A Rare Case Report of Waugh Syndrome in an Eight Months Old Male

Abstract: The author reports a case of Waugh syndrome in an 8-month-old baby boy. Although Waugh's Syndrome is rare, most of the documented cases are in children. Waugh syndrome, first described in 1991 by Georges Waugh, is an intussusception associated with abnormal bowel rotation (Behera, C. R., & Mohanty, S. K. 2014; & Henderson, A. A. et al., 2013). Less than 100 cases have been described so far worldwide. Intussusception is one of the most common causes of bowel obstruction in pediatrics. Although this intussusception is idiopathic in 81% of cases, it can sometimes be secondary to various etiologies such as Meckel's diverticulum, congenital bridles, gastroenteritis, polyposis and intestinal lymphomas. The authors discuss treatment options and discuss the probability of Waugh's Syndrome during any intussusception in children (Behera, C. R., & Mohanty, S. K. 2014).

Keywords: Waugh Syndrome, Intussusception, intestinal rotation malfunction.

INTRODUCTION:

Waugh Syndrome is the rare association of intussusception with abnormal bowel rotation (Behera, C. R., & Rezende Caino de Oliveira, F. et al., 2014). Waugh Syndrome is omitted if there is reduced intussusception and may be a reason for recurrence which may result in acute midgut volvulus with ischemic bowel injury. Although it is a rare entity, the likelihood of this syndrome should be kept in mind during hydrostatic reduction surgery for any recurrent intussusception (Behera, C. R., & Mohanty, S. K. 2014; & Henderson, A. A. et al., 2013). The objective of this observation is to report the case of an 8-month-old infant with Waugh Syndrome and to discuss its clinical presentation and management.

CASE DESCRIPTION:

This is a male infant, 8 months old, well vaccinated according to the national vaccination program, breastfeed and having no history or known allergy, referred by his pediatrician for a crying fit that lasted more than 20 minutes, 2 episodes of vomiting, lethargy and pallor. In his history, the pregnancy went well and the cesarean delivery was at term. The interrogation noted normal bowel movements, no trauma, no fever or recent illnesses. The admission examination found normal vital signs but poor general health, pallor and persistent lethargy. It should also be noted that since parents were not vigilant enough, they could not be relied upon as a trusted source of information, thus history was not accurate. Physical examination showed a bloated, soft abdomen with presence of bowel sounds, non-erythematous tonsils with normal cardiopulmonary auscultation.

How to Cite the Article:
The differential diagnoses were: dehydration, excessive head trauma, metabolic disease, food intolerance or intussusception. Biologically, the blood count showed a slight increase in white blood cells (14,000) with normal electrolytes, hepatic transaminases and blood sugar. The infant, hospitalized for monitoring and hydration, got admitted to the pediatric ward. The evolution of his condition was marked by the onset of rectal bleeding and recurrent vomiting which required urgent advice from a pediatric surgeon. Intussusception is diagnosed requiring hydrostatic reduction with a barium enema. A discovered intestinal malformation complicated the management, requiring surgical management on third day after the enema. The surgery consisted of excision of the flanges from the duodenum to the coecum, an anastomosis going from the Treitz angle to the right colon with appendectomy. The postoperative follow-up went well. The child resumes feeding 48 hours after surgery and returns home after the surgeon's advice (Ladd procedure). The intussusception accompanied by a malformation suggests Waugh Syndrome.

**DISCUSSION:**

The age of manifestation of Waugh Syndrome is very variable and ranges from 13 days to 17 years (Henderson, A. A. *et al.*, 2013). However, cases of Waugh in adults have been reported. Clinically, the person has acute intussusception and general gastrointestinal symptoms (nausea, vomiting, periodic abdominal pain, distended and tender abdomen). Subsequently, the patient may develop rectal bleeding and sometimes dyspnea in children. The abnormal rotation that accompanies this clinical presentation is usually discovered later during exploration or surgery (Henderson, A. A. *et al.*, 2013; Tackett, J. J. *et al.*, 2014; & Saxena, R. *et al.*, 2015). The pathophysiology involves ileocolic prolapse in the unbound region, an abnormal rotating ascending colon found in children and poorly attached to the retro peritoneum, and invagination of the descending colon and rectum compromising the vascularization of the intestine hence the urgency of surgical management (Henderson, A. A. *et al.*, 2013). In Waugh Syndrome, anal crypts are associated with rectal prolapse and not with simple intussusception (Tackett, J. J. *et al.*, 2014). A rectal examination between the prolapse and the anus in patients presenting with intussusception and without a rectal prolapse may guide
the doctor in his/her diagnosis (Saxena, R. et al., 2015). Our infant's abnormal bowel rotation was asymptomatic until the age of eight months apart from the occasional bout of crying noted by the mother and episodes of diarrhea. It was the intussusception and acute clinical presentation that led the parents to the consultation. Although the clinical presentation of Waugh Syndrome is marked by a tender and bloated abdomen, ultrasound remains the imaging modality of choice to confirm the diagnosis and exclude simple intussusception (Khan, Y. A. et al., 2017). The classic image of intussusception on ultrasound is a "bull's eye" or "coiled spring" lesion representing layers (Baltazar, G. et al., 2012). The radiological signs compatible with intussusception are those of intestinal obstruction (hydro-aeric levels, distended intestine with absence of intestinal ventilation). In addition, the barium enema shows a classic image of a "coiled spring" with obstacle to the contrast product at the level of the invagination and localization of the coecum. In 80 to 87% of Waugh Syndrome cases proven surgically the barium enema was abnormal (Singh, A. P. et al., 2015). The abnormal rotation has a variety of clinical presentations, ranging from abdominal pain to acute midgut volvulus with ischemic bowel injury. The child may present bilious vomiting, diarrhea, growth retardation and malabsorption (Khan, Y. A. et al., 2017). Patients with intussusception are optimally treated with air insufflation or barium enema in pediatrics. In adults, surgery is absolute, especially if the patient has unmistakable peritonitis and the intussusception cannot be reduced manually (Ahsino, F. 2016). An anastomosis is performed. In our case, manual reduction by compression of the colon was not possible because intussusception was irreducible and end-to-end anastomosis resection was performed. At the time of operation for this dual condition, the diagnosis was made and confirmed by the location of the coecum and the pathognomonic presence of the peritoneal flanges of the ascending colon through the duodenum. In addition, a laparoscopic approach to Waugh Syndrome has also been recently developed (Hardy, D. et al., 2011).

**CONCLUSION:**

Waugh's Syndrome or the association of abnormal bowel rotation with intussusception should be considered in the management of any recurrent intussusception. Abnormal rotation and poor fixation of the intestine and its mesentery are an important factor in the etiology of idiopathic intussusception with risk of recurrence associated with non-surgical methods (Khan, Y. A. et al., 2017; & Behera, C. R., & Mohanty, S. K. 2014). Misdiagnosis can increase morbidity and mortality (Khan, Y. A. et al., 2017).

**REFERENCES:**