

# Chronic Ileocecal Intussusception Secondary to Non-Hodgkins Lymphoma

Wamalwa A<sup>1</sup>, Siwo EA<sup>2</sup>, Mburugu PM<sup>2</sup>, Mohamed M<sup>2</sup>

1. Garissa University College
2. Provincial General Hospital, Garissa

**Correspondence to:** Dr Alex Wamalwa, P.O Box 1801-70100, Garissa. Email: aleoke@gmail.com

## Summary

Chronic intussusception is a distinct clinical entity which is unfortunately poorly recognized due to its atypical presentation. A 5½ year old female patient presented with a 5 month history of abdominal pain associated with occasional vomiting, anorexia and progressive weight loss. 5 days prior to admission she developed loose, non-mucoid, non-blood stained stool associated with refusal to feed and irritability. Systemic examination, an abdominal ultrasound and abdominal radiograph were non-contributory. During treatment for malnutrition and dehydration she was noted to clinically improve, however, 6 days post-admission she developed abdominal pain, a palpable sausage-shaped abdominal mass, and currant-jelly stool. Emergency laparotomy revealed an ileocecal intussusception with oedematous non-

viable, aperistaltic terminal ileum and mobile cecum, a cecal mass and mesenteric lymphadenopathy was found. We performed a right hemicolectomy with an ileocolic anastomosis and mesenteric lymph node sampling. Histology results revealed the cecal mass as a lymphoproliferative neoplasm with a diagnostic consideration of intermediate to large cell Non-Hodgkins Lymphoma; and reactive hyperplasia of mesenteric lymph nodes. There is need for a high index of suspicion in children with non-specific abdominal symptoms for which no cause can be found. Further, all suspicious masses should have immunohistological histological evaluation.

**Key Words:** Chronic Ileocecal Intussusception, Non Hodgkins Lymphoma

## Introduction

Intussusception, which is the telescoping or invagination of a proximal portion of intestine (intussusceptum) into a more distal portion (intussusciens), is one of the most common causes of bowel obstruction in infants and toddlers. The acute type is more frequently diagnosed due to its classical symptomatology of abdominal pain, a palpable sausage-shaped abdominal mass, and currant-jelly stool (1,2). Chronic intussusception, on the other hand, presents no such typical picture and is often vague and the signs of intestinal obstruction are frequently lacking (3). Ultrasonography is used in the diagnosis of intussusception; with a sensitivity and specificity of up to 100% (4). Stable patients with a high clinical suspicion and/or radiographic evidence of intussusception and no evidence of bowel perforation should be treated with non-operative reduction. When an intestinal mass is encountered during surgery; histological evaluation

is required to exclude the possibility of malignancy (5). We describe a rare case of symptomatic cecal non-Hodgkin's lymphoma associated with chronic ileocecal intussusception; that resulted in severe malnutrition. We reviewed some aspects of the diagnosis and treatment.

## Case Presentation

A 5 ½ year old female child with no notable prior medical or surgical history presented to the pediatrics team with a 5 month history of abdominal pain that was of insidious onset, generalized, colicky in nature, with intermittent episodes of severe intensity pain especially after feeding. This was associated with occasional non-projectile, non-bilious vomiting and marked weight loss. Five days prior to admission she developed loose, non-mucoid, non-blood stained stool associated with refusal to feed and irritability. On examination she was sick looking, in poor nutrition (wasted), had mild pallor, was severely dehydrated

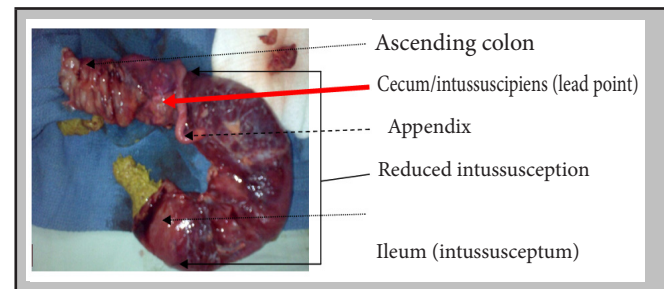
and weighed 12.5 kg. She was hypothermic at 35.4 °C, had tachycardia of 124beats per min and tachypnea of 30 breathes per min. The abdomen was soft, non-tender with no palpable masses. Other systems were normal. She was subsequently put on management for severe malnutrition and severe dehydration due to acute childhood diarrhea.

Complete blood count indicated anaemia of 8.9g/dl, normal white blood count and thrombocytosis of  $548 \times 10^3/\mu\text{L}$ . Biochemistry revealed high creatinine of 203.56 with the other indices being within normal limits. Two days post-admission the surgical team was called to review after her abdomen was found to have visible peristalsis. The surgical review found non-contributory findings. A digital rectal exam was normal. Abdominal ultrasound findings indicated dilated bowel loops throughout the abdomen with reduced motion and localized free fluid at both lumbar regions. Abdominal x-ray was normal. We decided to continue with pediatric management as her diarrhea stopped soon after the review and she started feeding normally with noted general improvement within 2 days.

Four days after initial surgical review, the surgical team was called to review the patient. Examination revealed a sick looking, irritable child in obvious pain, with a distended abdomen and visible peristalsis. On palpation, there was a non-tender, oval-sausage shaped mass at the right iliac fossa region and the bowel sounds were increased in frequency and intensity. On digital rectal examination; the anal tone was normal with a palpable non-tender mass in the right iliac fossa. There was currant-jelly stool on the examining finger. She was put on nil per oral, and had a nasogastric tube and urethral catheter inserted. She was also started on intravenous ceftriaxone and metronidazole.

Once stabilized, she underwent a laparotomy. Intra-operative findings were an ileocecal intussusception with oedematous non-viable, aperistaltic terminal ileum and mobile cecum; a cecal mass and mesenteric lymphadenopathy. A right hemicolectomy with an end-to-side ileocolic anastomosis was performed. The mesenteric lymph node were sampled for histology. Post operatively, she was transfused blood, started oral sips on the 4th day and graduated to normal feeds by the 7th day. A pediatric consultation facilitated provision of essential micronutrients and fortified feeds (6). Prompt weight gain was subsequently noted. Histology results revealed the cecal mass as a lymphoproliferative neoplasm with a diagnostic consideration of intermediate to large cell

Non-Hodgkins Lymphoma; and reactive hyperplasia of mesenteric lymph nodes. The child was referred to a pediatric oncologist for chemotherapy, under whose care she currently is.



## Discussion

Intussusception is the most common abdominal emergency in early childhood, particularly in children younger than two years of age (7). The incidence of intussusception is 1.5-4 cases per 1000 live births, with a male-to-female ratio of 3:2. In approximately 25 percent of cases, an underlying disease causes a pathological lead point for the intussusception, which may be focal or diffuse. These triggers account for a greater proportion of cases of intussusception in children younger than three months or older than five years (8,9). Some of the conditions which have been associated with intussusception, include Meckel diverticulum, polyps, small bowel lymphoma (10–12), duplication cysts, vascular malformations, inverted appendiceal stumps, parasites (eg, *Ascaris lumbricoides*), Henoch-Schönlein purpura, cystic fibrosis, and hemolytic-uremic syndrome.

The distinct clinical entity of chronic intussusception is poorly recognized and rarely described (13). Rafinesque (1878) classified intussusception into 4 types: (a) hyper acute (dying within 2 days of onset), (b) acute (dying within a week), (c) sub-acute (surviving for 7-14days) and (d) chronic (surviving for more than 14 days). Chronic intussusception never completely reduces and the variation in severity of the illness can be explained by the intermittent further progression of the intussusception. The peritoneal attachments usually have time to stretch, and as a result, the pull of the mesentery is less marked and the contractions of the bowel are less violent; consequently there may be little or no circulatory disturbance. Oedema, necrosis and gangrene are infrequently seen.

A review of chronic intussusception in children found that the only reliable clinical feature being history of repeated attacks characterized by sudden

onset of gripping abdominal pain. All other signs may be absent and are unreliable (14). Weight loss has also been found as a prominent feature of chronic intussusception (14). In our case, there was evidence of malnutrition which could be attributed to prolonged anorexia and vomiting. The history also indicated intermittent attacks of abdominal pain. Vomiting is relatively less common in chronic than in acute cases. Abdominal tenderness is not marked and may be absent. Our patient exhibited the classic triad of symptomatology when the intussusception was irreducible. We report a delay in diagnosis of our case; as has also been previously noted over the years since the description of chronic intussusception (15,16). The atypical presentation of chronic intussusception may be responsible for delay in diagnosis.

Ultrasonography is the method of choice to detect intussusception (17). The presence of ascites and long segments of intussusception can be used as sonographic predictors of failure for non-operative management (18). In our case, the ultrasound image was not of the classical "bull's eye" or "coiled spring" lesion; rather it only revealed the presence of ascites. In our set up, we lacked the facilities and the experience in contrast enema and CT scan as diagnostic means.

Surgical treatment is indicated as a primary intervention for patients with suspected intussusception who are acutely ill or have evidence of perforation. Surgery also may be appropriate when the patient is treated in a location where the radiographic facilities and expertise to perform non operative reduction are not readily available. We also lacked the experience in enema-based reduction of intussusception. Other considerations also made at deciding on operative management were a presentation of severely ill child with an acute intussusception; albeit at its late stage. We advocate for operative reduction and resection in cases of chronic intussusception to avoid the possibility of missing any predisposing lesion. The finding of a long segment of ischemic intussusception necessitated a right hemicolectomy. We further ensured a histologic assessment of the cecal mass was undertaken. The timely diagnosis of non-Hodgkin's lymphoma facilitated prompt referral and institution of appropriate chemotherapy. It was evident that the clinical symptoms had severely affected the child's feeding habits; resulting in severe malnutrition. After

correction of the underlying disease process, normal feeding habits were noted and a return normal weight was achieved within a short period of time.

We advocate for a high index of suspicion of chronic intussusception in children with non-specific abdominal pain, vomiting and weight loss. It is hoped this article will draw attention to chronic intussusception as a real clinical entity.

### **Conclusion**

Despite the obscure and indefinite symptomatology, careful history and physical examination often guides to an accurate diagnosis in chronic intussusception. We further recommend histological evaluation of all suspicious masses to ensure early diagnosis and treatment of tumors found acting as lead points in cases of intussusception. A multidisciplinary approach in the management of childhood intussusception is recommended to facilitate a quick recovery.

### **References**

1. West KW, Stephens B, Vane DW, et al. Intussusception: current management in infants and children. *Surgery*. 1987;102(4):704-10.
2. Yamamoto LG, Morita SY, Boychuk RB, et al. Stool appearance in intussusception: assessing the value of the term 'currant jelly'. *Am J Emerg Med*. 1997;15(3):293-8.
3. Reisman Ha, Wolk Ad. Chronic Intussusception. *Pediatrics*. 1949;4(2):183-7.
4. Burke LF, Clark E. Ileocolic intussusception—A case report. *J Clin Ultrasound*. 1977;5(5):346-7.
5. Akyıldız H, Biri I, Akcan A, et al. Ileal lipoma: case report. *Erciyes Med J*. 2011;33(1):83-6.
6. World Health Organization. Pocket book of hospital care for children: guidelines for the management of common illnesses with limited resources. 2005 [cited 2015 Feb 7]; Available from: <http://apps.who.int/iris/handle/10665/43206>
7. Walker WA. Pediatric gastrointestinal disease: pathophysiology, diagnosis, management [Internet]. PMPH-USA; 2004 [cited 2015 Feb 7].
8. Blakelock RT, Beasley SW. The clinical implications of non-idiopathic intussusception. *Pediatr Surg Int*. 1998;14(3):163-7.
9. Navarro O, Daneman A. Intussusception. Part 3: Diagnosis and management of those with an identifiable or predisposing cause and those that reduce spontaneously. *Pediatr Radiol*.

- 2004;34(4):305–12; quiz 369.
10. Abbasoğlu L, Gün F, Salman FT, et al. The role of surgery in intraabdominal Burkitt's lymphoma in children. *Eur J Pediatr Surg Off J Austrian Assoc Pediatr Surg Al Z Kinderchir.* 2003;13(4):236–9.
  11. Brichon P, Bertrand Y, Plantaz D. Burkitt's lymphoma revealed by acute intussusception in children. *Annales de chirurgie [Internet].* 2001 [cited 2015 Feb 7]. p. 649–53. Available from: <http://europepmc.org/abstract/med/11676236>
  12. Gupta H, Davidoff AM, Pui C-H, et al. Clinical implications and surgical management of intussusception in pediatric patients with Burkitt lymphoma. *J Pediatr Surg.* 2007;42(6):998–1001.
  13. Rees BI, Lari J. Chronic intussusception in children. *Br J Surg.* 1976;63(1):33–5.
  14. Garvie JM, Kemp FH. The diagnosis of chronic intussusception in children. *Arch Dis Child.* 1945 ;20(102):73–80.
  15. Thomasson B, Sutinen S. Chronic primary intussusception in an infant. *J Pediatr Surg.* 1972;7(3):299–301.
  16. Hays DM, Geller F, Norris W, et al. A Review of the Management of Intussusception in a Pediatric Center (1938-1958): Three Hundred Eighty-Two Consecutive Cases. *AMA Arch Surg.* 1960;80(5):788–805.
  17. Ko HS, Schenk JP, Tröger J, et al. Current radiological management of intussusception in children. *Eur Radiol.* 2007;17(9):2411–21.
  18. Munden MM, Bruzzi JF, Coley BD, et al. Sonography of pediatric small-bowel intussusception: differentiating surgical from nonsurgical cases. *Am J Roentgenol.* 2007;188(1):275–9.