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Case Report

Intussusception associated with celiac disease in a child: case report and review of literature

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ABSTRACT

Intussusception and celiac disease both are common in children. Intussusception has been rarely associated with celiac disease in children, which mostly involved small bowel, are transient without classical symptoms, reduced spontaneously and rarely needs intervention. Authors are reporting a 2-year girl who presented initially as gastroenteritis and later on intestinal obstruction and on investigation revealed ileo-ileal intussusception which resolved spontaneously during laparotomy preparation. The patient had growth retardation and microcytic hypochromic anemia and on further evaluation diagnosed with celiac disease. This case report highlights the need of celiac testing in children with intussusception, and such intussusception resolved spontaneously with gluten free diet and rarely needs any active intervention, thus preventing from undue nonsurgical or surgical interventions for intussusception.

Keywords: Celiac disease, Children, Gluten free diet, Intussusception

INTRODUCTION

Intussusception is the most common cause of intestinal obstruction between 5 months and 3 years of age and the most common abdominal emergency in children younger than 2 years. Approximately 90% cases of intussusception in children are idiopathic.[1] Celiac disease (CD) is an immune-mediated chronic inflammatory systemic disorder elicited by gluten and related prolamins found in wheat, rye and barley in genetically susceptible individuals and characterized by the CD specific antibodies, enteropathy and variable combination of clinical manifestations.[2]

Intussusception and CD has been reported in association in adults in many case reports and rarely in children and adolescents also.[2-4] Authors are reporting a 2-year-girl presented initially as gastroenteritis followed by intussusception and intestinal obstruction for more than 48 hours and resolved spontaneously during laparotomy. She was later diagnosed with CD when evaluated for concomitant malnutrition and anemia. This case report highlights the need of celiac testing in children with intussusception, and such intussusception resolved spontaneously with gluten free diet and rarely needs any active intervention, thus preventing from undue nonsurgical or surgical interventions.

CASE REPORT

A 2-year-girl had history of fever, diarrhea, abdominal pain and distension for 3 days. Stool examination showed

3-5 polymorphonuclear leucocytes/ hpf with undigested starch and fat globules. Ultrasound revealed fluid filled distended bowel loops. She was treated initially as gastroenteritis, but later developed constipation and bilious vomiting since last 2 days. There was no history suggestive of blood in stool. Her weight was 8.8 kg (below 3rd percentile of weight for age), height 80 cm (below 3rd percentile of height for age), and was some dehydrated, febrile, having tachycardia and tachypnoea. Abdominal distension with diffuse tenderness and guarding noted without any palpable mass. Bowel sounds were absent. Hemoglobin was 9.1g/dl, total leukocyte counts 9980/mm^3 (neutrophils 69.7%, lymphocytes 16.6%, monocytes 7.3%, eosinophils 6.1%), platelets 4.9 lac/mm^3, MCV 72.3 fm, MCH 21 pg, MCHC 29.1%, blood sugar 74 g/dL, serum glutamic pyruvic transaminase 23 U/L, serum glutamic oxaloacetic transaminase 46 U/L, sodium 141 mmol/L, potassium 3.1 mmol/L, chloride 93 mmol/L. HIV and widal test were non-reactive. Abdominal X-ray showed multiple air-fluid levels. Abdominal CT revealed ileo-ileal intussusception without any distal lead point causing intestinal obstruction with twisting of mesenteric vessels of involved segment around the inner most lumen (Figure 1).

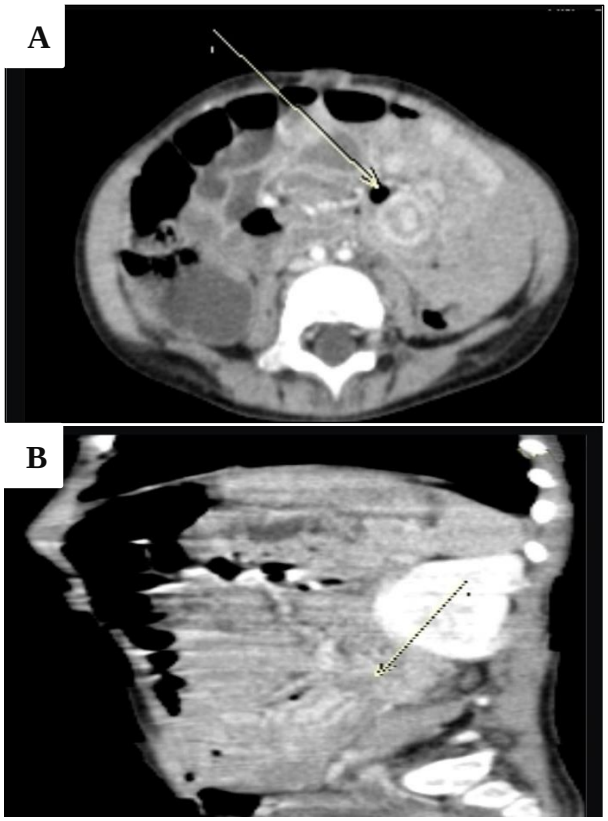

Figure 1: CT scan showing (a): intussusception with target sign in axial view, (b): and sausage sign in sagittal view.

Proximal small bowel was dilated with mild free fluid in peritoneal cavity and pelvis. Laparoscopy done, but didn't find any intussusception or obstruction, probably relieved spontaneously.

Patient was treated symptomatically, hypokalemia corrected, and further evaluated for concomitant microcytic hypochromic anaemia and underweight. Serum thyroid stimulating hormone was 0.63 mIU/L. Serum tissue transglutaminase levels (tTG IgA) was found greatly elevated, 174.4 U (normal range <12 U). CD was confirmed with endoscopic duodenal biopsies, showing histopathology of marsh III. After 6 months of gluten free diet (GFD), she was asymptomatic, abdominal distension resolved, had good appetite and gained 2 kg of weight.

DISCUSSION

Intussusception is the most common cause of intestinal obstruction in late infancy and toddlers.[1] CD is an immune mediated gluten sensitive enteropathy. Both are among the most common disease among children. Biopsy-proven CD had 1% prevalence in children.[2,5] Earlier celiac disease was considered uncommon in India, but studies showed overall sero-prevalence of CD as 1.44% and the overall prevalence of CD as 1.04%. The overall prevalence of CD in the north Indian community was 1 in 96, and 1 in 310 in school children of north India.[6,7]

For the first time, the association between intussusception and CD was demonstrated by Ruoff M et al, in adult in and by Germann R et al, in children in 1997.[3,4,8] Thereafter, multiple case reports stated association between intussusception and CD in adults, and less frequently in children also. Reported prevalence of intussusception with CD is around 1.6-20% and 1.2-25% in adults and children, respectively, but actual prevalence can be higher because of transient and asymptomatic presentation.[5,8-11] Rarely, intussusception can be the initial presenting symptom of pediatric CD.[8,9] Spontaneous reduction of intussusception occurs in 4-10% of children and not uncommon even during preparation for operation.[1]

If not promptly diagnosed and treated, it can lead to arterial obstruction, bowel necrosis and perforation. Ultrasound guided hydrostatic reduction is the currently favored treatment modality in uncomplicated intussusception. Surgical intervention needed in case of prolonged intussusception, shock, peritonitis and perforation or when non-operative intervention is either unsuccessful or lead to perforation, or in situations of lack of a trained professional for non-invasive approaches.[1,12,13]

Gonda TA et al, determined 14 cases of intussusception among a cohort of 880 patients with CD (mean age of 47±17.5 years).[9] Eleven of them underwent imaging for abdominal pain and three were incidental. Intussusception as an initial manifestation of CD was

present in 57% (8/14). Reilly NR et al, found that intussusception was more common with untreated CD and majority of them had no evidence of nutritional deficit at the time of intussusception.[10] Abdominal imaging done in 21% among 254 biopsy-proven CD children, mainly because of abdominal pain.1.2% experienced intussusception <9 months before their diagnosis compared to 0.07% in general pediatric population. Gheibi S et al, found that 5% of pediatric CD presented with intussusception in referral pediatric gastrointestinal ward.[8] The high prevalence was related to small number of patients, genetic or environmental differences, or awareness to similar subject. Intussusceptions were not reported after adherence to GFD. Recurrence of IS may be a sign of non-compliance to GFD.[11]

Despite previously reported intussusception associated with CD, Ludvigsson JF et al, found no such association, but a modest increased risk of intussusception after diagnosis of CD.[14]

In one prospective study, where all 150 serology-positive and biopsy-proven CD children with a median age of 72 months were screened irrespective of symptoms, intussusception was present in 25% with 95% involving small bowel. All but one had asymptomatic intussusception. On GFD, intussusception resolved spontaneously within 7 days in 65%, within 14 days in 84%, and within 28 days in 92% of cases, and none required surgical or hydrostatic reduction. Intussusceptions were more commonly present in children with younger age, presentation with diarrhea and abdominal distension, and who were more severely affected by malabsorption with low weight, rickets, hypoalbuminemia, more severe histopathological changes in form of villous atrophy, and refeeding syndrome.[5]

Intussusception associated with CD involve small bowel, may be multiple or recurrent, and mostly were transient non-obstructive with spontaneous resolution, but may leads to obstruction.[5,8-10,15] The exact mechanism of intussusception in CD is yet to be determined. Most intussusception in CD occur without a lead point. But multiple enlarged mesenteric lymph nodes as reported during surgery, diffuse chronic inflammation with stiffened and thickened bowel, disturbed peristalsis in the hypotonic intestinal loop, dilated bowel, ulceration and weakening of intestinal wall may act as lead point for intussusception.[5,6,8,13]

Intussusception in celiac patients are transient and may be chronic and painless, so can easily be missed in single examination and thus mostly underdiagnosed. In adults, most cases of intussusception are investigated because it occurs due to pathological condition. While in children, mostly they idiopathic and identified by chance when undergoes imaging for abdominal pain, otherwise investigated only when an abnormal clinical presentation or physical examination ensues.[6,8,13] Whether spontaneous resolution in CD is more frequent, will be determined only when routine abdominal ultrasound becomes protocol in CD workup and a large serial study on pediatric populations might help in resolving this question.

CONCLUSION

Transient non-obstructive small bowel intussusception may occur in association with CD, so that celiac testing is recommended in children with intussusception, especially in unusual age range and with concomitant growth failure.

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