Transient small bowel intussusception in adults: an overlooked feature of coeliac disease

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SUMMARY

Coeliac disease is the commonest immunological gastrointestinal disorder in the Western world. The symptoms of coeliac disease in adults are often non-specific, and a high index of suspicion may be required for timely diagnosis. We describe the case of a 46-year-old woman, with known dilated cardiomyopathy and pulmonary hypertension, who presented with non-specific abdominal symptoms, not initially attributed to gastrointestinal disease. Radiological investigations demonstrated transient small bowel intussusception without other abnormality, leading to the suggestion of coeliac disease as a cause, which was subsequently confirmed as the diagnosis.

BACKGROUND

Although transient intussusception is a reported feature of adult coeliac disease (CD), it is not widely recognised, and appreciation of this association can be critical for the diagnosis of CD and to avoid unnecessary surgical intervention for intussusception. Expedient diagnosis of CD is crucial, as earlier treatment improves symptoms and reduces the risk of complications associated with this condition.

CASE PRESENTATION

A 47-year-old woman with known dilated cardiomyopathy, severe pulmonary hypertension and renal failure was admitted to hospital via the emergency department with symptoms of vomiting. Her medical records included one previous admission with similar symptoms which had resolved spontaneously.

She was admitted under the cardiology team, where her symptoms were attributed to viral gastroenteritis and subsequent decompensation of her cardiac and renal function. She was treated with sildenafil, which significantly improved her cardiac function, but despite this suffered a further episode of abdominal pain and vomiting.

INVESTIGATIONS

A CT scan of the abdomen revealed multiple areas of jejunal intussusception as evidenced by target-like lesions with crescents of intraluminal fat (figure 1), with proximal bowel dilation and minor mesenteric lymphadenopathy. Subsequent barium small bowel enteroclysis undertaken 7 days later showed only transient areas of minimal invagination of the jejunal wall (figure 2) with increased number of jejunal folds (figure 3). The enteroclysis report raised the possibility of CD as a cause for the definite intussusceptions seen on the CT.

Figure 1 Contrast-enhanced CT scan demonstrating a target-like lesion of intussusception (indicated by the white arrow).

DIFFERENTIAL DIAGNOSIS

Small bowel intussusception in adults is usually secondary to a pathological lesion acting as a 'lead point'. Common underlying causes include lymphoma, carcinoid tumour, primary adenocarcinoma or metastatic disease, for example, from melanoma. Non-neoplastic conditions such as lymphoid hyperplasia, infection or coeliac disease have also been reported.

OUTCOME AND FOLLOW-UP

The vomiting and abdominal pain again subsided and she was discharged home.

At cardiology follow-up consultation 1 month subsequently, the patient reported a further episode of vomiting preceded by abdominal pain and

Figure 2 Image from small bowel enema series demonstrates an area of invagination of the bowel wall (white arrow), resulting in transient intussusception.

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Figure 3 Image from small bowel enema demonstrating increased mucosal folds.

bloating, but otherwise was well. Her symptoms of abdominal distension were again attributed to fluid retention, secondary to right heart failure. However, in view of the radiological suggestion of CD, a blood sample was drawn and tested for antiendomysial antibodies and consultation with a gastroenterologist for a period of 1 month was arranged. In the meantime, the patient was advised to undertake a trial gluten-free diet. Routine blood tests at this time also demonstrated an iron deficiency anaemia, and oral iron supplement therapy was started.

When seen at gastroenterology clinic, direct questioning revealed that the patient had, in fact, suffered from numerous episodes of vomiting with associated diarrhoea, colicky abdominal pain and sweating earlier in the year. Since removing gluten from her diet the symptoms had virtually resolved. Her endomysial antibody test was positive and a formal diagnosis of CD was made. Routine screening for the complications of CD showed osteopaenia, for which calcium and vitamin D supplementation was started. At follow-up 1 year later, after remaining on a gluten-free diet, the patient suffers no further symptoms and remains well.

DISCUSSION

Coeliac disease

CD is an autoimmune condition characterised by abnormal small bowel mucosa, reverting to normal when patients are treated with a gluten-free diet, and that relapses when gluten is reintroduced. It is very common in the western world, with a prevalence of around 1:300, and is strongly associated with the histocompatibility antigen HLA-DQ2. The precise structure of the causative antigen remains unknown, but studies suggest that enterotoxicity results from a peptide corresponding to A-gliadin. CD can present at any age, but most adult cases occur in the third and fourth decades. Clinical features are varied and non-specific; diarrhoea, abdominal pain and bloating, weight loss, glossitis and symptoms of anaemia are among the most common symptoms. Other features include neuropathy, amenorrhoea and oligospermia.

Serological testing for antiendomysial antibody is highly sensitive (90%) and specific (99%) for diagnosis, although small bowel biopsy is still regarded as the gold standard test.¹ Radiological tests may also suggest a diagnosis of CD. While barium studies have long been used in the diagnosis and assessment of small bowel mucosal disease, more recently CT features of CD have also been reported.² The most commonly described

radiological features of CD are loss of normal mucosal pattern and lymphadenopathy.³

Treatment is primarily with gluten-free diet, and corticosteroid therapy is required in only a small portion of cases. The complications of CD are more likely in undiagnosed disease or if dietary compliance is poor, and include ulcerative jejunitis, osteomalacia and cavitating lymph node syndrome. Untreated or poorly controlled CD also poses an increased risk of malignancy, including lymphoma, adenocarcinoma and squamous cell carcinoma.³

Intussusception

Intussusception, first described in 1674, is defined as the invagination of a bowel loop with its mesenteric fold (intussusceptum) into the lumen of a contiguous portion of bowel (intussusciens) as a result of peristalsis.⁴

The process of intussusception is most commonly perceived as secondary to mechanical factors, such as a bowel wall mass or kink acting as a 'lead point'. It should also be remembered, however, that functional disturbance may result in intussusception without gross mural abnormality.⁵

Intussusception is most commonly seen in children, in whom it classically presents with a triad of cramping abdominal pain, bloody diarrhoea and palpable abdominal mass. In this age group, around 90% are idiopathic in nature.

In contrast, while intussusception is much rarer in adults (accounting for only 5% of cases), a far higher proportion (up to 90%)⁶ are associated with a detectable underlying cause. The clinical presentation in adults is also different and highly variable; nausea, vomiting, gastrointestinal bleeding, change in bowel habit and abdominal distension all feature. The non-specific nature of these symptoms, in conjunction with the relatively low incidence, makes early diagnosis challenging. One study found that the mean time from onset of symptoms to diagnosis of CD to be 4.9 years.⁷

Intussusception most commonly occurs at the junctions between freely moving and fixed segments of bowel and can be classified according to site as enterenteric, colocolic, ileocolic or ileocaecal. It may also be classified by underlying aetiology (benign, malignant or idiopathic).⁸

CT imaging is most often the examination to suggest a diagnosis of intussusception.⁹ Characteristic features include inhomogeneous 'target' or 'sausage-shaped' soft tissue mass. Intussusception may be a suspected diagnosis confirmed by CT, but more commonly it is an unexpected finding when investigating another clinically suspected condition.

The management of adult intussusception is dependent on the site and underlying cause. Historically, it is reported that most surgeons accept that adult intussusception requires operative intervention to reduce, although debate exists regarding the necessity and extent of bowel resection and manipulation during reduction.⁶ However, increasing usage of CT means that intussusception is increasingly seen as an 'incidental' finding, and operative management is not always appropriate.

Intussusception in CD

The relationship between transient small bowel (enteroenteric) intussusception and adult CD is not new, having first been described in 1978 by Cohen and Lintott,¹⁰ and subsequently with relation to imaging findings. Not only is this association long recorded, it is not uncommon. Case series report that intussusception can be observed on CT in up to 21% of adults with CD.² In one series of patients with intussusception diagnosed on CT, 12.5%⁹ were found to have CD. A further series

of intussusceptions detected at ultrasound revealed that 9% were related to CD.

However, this association has not been well reported to date in the gastroenterological literature, and is not included in even the most comprehensive gastroenterology textbooks.

The mechanism of intussusception in CD is thought to relate to dilated flaccid loops of bowel disturbing normal peristaltic waves, leading to invagination of one segment of bowel into another.¹⁰

Knowledge and recognition of the link between CD and intussusception is important for several reasons. First, as described above, adult intussusception is associated with a high proportion of structural abnormalities and malignancy. As a result, there are many advocates of early surgical intervention for manual reduction of even benign enteric intussusception, with bowel resection if malignancy is suspected.^{8 11}

To date, at least one case of unnecessary laparotomy to treat transient intussusception in a patient subsequently diagnosed with CD has been reported in the literature, which resulted in significant complication and morbidity.¹² Other risks from intervention might also be avoided, for example, one study reports that 13.6% of patients with CD underwent endoscopy without duodenal biopsy before their diagnosis.⁷ We suggest that the appreciation of CD as a cause for enteric intussusception is important to avoid unnecessary operative intervention.

Second, CD itself is a cause of significant morbidity. Transient intussusception may be the only finding to suggest CD. Recognition of this and other features of the condition is important to expedite diagnosis and subsequent treatment. This is especially important where the clinical symptoms are not typical, or there are other disease processes with confounding symptoms, as described in our case.

The significance of timely diagnosis and intervention in CD lies not only in improvement of symptoms, but in allowing identification and treatment of the complications of CD, the risks of which increase without appropriate management. These include osteomalacia, cavitating lymph node syndrome, demyelinating neuropathy and malignancy such as lymphoma and adenocarcinoma.¹³

The growing use of CT for abdominal imaging means that transient intussusception without apparent structural disease will be more frequently encountered.¹³ Appropriate clinical interpretation of such findings cannot be undertaken without sound understanding of potential causes. We have found one previous case in the literature where a radiologist prospectively raised the possibility of clinically unsuspected CD, in a patient with intussusception on a CT, and where the diagnosis was subsequently confirmed.¹⁴ Ours is the second case of this type.

This case highlights the potential significance in appreciation of the link between transient intussusception and CD. It can be the key to timely diagnosis and expedient treatment, and, in some cases, will avoid unnecessary surgery.

Learning points

- ▶ Small bowel intussusception in adults is usually, but not always, due to an underlying lesion acting as a lead point. This often requires surgical treatment.
- ▶ However, non-surgical causes of small bowel intussusception should be considered, including coeliac disease.
- ▶ Knowledge of the above can expedite diagnosis and prevent unnecessary surgery.

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Competing interests

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

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