CASE REPORT

Coeliac disease in an adult presenting as intussusception without a lead point

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SUMMARY

A case of adult intussusception is presented in which previously undiagnosed coeliac disease was the cause. The diagnosis was made following microscopic examination of a resected segment of small intestine containing multiple intussusception sites. Adult intussusception is rare and in most cases associated with a 'lead point' lesion, often a tumour. As illustrated here, intussusception without a lead point in the adult patient may be the presenting sign of coeliac disease. Recognition of such may allow correct diagnosis and thus prevent unnecessary surgery.

BACKGROUND

Intussusception in adults is rare. The majority of cases are due to a mass or 'lead point' of which approximately half are malignant tumours. A much smaller number occurs without a demonstrable lead point and is ascribed to adhesions, inflammatory or idiopathic causes.^{1 2} Intussusception is recognised as a complication in known coeliac disease, but intussusception as the presenting sign in undiagnosed coeliac disease is little appreciated³ and, as such, may fail to prompt investigation for this disorder. This case illustrates an adult patient who presented with multiple intussusceptions and whose subsequent small bowel resection allowed an initial pathological diagnosis of coeliac disease.

CASE PRESENTATION

A 39-year-old woman was admitted to the hospital with a 1-month history of weight loss, epigastric discomfort, nausea with vomiting of undigested food, night sweats and marked fatigue. Her medical history was unremarkable. Abdominal and pelvic CT

imaging with contrast revealed three jejunojejunal intussusceptions and multiple enlarged mesenteric lymph nodes (**figure 1**). Oesophagogastric-duodenal endoscopy was performed with biopsies of the stomach and duodenum. Shortly thereafter, the abdominal pain increased significantly to the extent that emergency laparotomy was performed (without knowledge of the pathological diagnosis of the biopsy specimens). A 10 cm segment of jejunum with the three intussusception sites was resected. Palpation of the rest of the small intestine revealed no other anomalies. The resected specimen was macroscopically normal, but microscopic examination revealed villous atrophy and inflammatory changes compatible with coeliac disease (**figure 2A**). Of note, the findings of the endoscopic duodenal biopsy showed similar villous atrophy. The postoperative course was uneventful except for prolonged ileus.

Figure 1 Abdominal CT without contrast showing two of the intussusception sites (arrows).

Figure 2 (A) Mucosa from the resected small bowel specimen showing marked villous atrophy and increased chronic inflammation characteristic of coeliac disease. (B) Normal duodenal biopsy three years after instigation of a gluten-free diet.

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INVESTIGATIONS

Shortly following surgery serological studies were performed: anti-deamidated gliadin IgA antibodies were negative (<20 U/mL) as were anti-transglutaminase IgA antibodies (<5 U/mL). Human leucocyte antigen (HLA) typing showed the patient to be HLA DQ2 positive. Repeat abdominal and pelvic scanning 3 months following surgery were normal.

OUTCOME AND FOLLOW-UP

A gluten-free diet was started. One year later the patient was asymptomatic and had gained weight. Three years later an endoscopic duodenal biopsy showed a reversion to normal histology (figure 2B).

DISCUSSION

Intussusception in adults is rare. The small intestine, colon or both may be involved. As many as 90% are due to mass or organic lesions which create 'lead points'.^{1 2} Of these, approximately half are malignant lesions. In the small intestine metastatic tumours (often melanoma) are common causes whereas in the colon, primary adenocarcinoma and lipomas are responsible. In the minority of adult intussusception cases without a lead point an inflammatory or 'idiopathic' cause is often invoked.

Coeliac disease (gluten-sensitive enteropathy) is an autoimmune disease whose varied clinical features, or conversely, subclinical course, are characteristic of an entity with 'protean manifestations'.^{4 5} Of these, intussusception is well described in patients with previously diagnosed coeliac disease. As noted in the diagnostic radiology literature, it is frequently asymptomatic and is often multiple, transient and repetitive.^{6–8} Conversely, small intestinal lymphoma or adenocarcinoma developing secondary to coeliac disease may result in a 'lead point' cause of intussusception.⁸

Much less appreciated is intussusception as the presenting symptom of previously undiagnosed coeliac disease.^{3 9 10} This is illustrated by the paucity of reported cases and absence of mention of this complication in several major clinical reviews.^{4 5} Theories as to its development include 'flaccid, dilated small bowel', fold abnormalities, lymphadenopathy and fluid excess.⁷

Of note, although serological testing for coeliac disease markers was negative, the clinical presentation, IgA negativity, positive HLA DQ2 status, typical histological findings and the reversion of the duodenal mucosa to normal on a gluten-free diet allowed for a confident diagnosis of coeliac disease.

We suggest that all adult patients with intussusception without a lead point lesion undergo investigation for coeliac disease. Furthermore, awareness of this association may prevent unnecessary surgery as intussusception in coeliac disease often resolves spontaneously. The presence of multiple concomitant intussusceptions may provide a clue to making the correct diagnosis.

Learning points

- ▶ Coeliac disease in adults may initially present as intussusception.
- ▶ Intussusception without a 'lead point' in the adult patient should prompt investigation for coeliac disease.
- ▶ Intussusception in coeliac disease is often multiple, transient and repetitive; surgical intervention should be avoided if at all possible.

Contributors AM was involved in writing the manuscript and is responsible for the pathologic discussion, including preparation of photomicrographs. FH and SD provided clinical details and wrote the clinical aspects of the manuscript.

Competing interests None.

Patient consent Obtained.

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