Adult Celiac Disease Presenting with Intussusception and Elevated Liver Enzymes

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Key words: celiac disease, intussusception, liver enzymes, dysmotility, gluten-free diet

IMAJ 2003;5:203–204

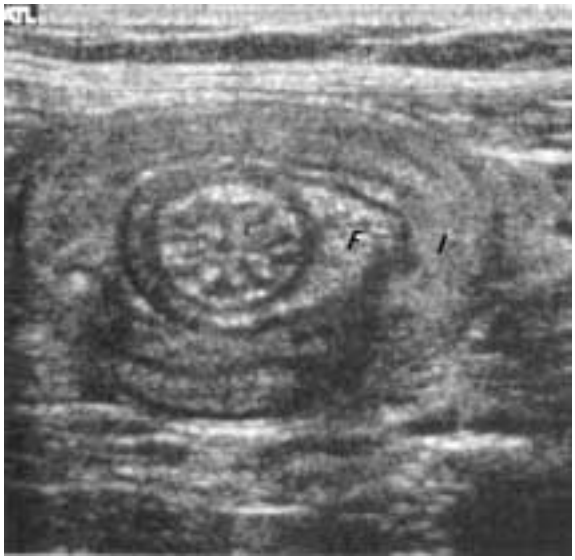
Intussusception in adults is a rare condition documented in less than 1% of patients with bowel obstruction and comprising only 5% of all intussusceptions. In 70–90% of cases, intussusception in adults is associated with an identifiable pathologic lesion or a lead point, 17–24% of which are found to be malignant [1]. The natural history of the 10–25% of cases of idiopathic intussusception has not been well described.

We present a patient who presented with subacute intussusception and abnormal liver enzymes, was subsequently diagnosed with celiac disease, and was managed successfully with conservative treatment.

Patient Description

A 34 year old woman presented with complaints of diarrhea and abdominal cramps of a few weeks duration. Her liver enzymes were known to be elevated for 1 year. She had no history of travel relevant to her condition, no antibiotic use or exposure to contaminated food. There was no familial history of gastrointestinal disease. Her height was 180 cm and her weight 75 kg. The physical examination was normal. Her blood chemistry levels were normal except for mildly elevated liver transaminases: aspartate aminotransferase 49 U/L (normal <35 U/L), alanine aminotransferase 52 U/L (normal <40 U/L); and an INR of 1.37 (normal 0.8–1.2). Investigation of the elevated liver enzymes revealed negative autoimmune and viral serology, as well as blood copper and ceruloplasmin levels. Abdominal sonogram showed a mildly fatty liver.

The patient was started on a lactose-free diet. Three months later, the patient's diarrhea partly ameliorated, but she developed increasing abdominal cramping and lost 13 kg. Physical examination revealed a distended abdomen with hypoactive bowel sounds. A repeated abdominal sonogram showed a mildly fatty liver and strikingly abnormal bowels. Much fluid filled small bowel loops with grossly edematous valvulae conniventes; three ileoileal intussusceptions were seen, along with associated lymphadenopathy (Figure).



Cross-sectional ultrasound image showing the intussusception (i) within the multiconcentric layers of the intussusceptien (I). Mesenteric fat (F) appears as an eliptical echogenicity.

A computed tomography scan demonstrated "target" lesions of small intestines, consistent with intussusceptions, which did not cause complete obstruction. The laboratory tests revealed calcium 8 mg/dl (normal 8.5–10.5), albumin 3.2 mg/dl (normal 3.8–5) and folate 3.1 mmol/L (normal 3.4–47). The hemoglobin level decreased, lymphocytopenia of 1,100/mm^3 (normal 1,500–400) developed and the INR and transaminases increased. At this point there was evidence of malabsorption, weight loss and lymphadenopathy. An anti-endomysial antibody titer was obtained, which was highly positive at 1:160. A duodenal biopsy showed intraepithelial lymphocytes, crypt hyperplasia, and villous blunting – all compatible with celiac disease. The patient was started on a gluten-free diet. Four months later, all her blood tests showed normal values. The diarrhea subsided, along with the abdominal pain and cramping, and the patient gained 10 kg, reaching her normal weight. Follow-up examination of the small intestine showed mild irregularity of the mucosa, suggesting celiac disease, but no evidence of intussusception.

Comment

In recent years a great deal of attention was drawn to the changing spectrum of celiac disease. Today, numerous patients are

diagnosed as having celiac disease because of symptoms other than diarrhea or iron deficiency [2]. In light of this information it is important to consider celiac in patients with atypical symptoms. In the 1960s and 1970s, when small bowel barium meals were routinely used to evaluate patients with celiac disease, intussusception was reported in up to 20% of cases [3]. Outside the radiologic field, however, there is a lack of awareness of this association. Only a few case reports have been published, most of them in the pediatric literature. In adults, in 70–90% of cases, intussusception is secondary to an identifiable lead point such as a polyp, metastatic deposit, or Meckel's diverticulum [1]. Although adult intussusception at presentation can be acute, it is usually subacute or chronic. A longer mean duration of symptoms has been reported in benign lesions than in malignant lesions and in enteric compared to colonic lesions [1]. Gastrointestinal dysmotility and intestinal wall abnormalities such as edematous thickening probably cause the intussusception in untreated patients with celiac disease [4]. In adults with untreated celiac disease, muscular hypotonia and flaccidity of the small bowel may explain the non- obstructive nature of the intussusception. Adult intussusception is usually treated with surgery. Most authors agree that laparotomy is necessary, based on the likelihood of identifying a pathologic lesion. In our patient the intussusception resolved after 3–4 months of a gluten-free diet, which partly reversed the dysmotility and hypotonia of her small intestine. Our patient's liver test abnormalities also resolved after 4 months of this diet. Bardella et al. [5] recently reported that about 40% of celiac patients have hypertransaminasemia at diagnosis. The same group reported a prevalence of 9.3% of occult celiac disease in patients with unexplained hypertransaminasemia, which resolved with a gluten-free diet.

In conclusion, celiac disease should be strongly suspected when intussusception is diagnosed in an adult in the absence of an overt lead point, especially when the presentation is chronic or subacute. When celiac disease is suspected and the signs and symptoms do not indicate a life-threatening situation, conservative management should be undertaken and further investigation carried out before any surgery is planned.

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