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Waugh Syndrome in a One-Month Old Patient

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Abstract

Introduction: Waugh Syndrome, a rarely encountered problem, is a combination of two un-rare conditions, intestinal malrotation and intussusception. This is a case of Waugh Syndrome diagnosed and treated in less than 48 hours.

Case Description: A 1 month 10 days baby girl presenting for vomiting and fresh blood from rectum 5 days after undergoing inguinal hernia repair.

Discussion: About 100 cases of Waugh Syndrome have been reported since 1911 when this entity was first described. One theory explaining why it is being more discovered is that imaging techniques are improving and physicians are having a greater index of suspicion for such associations.

Conclusion: Intussusception cases must be carefully investigated for a possible association with underlying pathology including intestinal malrotation. Early surgical treatment can protect the patient from short- and long-term complications.

Keywords: Waugh syndrome; Intussusception; Intestinal malrotation; Pediatrics

Introduction

Intestinal malrotation is the congenital failure of the midgut to rotate counter-clockwise leading to the abnormal rotation of the Superior Mesenteric Artery (SMA) and placement of the duodeno-jejunal junction to the right midline. It usually presents during the neonatal period and it is treated surgically by Ladd procedure [1]. Intussusception on the other hand is the telescoping of one part of the bowel into the adjacent part. In the pediatric population, intussusception is usually idiopathic involving the ileocecal segment of the bowel [2].

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In rare circumstances intussusception is caused by midgut malrotation resulting in a combination known as Waugh syndrome. Historically, this combination was first described by George E. Waugh in 1911 who reported the first known 3 cases of 3 children who suffered from recurrent episodes of abdominal pain radiating to the suprapubic region presented for persistent vomiting with "abnormal persistence of primitive mesenteries" [3]. Conservative management methods such as air and hydrostatic enemas have higher failure rates if malrotation is associated to intussusception [4].

Subsequently, failure of non-operative techniques to reduce intussuscepted bowel in Waugh syndrome raise the need for urgent surgical intervention through laparotomy and manual reduction because if left untreated it can result in bowel perforation, purulent peritonitis and septic shock [5].

A theory explaining why intussusception occurs in patients with intestinal malrotation was proposed by Waugh and Lond suggesting that the relative instability of the ascending and descending colon predisposes to an ileocecal invagination [6].

In 76 reported cases of Waugh syndrome between 1911 and 2016, only one case showed chronic presentation of symptoms while the rest showed acute onset of vomiting abdominal pain and bloody stools; only 7 cases showed definite diagnosis by abdominal ultrasound; only four cases were treated conservatively while the rest required surgical intervention [7].

Here we present a case of Waugh Syndrome diagnosed and treated in a patient during the second month of life.

Case Presentation

History and presentation

A 1 month 10 days old baby girl born by normal vaginal delivery without any pre- or post-natal complications, presented to the emergency department of Bahman University Hospital for one episode of massive fresh blood from rectum (Figure 1). This was associated with one episode of projectile post-prandial vomiting. Family mentioned decrease oral intake several hours before presentation, with irritability and excessive crying that ended up with hypoactivity. History goes back to 4 days prior to presentation when the patient underwent a bilateral inguinal hernia repair due to a congenital bilateral inguinal hernia. The surgical approach was smooth and with no post-surgical complications.

On physical examination, baby girl was pale, hypoactive and lethargic. Mucosal membranes were dry reflecting moderate dehydration. Palpation of the abdomen showed a soft non-distended abdomen with absent bowel sounds to auscultation.

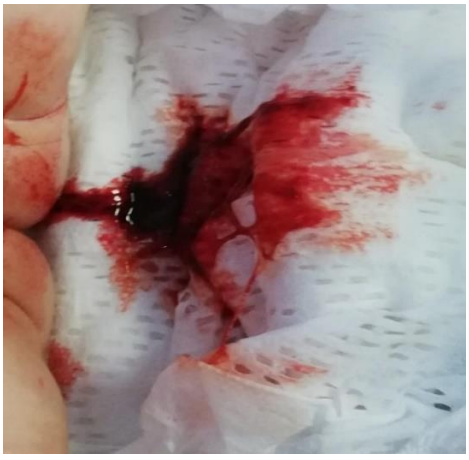

Figure 1: Fresh blood out of rectum on presentation.

Diagnostic focus and assessment

Several differential diagnoses were suggested including: intussusception, Meckel's diverticulum, and volvulus. Immediately, a standing abdominal X-ray was done and revealed signs of mechanical intestinal obstruction (Figure 2). Also, blood samples for hematologic and biochemistry workup were drawn out (Table 1 and 2).

Table 1: Hematology laboratory test results.

Haemoglobin (g/L)	11.8
White blood cells (cu/mm^3)	8950
Red blood cells (x1,000,000/mm^3)	4.11
Haematocrit (%)	35.2
Mean cell volume (fL)	85.5
Platelets (cu/mm^3)	408000
Neutrophil (%)	53.8
Lymphocyte (%)	35.9
Monocyte (%)	6.16
Eosinophil (%)	2.77
Basophil (%)	1.43
INR	1.2

Table 2: Biochemistry laboratory test results.

Sodium (mEq/L)	133
Potassium (mEq/L)	4.7
Chloride (mEq/L)	101
CO2 (mEq/L)	22
Creatinine (µmol/L)	0.18
CRP (mg/dL)	2.2

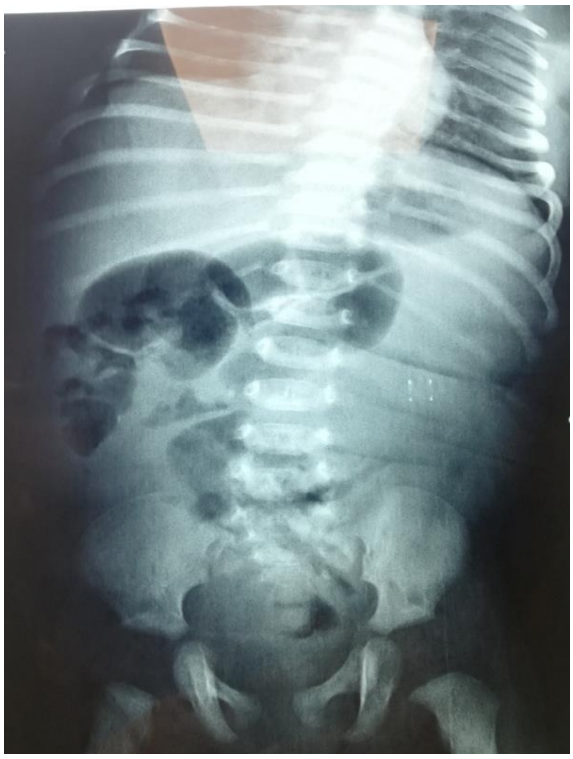

Figure 2: Standing abdominal X-ray showing intestinal obstruction.

For further elimination of other differential diagnosis, an abdominal echography was done and revealed at the splenic flexure a bowel invagination with wall thickening (peripheral hypoechoic ring with central echogenicity) suggesting intussusception (Figure 3). The fact that intussusception occurs mostly at the ileo-colic junction and thus at the right lower abdominal quadrant suggested the possibility of intestinal malrotation so a therapeutic and diagnostic gastrografin enema was done successfully reducing the intussusception and revealing an intestinal malrotation (Figure 4). Here, the diagnosis of the interesting Waugh Syndrome was established, a combination of intussusception and intestinal malrotation.

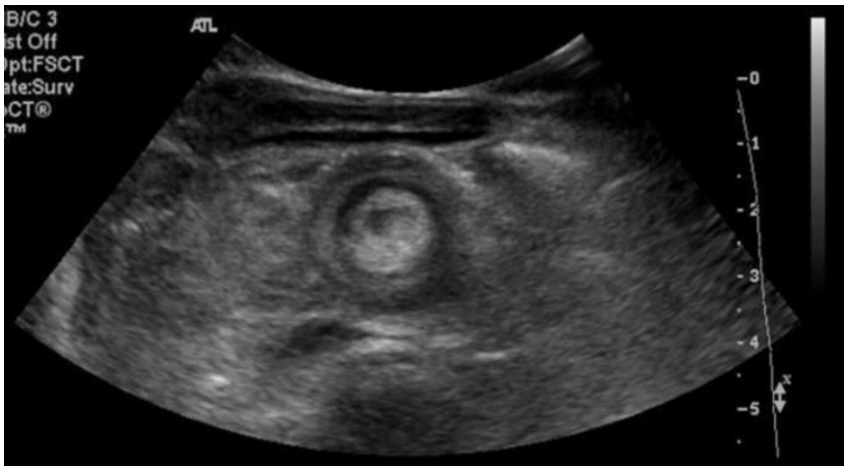

Figure 3: Abdominal echography showing target sign and pseudokidney sign.

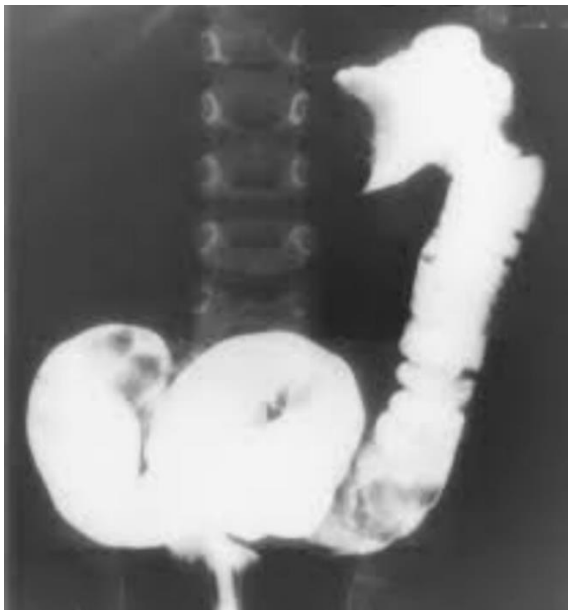

Figure 4: Abdominal X-ray post gastrografin enema showing intestinal malrotation.

Therapeutic focus and assessment

After confirming the diagnosis, patient was admitted to the hospital and intravenous antibiotics were started. Urgent surgical reduction was done few hours after admission due to recurrence of intussusception. Surgical report mentioned the following: laparotomy done, the transverse colon was black so excision of the necrotic part (partial colectomy) with anastomosis done. Also, reduction of intussusception was performed. The procedure also included correction of malrotation and positioning of the small intestine in the right and the colon in the left. In addition, appendectomy was done. Pathology specimens showed pattern consistent with infarct of the transverse colon wall tissue and inflammation of the appendiceal wall.

Antibiotics were continued after the surgical intervention. The baby passed stools normally and the bowels resumed its function. Two days after the operation feeding was started and was tolerated without vomiting. Patient finally discharged home and followed up regularly.

Discussion

Positioning of the midgut during the embryological life involves 4 phases: herniation, rotation, retraction and fixation. The first phase is the physiologic herniation of the midgut loop into extra-embryonic coelom due to its rapid growth when compared to the rest of the intestine. This is followed by the rotation phase where the intestine rotates a total of 270 degrees in a counterclockwise direction. This phase is also subdivided into three steps; the first one results in 90 degrees rotation of small intestine from a cephalic to a right-sided position and the cecum from a caudal to a left sided position and occurs outside the abdomen. This is followed by retraction of the intestinal loop into the abdomen and is accompanied by another 90 degrees counter-clock wise rotation movement. Subsequently the duodenojejunal junction acquires its final position at the left upper side and the cecum reaches its normal anatomic location at the right lower side of the abdomen. The midgut maturation will end up by the fixation of the ascending and descending colons into the posterior abdominal cavity [8].

Accordingly, we can divide intestinal malrotation into three types: The first type is reverse malrotation were the rotation occurred in a clockwise direction rather than counter-clock wise direction, the second form is the non-rotational one where no rotation took place, and the last form is incomplete rotation where the rotation of the intestines was arrested at 180 degrees counter-clock wise [6].

Intussusception is the invagination of one part of the intestine (proximal) into the adjacent part (distal). It usually involves the ileo-colic part of the intestine in the pediatric population. Epidemiologically, it is considered the most common abdominal emergency and the second most common cause of intestinal obstruction in the pediatrics population after pyloric stenosis. Most of the cases are considered idiopathic without identifiable cause and about 10% are due to a pathologic lead point [2]. The diagnosis can be made with both abdominal ultrasound and contrast enema, with ultrasound being more effective and less expensive than contrast enema [9]. Intussusception can be treated with air or hydrostatic enema in most cases and only few circumstances require laparotomy for reduction [2].

The combined presentation of intestinal malrotation and intussusception is known as Waugh Syndrome. The case reported in this paper lightens the importance of early surgical treatment that will end up in a better prognosis of the patient and safer outcome.

Conclusion

Waugh Syndrome is considered a rarely described entity in the literature, but it may be more frequently encountered than reported. Clinicians must have a high index of suspicion for Waugh Syndrome especially when facing a case of recurrent idiopathic intussusception without a clear pathological explanation. Early diagnosis and treatment will help the patient avoid further intussusception episodes and will improve the quality of life of both the patient and the family

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