Case Report

Intussusception in children with celiac disease

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Abstract

Celiac disease (CD) is a chronic illness. Blood testing for tissue transglutaminase antibodies is the initial screening test for the diagnosis of CD, and upper gastrointestinal endoscopy and duodenal/jejunal biopsy are used to confirm CD. Intussusception (IS) is the process in which a proximal segment of the bowel invaginates through the lumen of a distal segment. The association between pediatric IS and CD has been described but is still not widely recognized. Herein, we report a case of IS as the first manifestation of CD in a child. A 3-year-old girl presented to the emergency department with a 1-week history of marked abdominal distention and lethargy, but there was no history of fever, bleeding per rectum, or jaundice. A second-degree relative had a family history of CD. Clinical examination: The patient was a lethargic child with pale conjunctiva and bilateral lower limb edema. She was a febrile and had a normal hemodynamic status. The adipose tissue was diminished throughout the patient's body; her weight was 8 kg (<5% weight percentile for girls), and her height was 81 cm (<5% height percentile for girls). Laboratory results included the following: Hb of 9 g/dL, serum ferritin of 10 ng/mL (30–400 ng/mL), normal liver function test results except for hypoalbuminemia at 21 g/L (35–52 g/L), and low blood cholesterol of 0.94 g/L (1.54–2.01 g/L). The patient's blood sugar level was 98 mg/dL, and her renal function test results were normal, with negative septic screening. Abdominal radiography revealed several air–fluid levels, suggestive of an obstruction in the small bowel. Abdominal ultrasonography revealed typical features confirming the diagnosis of IS. Abdominal computed tomography demonstrated an enteroenteric IS with no other signs of organic causes, such as lymphoma or other tumors. Based on the high index of suspicion of CD, a workup confirmed the diagnosis. A gluten-free diet [A1] was started during the hospital course, and the patient improved dramatically regarding her symptoms and was discharged home. In conclusion, this case highlights the association between IS and CD. Intussusception is an emergency condition and usually idiopathic. However, in atypical or recurrent typical presentations underlying causes, we should do proper investigations to initiate appropriate management. [A1]Abbreviations are generally avoided for terms that are not repeated in the text. In this case, this abbreviation is not used elsewhere in the Abstract. Hence, I have deleted the abbreviation. Categories: Pediatrics, Gastroenterology.

Keywords: Celiac disease, child nutrition, gluten-free diet, intussusception, recurrent small-bowel obstruction

Introduction

Celiac disease (CD) is a chronic immune-mediated disorder triggered by gluten ingestion in genetically predisposed individuals with the human leukocyte antigen (HLA)-DQ2 or HLA-DQ8 genotype on chromosome 6.[1] Serum tissue transglutaminase antibody testing is used as a screening test for CD diagnosis, and upper gastrointestinal (GI) endoscopy and duodenal/jejunal biopsy are used to confirm the diagnosis. Classical (GI) celiac manifestations include diarrhea, abdominal pain, distention, and weight loss. However, CD can present in other forms, such as nonclassic (non-GI), latent, silent, potential, or refractory CD.[2] Patients with nonclassical CD are usually diagnosed late. The prevalence of CD in most regions is <1%, whereas its prevalence in Saudi Arabia has been reported to be 1.5%[3] and 10%–26% in children in northern India.[4]

Patients with CD may develop intussusception (IS), although this condition is mostly asymptomatic. IS is a pathological in which

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one segment of the small bowel invaginates through the lumen of a nearby segment of the intestine.

Idiopathic IS is a common disease in young children and usually treated by nonsurgical reduction like using pneumatic and/or hydrostatic enemas[4-6]; however, the etiology of IS remains idiopathic and is still under investigations. Moreover, the recurrence of small-bowel IS is associated with specific pathologies.[5,6] Asymptomatic and transient, asymptomatic IS without a leading point in patients with CD has been reported in radiological studies.[7-10] However, rare cases of chronic or recurrent IS require further investigation; furthermore, chronic/recurrent IS has rarely been associated with CD. Whether all IS patients should be screened for CD remains controversial; in pediatric patients, the association between pediatric IS and CD has been described but remains unresolved. Therefore, here, we report the case of a patient with pediatric IS who was diagnosed using radiology; the diagnosis of CD was subsequently confirmed via duodenal/jejunal biopsy. The present case demonstrates an association between these two conditions. We believe such a case presenting an important lesson for the general physician and general pediatrician to consider the possibility of IS in patients with CD.

Case Presentation

An undiagnosed 3-year-old Yemeni girl presented with complaints of chronic diarrhea, abdominal distension, and failure to gain weight over 8 months. She had a history of abdominal pain that had started 6 months prior to arrival at our hospital and a 1-week history of abdominal distention and lethargy with no fever, night sweats, passage of fresh blood per rectum, blackish tarry stool, or yellowish discoloration of the sclera. A second-degree relative had a family history of CD.

On clinical examination, the patient was lethargic, with pale conjunctiva and bilateral lower limb edema. She was afebrile and had a stable hemodynamic status. The patient had reduced adipose tissue, she weighed 8 kg, and her height was 81 cm (both parameters, <5% percentile for 3-year-old girls). Her abdomen was distended with diminished bowel sounds and no masses, hepatomegaly, or splenomegaly.

The laboratory results revealed hemoglobin, 9 g/dL, serum ferritin (10 ng/mL; normal range, 30–400 ng/mL), normal liver function test results except for hypoalbuminemia (21 g/L; normal range, 35–52 g/L), and low blood cholesterol (0.94 g/L; normal range, 1.54–2.01 g/L). Her blood sugar level was 98 mg/dL, and her renal function tests were normal, with negative septic screening. Abdominal radiography revealed multiple air–fluid levels, suggestive of an obstruction in the small bowel [Figure 1].

Abdominal ultrasonography [Figure 2] revealed features suggestive of IS; the presence of enteroenteric IS was confirmed using abdominal computed tomography (CT), with no other signs of organic causes, such as lymphoma or other tumors.

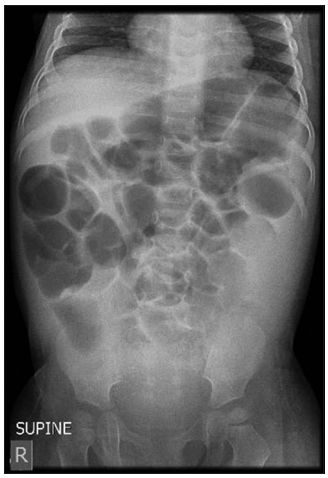

Figure 1: Abdominal X-ray showing a dilated small bowel with the absence of air in the large bowel, suggestive of intestinal obstruction

During admission to our CD center, a diagnosis of CD was highly suspected; hence, tests for specific serum autoantibodies were requested [immunoglobulin A (IgA) tissue transglutaminase >560 UI/mL and endomysium antibodies >350 UI/mL, with normal serum IgA levels]. Upper GI endoscopy revealed scalloping of the duodenal folds. Duodenal biopsy revealed total villous atrophy, crypt hyperplasia, and mucosal inflammation (Marsh 3c), confirming the diagnosis of CD [Figure 3].

The patient was treated conservatively with gradual resolution of the obstruction, and a gluten-free diet (GFD) was started. The patient was discharged after complete resolution of her symptoms and remained asymptomatic during the follow-up period of 12 months, with reasonable weight gain.

Discussion

The CD Center at King Abdulaziz University Hospital (KAU) was launched in February 2017. All patients with CD are followed up there, with more than 500 cases reported through September 2023. The prevalence of CD is high worldwide, at approximately 1.5% in Saudi Arabia.[3] There are case reports of transient nonobstructive IS in adults with CD (10). However, IS as an initial manifestation of CD is rarely reported.[11] Ruoff *et al*.[12] first demonstrated the association between CD and IS in 1968. IS is a common emergency in pediatrics and is usually idiopathic.[4-6] The classical clinical presentation includes abdominal pain, abdominal mass, and red currant jelly stools in children with a normal nutrition and body weight.[13-15] IS requires surgical or nonsurgical intervention. Surgical interventions indicated IS with complications like peritonitis, perforation, or shock. Other noninvasive modalities, like pneumatic reduction or enema, can be used in most of the cases with a high success rate. The link between IS and CD has been reported in some studies.[7-9] If the presenting symptoms are unusual, or if there is a chronic diarrhea without weight gain, abdominal distention, recurrent abdominal pain, recurrent IS, or worsening of clinical

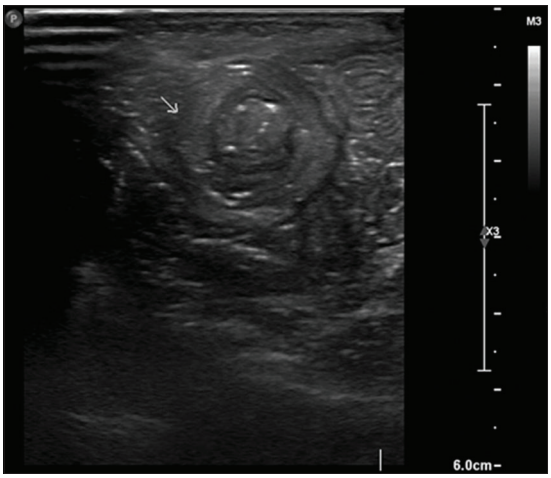

Figure 2: Abdominal ultrasound showing a round mass with a target bowel-in-bowel appearance in the transverse plane (doughnut sign – indicated by the arrow)

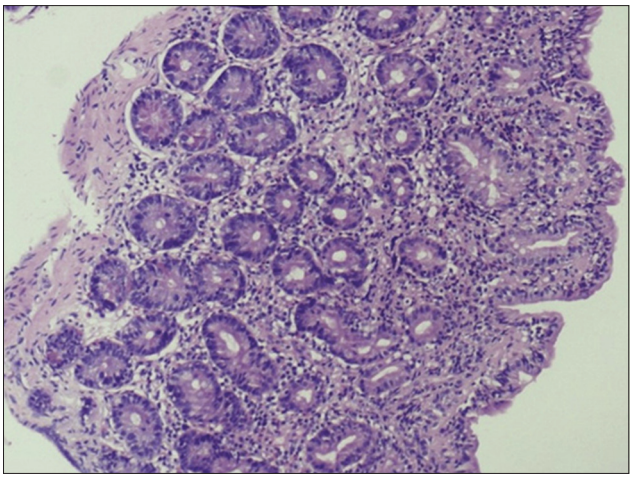

Figure 3: Duodenal biopsy (H and E stain) showing total villous atrophy, crypt hyperplasia, and mucosal inflammation with marked lymphocytic infiltration (Marsh 3c)

condition, further tests should be done after the treatment of the IS, like the management of the patient in this case. In this report, CD was the primary pathology, with atypical clinical presentation of recurrent IS. The link between recurrent IS and CD was clear, and the symptoms resolved after the child commenced on a GFD. Another peculiarity is that the IS in CD is usually temporary. The success rate is high for nonsurgical modalities like reduction, and the land complication rate is low. However, surgical intervention is indicated when the patient develops complications like perforation or abdominal mass of noninvasive interventions are unsuccessful.[15]

Intussusception is underdiagnosed in pediatric patients with CD because it may be chronic and painless and it is important for initial necessary investigation for early diagnosis of IS where there is a high clinical suspicion. Since CD is a disease associated with chronic inflammation, the suggestive mechanism of IS in CD is related inflammation, which causes increased thickening of the intestinal wall; this will lead to hyperperistalsis and dilation of the small bowel. This might be the leading point for the link between CD and IS. The early diagnosis of IS in patients with CD is important, so immediate management can be started to preserve the bowel. Long-term management is important to prevent recurrence and requires a multidisciplinary team including a gastroenterologist, radiologist, surgeon, and dietitian. However, most of the available data regarding this association only come from small studies in the form of case reports or case series case reports. Further studies in bigger scales are need to clarify the link between IS and CD.

Conclusions

Idiopathic acute small-bowel IS is associated with CD. Therefore, we suggest that celiac serology combined with upper GI endoscopy be performed in older children with IS in cases where a secondary cause should be suspected. IS in CD is usually transient and should be managed conservatively rather than with early surgery; additionally, IS should be suspected in patients with known CD presenting with abdominal pain or abdominal distention.

Abbreviations

CDC = Celiac disease
GFD = Gluten-free diet
KAU = King Abdulaziz University
GI = Gastrointestinal
IS = Intussusception.

Ethical approval and informed consent

A phone consent was obtained from the patient's legal guardian and the form was signed by the most responsible physician. Availability of data and materials sharing is not applicable to this article, as no datasets were generated or analyzed during this case report.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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