

Ileo-Ileal Intussusception caused by an Inverted Meckel's Diverticulum Eliciting a Systemic Vasculitis-Like Response

Avinoam Nevler MD^{1,2}, Sagi Arieh Shpitzer BMedSc¹, Daniel Shinhar MD³ and Ron Bilik MD³

¹Department of Surgery and Transplantation, Sheba Medical Center, Tel Hashomer, Israel

²Borenstein Talpiot Medical Leadership Program 2012, Sheba Medical Center, Tel Hashomer, Israel

³Department of Pediatric Surgery, Safra Children's Hospital, Sheba Medical Center, Tel Hashomer, Israel

All affiliated with Sackler Faculty of Medicine, Tel Aviv University, Tel Aviv, Israel

KEY WORDS: intussusception, vasculitis, juvenile idiopathic arthritis (JIA), Meckel's diverticulum

IMAJ 2015; 17: 646-647

Intussusception is the most common cause of bowel obstruction in children, especially under age 2 years. The prompt diagnosis and treatment of intussusception is hugely important, since a delay in diagnosis entails greater morbidity and the need for more invasive treatment. We present here the case of a 15 year old adolescent in whom the final diagnosis of intussusception was delayed due to a highly unusual vasculitis-like presentation that included fever, pericardial effusion and a seizure, ultimately resulting in the need for surgical intervention and small bowel resection.

PATIENT DESCRIPTION

A 15 year old Caucasian male presented to the emergency department (ED) with severe nausea and vomiting, hypertension and mild facial swelling. His medical history was unremarkable except for an acute event of polyarticular juvenile idiopathic arthritis (JIA) at the age of 13, and he took no regular medications. His vital signs included blood pressure of 182/109 mmHg, heart rate 72 beats/min and body temperature 37.1°C which rose to 40.1°C during his stay in the ED. Physical examination

revealed a 2/6 systolic heart murmur. The abdomen was soft with no signs of peritonitis and there were no signs of arthritis or rash.

Laboratory tests showed hyponatremia (127–131 mEq/L), leukocytosis (18.7 K/ μ l), elevated troponin-I (0.11 μ g/L), slightly elevated C-reactive protein levels (6.7 mg/ml) and microhematuria. Lactate levels were within normal range. Abdominal X-ray and abdominal ultrasound demonstrated several distended edematous small bowel loops and a moderate amount of abdominal fluid. Cardiac sonography was performed, revealing a hyperdynamic heart with mild aortic insufficiency, mildly hypertrophic left ventricle and mild pericardial effusion. After completing the cardiac assessment the patient returned to the ED where he experienced an absence-like seizure lasting several minutes, followed by a 10 minute post-ictal period. Capillary blood glucose was 128 mg/dl and a computed tomography (CT) scan of his head revealed no acute findings.

Due to the complex clinical presentation of serositis, possible nephritis, bowel edema and seizures, a systemic vasculitis like syndrome was suspected and the patient was admitted to the pediatric intensive care unit for stabilization, corticosteroid treatment and further assessment and treatment. Additional laboratory studies performed at this time were negative for serum antinuclear antibodies (ANA) and anticytoplasmic antibodies (ANCA). C3 levels were mildly decreased (74–80 mg/dl) while C4 and

C1-esterase inhibitor levels were within normal range. An abdominal CT angiography was performed which showed an ileo-ileal intussusception with normal abdominal vasculature [Figure 1A].

The patient was taken to the operating room and was found to have a severely edematous obstructed small bowel due to ileo-ileal intussusception caused by an inverted Meckel's diverticulum as the lead point [Figure 1B]. Due to areas of bowel necrosis in the intussusceptum, the segment was resected and primarily anastomosed. Post-operatively, all signs and symptoms apparent at presentation had subsided, no further seizures occurred, blood pressure returned to normal, troponin-I levels normalized, and a repeat urinalysis was devoid of red blood cells and leukocytes.

Follow-up echocardiography on post-operative day 3 showed a reduction in the size of the pericardial effusion. The patient gradually returned to oral intake and was discharged on the 8th day post-surgery. At the follow-up visit 1 month after the surgery, he reported a relapse of the juvenile idiopathic arthritis in the lower limbs bilaterally and was referred to his treating rheumatologist.

COMMENT

Intussusception is a result of the invagination of a proximal bowel segment into the lumen of a more distal segment. The majority of intussusceptions are of an idiopathic nature, while in some a pathologic

Figure 1. [A] Coronal section of abdominal CT showing intussusception (white arrow)



Figure 1. [B] Surgical specimen showing resected section of necrotic small bowel with Meckel's diverticulum



lead point such as a Meckel's diverticulum or an enlarged lymph node is identified. The classic findings include abdominal pain, vomiting, a palpable abdominal mass and currant-jelly stool [1]. These findings are not always present and at times are shadowed by other symptoms, obscuring and delaying the correct diagnosis.

In our case the patient arrived at the emergency department with relatively common and non-specific complaints of nausea and vomiting, but had additional findings of elevated blood pressure, micro-hematuria and pericardial effusion, and during his stay in the ED also spiked a fever and had a seizure. The combination of these features together with the patient's history of an autoimmune disease led the treating physicians to the presumptive diagnosis of a vasculitic disease, which could explain most of his symptoms. His initial complaints of nausea and vomiting were attributed to mesenteric vasculitis, which could be supported by the bowel edema shown on ultrasound. Another possible diagnosis considered was hereditary angioedema which may present with bouts of abdominal pain and bowel edema leading up to bowel obstruction [2]. In this case, however, complement levels did not

exhibit the typical decrease in C3, C4 and C1-esterase inhibitor. Intussusception is a well-known complication of vasculitides such as Henoch-Schonlein purpura [3] and has occasionally been linked to other rheumatic conditions such as Kawasaki disease and lupus erythematosus. In these cases intussusception usually arises due to mesenteric lymphadenopathy which provides a pathological lead point. In our case the intussusception was caused by a preexisting medical condition – a Meckel's diverticulum that was inverted and was not related to a systemic vasculitic process. This leads us to believe that the patient's symptoms (including pericardial effusion, seizure and possible glomerulonephritis) were perhaps a complication of the intussusception and the major physiological stress it caused. We did not find a connection in the literature between JIA and intussusception.

There are several reports of intussusception accompanied by neurological symptoms [4] as in our case. Seizures in some of these cases were caused by hyponatremia, secondary to third spacing in bowel obstructions, although in our case serum sodium was only mildly decreased. An additional point of interest in this case is the lack of abdominal pain and abdominal findings on initial physical examination. Despite being one of the three key symptoms in intussusception, there are descriptions of painless intussusceptions that are associated with poorer outcomes and greater need for bowel resection, such as in our case [5].

CONCLUSIONS

Intussusception is a diagnosis that requires prompt recognition and treatment in order to avoid excess morbidity and the need for surgical treatment. In our case, the myriad of symptoms and findings, some of which are not typical of intussusception, were confounders which delayed the correct diagnosis. In addition to knowing the classical symptoms and presentation of intussusception, the prudent clinician must also consider a diagnosis of intussusception in more complicated and atypical cases such as the present case.

Correspondence

Dr. A. Nevler
 Dept. of Surgery and Transplantation, Sheba Medical Center, Tel Hashomer 52621,
Phone: (972-3) 530-2247
Fax: (972-3) 530-8157
email: Dr.Nevly@gmail.com
 avinoam.nevler@sheba.health.gov.il

References

1. Beasley SW, Auldism AW, Stokes KB. The diagnostically difficult intussusception: its characteristics and consequences. *Pediatr Surg Int* 1988; 3: 135-8.
2. Zingale LC, Zanichelli A, Deliliers DL, et al. Successful resolution of bowel obstruction in a patient with hereditary angioedema. *Eur J Gastroenterol Hepatol* 2008; 20: 583-7.
3. Peru H, Soylemezoglu O, Bakkaloglu SA, et al. Henoch Schonlein purpura in childhood: clinical analysis of 254 cases over a 3-year period. *Clin Rheumatol* 2008; 27: 1087-92.
4. Ko SF, Lee TY, Ng SH, et al. Small bowel intussusception in symptomatic pediatric patients: experiences with 19 surgically proven cases. *World J Surg* 2002; 26: 438-43.
5. Ein SH, Stephens CA, Minor A. The painless intussusception. *J Pediatr Surg* 1976; 11: 563-4.