A Rare Symptom of Celiac Disease: Invagination

Nafiye Urgancı, Sinem Polat, Banu Yılmaz

Clinic of Pediatrics, Şişli Hamidiye Etfal Training and Research Hospital, İstanbul, Turkey

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ABSTRACT

Association between celiac disease and invagination is common in adults, but there are few cases have been reported in pediatric patients as well. In this case we would like to discuss the correlation between celiac disease and invagination.

Keywords: Celiac disease, invagination, childhood

ORCID IDs of the authors: N.U. 0000-0003-4854-507X; S.P. 0000-0001-7107-5489; B.Y. 0000-0002-3540-4772

INTRODUCTION

One of the most common causes of gastrointestinal obstruction in children is invagination. In children between 3 months and 5 years old, small intestinal obstruction is observed with a frequency of 22–56/100,000. Nausea, vomiting, and abdominal pain are the clinical findings of invagination, and it usually recovers spontaneously (1, 2).

However, in patients who have celiac disease and do not receive any treatment, it has been reported in recent years that invagination is seen as a rare clinical finding of the disease (3). While the association of celiac disease with invagination was first described in 1968 in an adult patient with celiac disease (4), it was reported in a small number of children (5–10).

A patient with celiac disease who presented with abdominal pain, vomiting, and invagination was examined in this case report.

CASE REPORT

A four-and-a-half-year-old girl was admitted to our pediatric emergency polyclinic with complaints of severe abdominal pain and vomiting. No postnatal features were found in the baby who was born of parents with first-degree cousin marriage, in term, 3100 g, and in a spontaneous natural birth. Her medical history revealed she had constipation and diarrhea attacks in addition to intermittent abdominal pain that continued for 1 year. She took the antibiotics and antiparasitosis prescribed by the doctors they repeatedly consulted. However, the frequency of abdominal pain that started in the last few days and regressed spontaneously gradually increased, and she lost weight along with vomiting.

On physical examination, the general condition of the patient with a body weight of 12 kg and height of 96 cm was moderate. She had a cachectic appearance, and her eyeballs were slightly depressed. The respiratory system examination was natural; compression, airway, and breathing was 130/min rhythmic; and a systolic murmur with 1/6 severity was detected in the cardiovascular system. Organomegaly could not be palpated because her abdomen was highly distended. Laboratory tests showed hemoglobin 10 g/dL, hematocrit 31%, leukocyte count 9380/mm^3, platelet count 544,000/mm^3, aspartate aminotransferase 56 U/L, alanine aminotransferase 49 U/L, gamma-glutamyl transpeptidase 20 U/L, alkaline phosphatase 279 U/L, total protein 5.4 g/dL, albumin 2.9 g/dL, urea 28 mg/dL, creatinine 0.17 mg/dL, iron 30 ng/dL, iron-binding capacity 406 ng/dL, ferritin 10 ng/dL, vitamin B12 189 pg/mL, folic acid 7 ng/mL, IgA 208 mg/mg/dL, and IgM 49 mg/dL. Microcytic anemia was detected in the peripheral spread. On ultrasonography of the abdomen (Siemens, Germany), the abdomen was highly distended, and a concentric, lamellar echogenic structure of invaginated intestinal segments and a 40 mm target lesion compatible with invagination were observed in intensive gas appearance and thickened hypoechoic intestinal wall (Figure 1). The patient was diagnosed with invagination by pediatric surgery. On follow-up of the patient, it was observed that the appearance of invagination spontaneously recovered. In the meantime, anti-endomysium antibody (EMA), one of the celiac antibodies examined for growth retardation, EMA IgA, and EMA IgG were found to be positive. Endoscopic examination of the upper gastrointestinal tract system revealed no pathology other than a comb-like appearance in the duodenum. Histopathological examination of biopsy specimens revealed total villous atrophy, increased intraepithelial lymphocyte count, and crypt hyperplasia (Figures 2, 3). The patient was diagnosed with celiac disease, and a celiac diet was started. The patient had been followed up for 2 years, and EMA IgA and EMA IgG were negative, her height and weight were 10–25 p, and no findings of invagination were further seen in our clinic. Verbal consent was obtained from the family.

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Address for Correspondence: Nafiye Urgancı, E-mail: nafiyeurganci@yahoo.com

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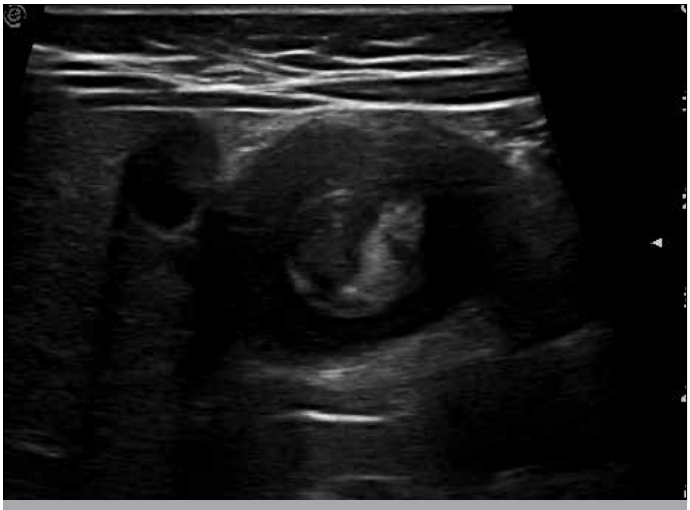

Figure 1. Typical sonographic image of invagination. Typical "target image" in transverse sections with convex and linear probe

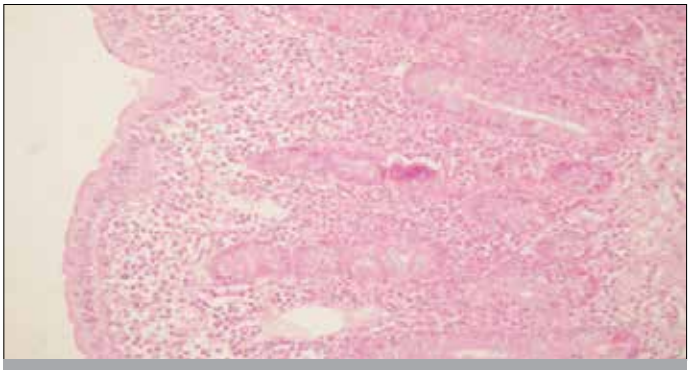

Figure 2. Flattened villi, crypt hyperplasia, and intraepithelial lymphocyte increase (H&E, ×200 magnification)

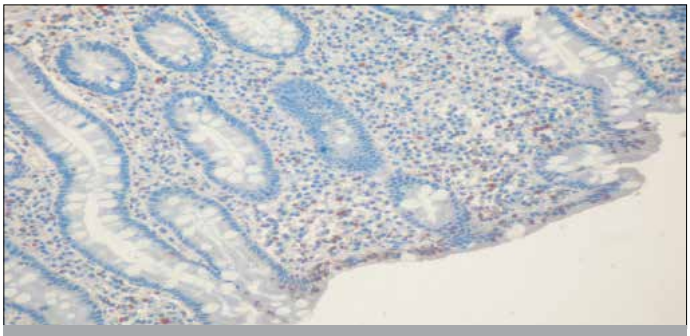

Figure 3. CD3 immunoreactivities in increased intraepithelial lymphocytes (H&E, ×200 magnification)

DISCUSSION

Invagination is an important cause of rectal bleeding and bowel obstruction in infants and children. It is an acute abdomen clinical condition that is most commonly seen in infants between 3 months and 2 years old. Its specific etiological factor is not known in classical invagination. Hypertrophy of Peyer's plaques, mesenteric lymphadenopathy, and rotavirus gastroenteritis are most commonly considered as responsible in cases referred to as idiopathic invagination (2, 11, 12). Celiac disease is a genetically transmitted inflammatory disease caused by gluten intolerance. In addition to classical findings, it is also accompanied by extraintestinal findings, such as iron deficiency anemia that does not respond to treatment, puberty delay, osteoporosis, cryptogenic hypertransaminasemia, and peripheral neuropathy, as well as autoimmune diseases, such as autoimmune thyroiditis, type 1 diabetes mellitus, and Sjögren's syndrome. In recent years, invagination has been one of the rare atypical gastrointestinal findings accompanying celiac disease in patients with untreated celiac disease (5, 6, 13). Celiac disease causes impaired intestinal motor function. As a result, it is believed that impaired intestinal peristaltism in hypotonic intestinal loops may lead to invagination (5). The incidence of invagination in patients with celiac disease is reported to be 1%–2% higher than that in normal children (3, 10). Only 1 (0.4%) of our 236 patients who we followed up with a diagnosis of celiac disease consulted to our clinic with invagination. We can explain this rate, which is lower than that reported in the literature, by the fact that the association of celiac disease and invagination is ignored by both pediatric surgeons and pediatricians because of the spontaneous recovery in 75% of invagination. Although the association of celiac disease and invagination is reported in a 9-month-old girl, as the youngest patient in the literature, it can be seen at any age as in our patient (5–7, 9, 10). Ultrasonography is very important in the diagnosis of invagination. It is the most commonly used method for the confirmation of the diagnosis, and it has a high correlation with the clinical findings. Hydrostatic reduction with barium enema and reduction with air insufflation are frequently used non-operative treatment methods. Success rates are reported to be quite high in many studies (12). Since growth retardation and anemia were also detected in our patient who was diagnosed with invagination through our clinic and ultrasonography, the antibodies related to celiac disease were investigated, and the patient was followed up. The patient who recovered spontaneously without any requirement for reduction or surgery was diagnosed with celiac disease serologically and histopathologically, and a gluten-free diet was initiated.

CONCLUSION

While patients with clinical findings, such as growth retardation, diarrhea, malabsorption, and abdominal distention, are diagnosed with typical celiac disease, in recent years, it is possible to diagnose cases with "atypical" and "silent" clinical findings through advanced serologic methods. Therefore, celiac disease should be kept in mind when growth retardation and anemia are detected in patients with invagination, which is a rare clinical finding of atypical celiac disease, and the celiac serology should be thoroughly investigated.

Informed Consent: Verbal informed consent was obtained from patients' parents who participated in this case.

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