT

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Presentations of Waugh's syndrome: intra-luminal cecal cyst and trans-anal prolapsing intussusception: a case report

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Abstract

Background Intussusception with intestinal malrotation is termed as Waugh's syndrome. The incidence of Waugh's syndrome is less than 1%. There are very few reported cases. Once presented, it is a pediatric surgical emergency.

Case presentation We present here two cases of Waugh's syndrome: an 11-month-old male patient of Punjabi descent and a 4-month-old female patient of Afghan descent who presented to us with abdominal pain and bleeding per rectum. Abdominal sonography revealed an intussusception with a target sign. They were explored and perioperatively had intestinal malrotation alongside intussusception, thus a diagnosis of Waugh's syndrome was made. A right hemicolectomy and Ladd's procedure was performed.

Conclusion Waugh syndrome is a rare congenital anomaly but can present with vague abdominal symptoms. Once presented, it is a pediatric surgical emergency. The patient should be optimized followed by surgical exploration.

Keywords Intussusception, Pediatric, Waugh syndrome

Introduction

Intussusception is a surgical emergency in which part of the gut telescopes into an adjacent part of the intestine [1]. It mostly occurs in the age range of 3 months to 3 years but can rarely occur in any age group. Patients usually present with a complaint of colicky abdominal pain with in-drawing of legs, red-currant jelly stool, and in some cases abdominal mass. Intussusception can be primary, where no lead point is observed, or secondary, due to a lead point. Waugh's syndrome is the association of intussusception with intestinal malrotation

[2]. Although intussusception is one of the most common causes of pediatric intestinal obstruction, there are very few reports on Waugh's syndrome. It was first reported in 1911 by George E. Waugh and named after him by Brereton *et al.* in their study [7]. The incidence of Waugh's syndrome is less than 1% in pediatric population [3]. Nonoperative management of intussusception may have masked many cases of Waugh's syndrome, owing to which data on this anomaly are scarce; to date, very few cases have been reported in literature [4].

We report herein two cases of Waugh's syndrome where the patient was diagnosed, operated upon, and managed at our facility. Both had uneventful postoperative course and were discharged home with instructions and remained on follow-up.

Case presentation

Case 1

An 11-month-old male child of Punjabi descent presented to us with complaint of loose stool from last

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5 days followed by excessive crying with in-drawing of legs and nongreenish vomiting from the last day with history of reflux since 3 months of life, for which he had multiple visits to local clinics and symptoms improved, and history of previous exploration for intussusception at 5 months of age. Perioperatively, previous exploration showed ileocecolic intussusception with edematous terminal ileum; manual reduction of intussusception and appendectomy was performed (Fig. 1).

On examination, the patient's vital signs were stable with soft abdomen and no distention, there was mild tenderness; on digital rectal examination, the patient passed watery stool with mucoid discharge.

X-ray of the abdomen was done and showed dilated gut loops, and ultrasound showed intussusception. The patient was explored following resuscitation, and basic laboratory investigations and perioperative ileocecolic intussusception was found, which was reduced manually; on reduction, a cecal mass was observed with clear fluid in it, which might have acted as a lead point, hence a limited right hemicolectomy was performed with anastomosis and specimen was sent for histopathology. Perioperatively, duodenojejunal junction was also observed to be on right side, hence Ladd's procedure was performed for intestinal malrotation. The postoperative course was uneventful, and the patient was discharged home on the sixth postoperative day with instructions and called for follow-up.

Case 2

A 4-month-old female child of Afghani descent presented with history of loose stool from the last 10 days, per-rectal bleed from last 6 days, and something coming out of anus (prolapsed intussusceptum) from last 3 days, and also vomiting from last 3 days, which initially contained milk but later turned greenish. As the patient presented

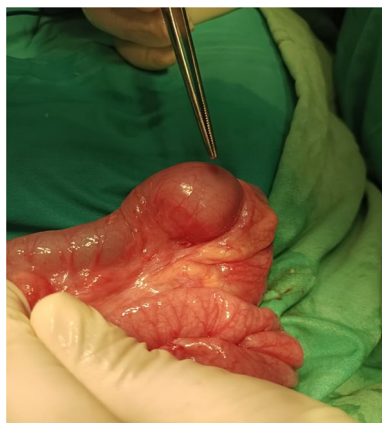


Fig. 1 Waugh syndrome with intraluminal cyst

from a rural area where proper medical facilities were not available and owing to nonaffordability, the patient presented very late. There was no previous significant medical or surgical history.

On examination, the patient was very sick-looking, dehydrated with tachycardia and hypotension, immediate resuscitation was started, and we were able to optimize the patient for surgery. Basic laboratory investigations were carried out, and blood was arranged; as history and clinical examination was enough, we did not send the patient for ultrasound or X-ray, and the patient was explored (Fig. 2). Perioperative findings were ileocecolic intussusception extending up to distal third transverse colon with gangrenous distal 13 cm of ileum and cecum, ascending colon perforated up to transverse colon, hence right extended hemicolectomy and end-to-end anastomosis was done.

Duodenojejunal junction was also observed to be on right side, and Ladd's procedure was also performed. Postoperatively, the patient remained admitted for 5 days. He was allowed oral intake after gut functions returned, followed by discharge with instructions. He remained on follow-up.

Discussion

Waugh's syndrome is the combination of intussusception with intestinal malrotation, first reported in 1911 by George E. Waugh regarding three patients who presented with such. Brereton later found that 40% of his patients presenting with intussusception had duodenojejunal flexure on right side (15 out of 37) and thus named the syndrome after Waugh, giving it the name of Waugh's syndrome [5]. The pathophysiology behind it is nonfixed cecum and ascending colon in children with malrotation, which provides an easy target to act as intussusception.

The age range of intussusception with or without malrotation is mostly between 3 months and 3 years, but it can present in any age group, as Waugh's syndrome has been reported in neonatal-age and



Fig. 2 Waugh syndrome with transanal prolapsing intussusception

school-age children too. The patients reported herein are also in the same age group as most others reported, that is, 11 months and 4 months [6].

The treatment of choice in Waugh's syndrome is manual reduction of intussusception plus straightening of gut and division of bands with widening of mesentery, but our reported cases presented with rare problems [7]. The first case, where the patient was 11 months old, was a male child who presented with recurrence of intussusception, thus not being an ideal candidate for nonsurgical management; we explored the patient and following manual reduction found cystic mass within cecum, which might previously have acted as a lead point, hence we carried out a limited hemicolectomy, removing a small part of the terminal ileum, cecum, and part of ascending colon, thus removing the lead point to limit further such episodes, and sent the specimen for histopathology. The other case also had an unusual presentation: a transanal prolapsing intussusception. He was also operated upon, with a laparotomy being performed. Peroperative findings showed ileocecolic intussusception extending up to distal third of transverse colon with gangrenous distal 13 cm of ileum and cecum, and ascending colon perforated, thus a right hemicolectomy was performed [8].

Whenever a case of Waugh's syndrome presents, it is deemed a surgical and diagnostic dilemma [9]. Owing to advancements in imaging technology and surgical knowledge, we have become wise regarding nonsurgical management for intussusception, but one should still bear Waugh's syndrome in mind for patients presenting with intussusception, as many cases go unnoticed because of this, while radiologists should be informed about also looking for malrotation in patients with intussusception to prevent recurrence [10].

Conclusion

Waugh syndrome is a rare anomaly but can present with vague abdominal symptoms. Once presented, it is a pediatric surgical emergency. The patient should be optimized followed by surgical exploration.

Supplementary Information

The online version contains supplementary material available at <https://doi.org/10.1186/s13256-024-04701-1>.

Supplementary Material 1.

Supplementary Material 2.

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Author contributions

Concept of study: MA, MH. Acquisition of data: HM, RR. Writing and drafting: MA, MH. Supervision: MAC.

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Data availability

The data that support the findings of this study are available from the corresponding author upon request to corresponding author.

Declarations

Consent for publication

Written informed consent was obtained from the patients' legal guardians for publication of this case report and any accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal.

Competing interests

The authors have no conflicts of interest relevant to this article to disclose.

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