

Unusual Initial Presentation of Celiac Disease in Children: Three Case Reports

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Abstract

Background: The togetherness of invagination with celiac disease is an extremely rare condition especially in children. However, invagination may be the presenting symptom of celiac disease. Moreover, recurrent invaginations have been also reported in patients with celiac disease. **Aim:** To increase the awareness of clinicians about togetherness of these conditions. **Case Presentation:** Herein, we will discuss three children with diagnosis of celiac disease who presented with invagination and intestinal pseudo-obstruction. **Conclusion:** Children with pseudo-obstruction and invagination without an underlying etiology should be evaluated for the presence of celiac disease, especially if they have accompanying growth retardation or anemia and if they are at an unusual age for invagination. Further studies are warranted to elucidate the exact relationship of invagination with celiac disease.

Keywords

Celiac Disease, Invagination, Intestinal Pseudo-Obstruction, Children

1. Introduction

Invagination is one of the most common causes of gastrointestinal obstruction in children [1]. Though it is usually idiopathic, tumors, Meckel diverticula, viral infections [2], cystic fibrosis, Crohn disease and celiac disease (CD) may rarely cause invagination [1].

Celiac disease is an autoimmune enteropathy affecting genetically predisposed patients [3]. The typical symptoms of CD include growth retardation, abdominal distention and anemia. However, patients may present with atypical symptoms such as neurological symptoms, or invagination [3] [4]. The togetherness of invagination

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with CD is a known but very rare condition especially in children. Invagination may be the presenting symptom of CD and moreover, CD patients with recurrent invaginations have been also reported [4] [5].

Herein, we will discuss three children with diagnosis of celiac disease who presented with invagination and intestinal pseudo-obstruction.

By this way we aimed to increase the awareness of clinicians about togetherness of these conditions.

2. Case Reports

2.1. Case 1

A nine-year-old boy was admitted to the hospital with abdominal pain and bilious vomiting for three days. His vital signs were normal. His weight was 20 kg (<5th percentile) and height was 124 cm (10th percentile). In physical examination, abdominal distention and diffuse tenderness on palpation were detected. Moreover, a mass lesion on the left lower quadrant was palpated. Laboratory test results were as follows: hemoglobin (Hgb), 9 g/dl (14 - 18); white blood count (WBC) , $22 \times 10^3/\mu\text{L}$ (4.5 - 11), C-reactive protein (CRP), 4.8 mg/dL (0 - 5), and Na 128 mmol/L (134 - 145). There was free air under the diaphragm in standing direct abdominal graph (SDAG) together with multiple air-fluid levels (Figure 1). Abdominal computed tomography (CT) revealed an

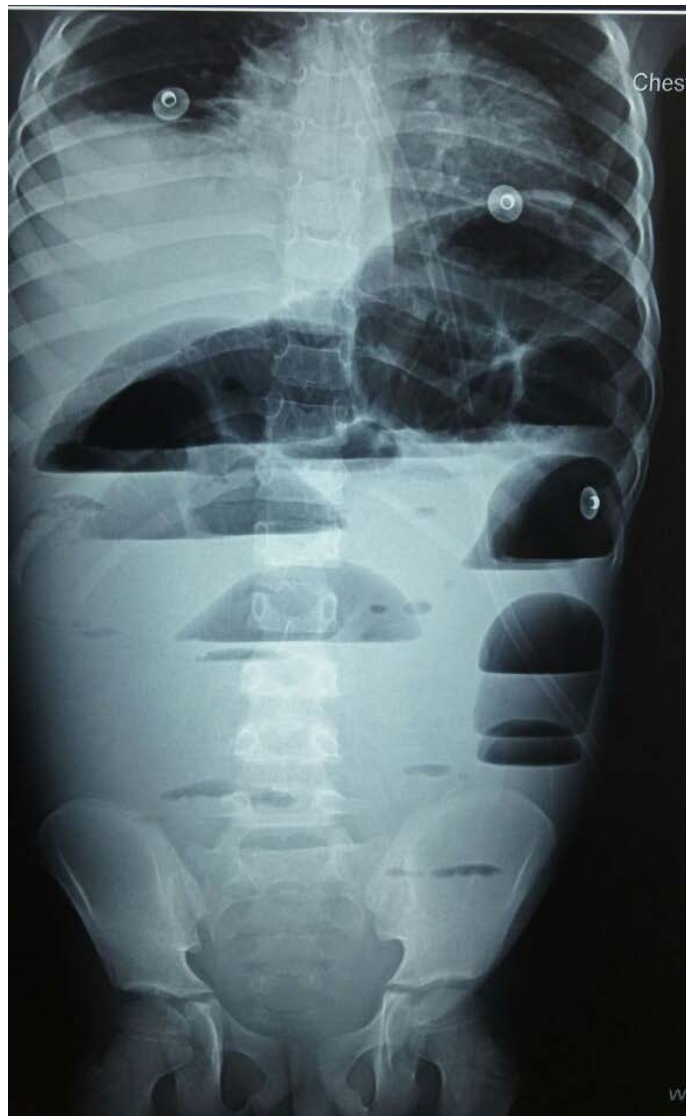


Figure 1. Free air under the diaphragm and multiple air-fluid levels in standing direct abdominal graph.

ileo-ileal invagination of 5.8 cm in length in the left lower quadrant, minimal free fluid between intestinal loops, an extensive dilatation between colonic segments (**Figure 2**) and fluid accumulation in Douglas pouch. Due to the presence of signs of perforation, laparotomy was performed. Since intestinal necrosis was found in ileo-ileal invaginated segment, about 40 cm proximal to the cecum (**Figure 3**) partial ileal resection compromising perforation and primary end to end anastomosis were performed. Laparotomy, did not reveal any other intestinal pathology or enlarged lymph nodes. After operation appropriate fluid and electrolyte replacement was ordered. However, on postoperative 8th day, abdominal distention was still present together with air-fluid levels in SDAG and the patient was consulted with pediatric gastroenterology department. Pediatric gastroenterologists obtained multiple biopsies from duodenum. The histopathology of biopsy specimens were reported as focal villus atrophy, intraepithelial lymphocytosis and hyperplasia and mitosis in crypts (**Figure 4**). His serum anti-tissue transglutaminase antibody (tTG-Ab) immunoglobulin (Ig) G level was 172 U/mL (normal range < 20) and antigliadin IgA level was 136 mg/dL (AGA) (normal range < 20). (IMMCO Diagnostics, Inc., Buffalo, NY). With approximately 1 year of gluten free diet (GFD), the patient gained 8 kg, and histopathology of terminal ileum biopsy was unremarkable. With in 24 months of follow-up after operation, signs of intestinal obstruction or abdominal fluid accumulation were not present on abdominal CT.



Figure 2. Abdominal computed tomography: an ileo-ileal invagination and an extensive dilatation between colonic segments.



Figure 3. Intestinal necrosis present in ileo-ileal invaginated segment.

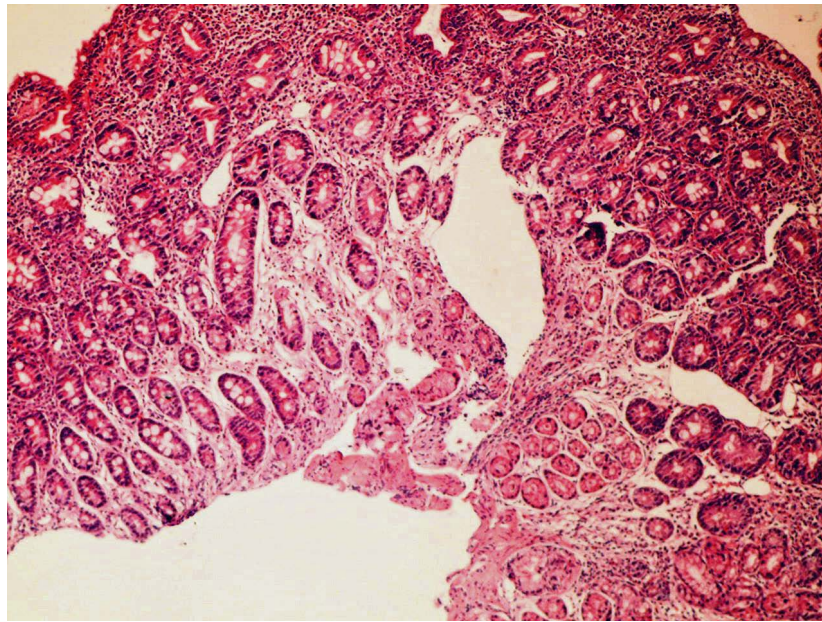


Figure 4. Total villous atrophy, intraepithelial lymphocytosis, crypt hyperplasia (marsh stage 3c), HE, X40.

2.2. Case 2

A six-year-old girl was admitted to the hospital with colic type abdominal pain and recurrent diarrhea. These self-limited episodes were lasting about one week. She was cachectic in appearance with a history of anorexia. Her weight was 17 kg (10th-25th percentile) and her height was 110 cm (25 percentile). Her abdomen was distended without any tenderness on palpation. Laboratory test results were as follows: Hemoglobin, 10.2 g/dl (14 - 18); WBC, 13.5×10^3 /uL (4.5 - 11), CRP, 10 (0 - 5) mg/dL, and Na 132 mmol/L (135 - 145). There were multiple fluid-air levels in SDAG. In abdominal ultrasound, distention in intestinal loops, and a reductable, transient ileo-ileal donut-shaped invagination with a 4 cm diameter was present in right upper quadrant. Control sonograms obtained after reduction of invagination were unremarkable. Her serum tTG IgG, and tTG IgA levels were 87 U/ml, and 100 U/ml., respectively. Endoscopic duodenal biopsy results revealed celiac disease with Marsh III histopathological appearance-intraepithelial lymphocytosis. With GFD, the asymptomatic patient gained 7 kg of weight within 1 year.

2.3. Case 3

A ten-year-old boy was admitted to the hospital with abdominal pain, constipation, vomiting and distention for 6 months. It was learnt that he had ileus attacks, and the last attack was 2 months ago which did not require surgical interventions. In physical examination, abdomen was distended with diffuse tenderness on palpation. In auscultation, the abdominal sounds were hyperactive. His Hgb level was 9.8 g/dl and CRP, erythrocyte sedimentation rate and electrolyte levels were within normal limits. His serum tTG IgG, and tTG IgA levels were 56 U/ml, and 68 U/ml, respectively. There was an air fluid level in abdominal X-ray as a sign of intestinal obstruction (**Figure 5**). In abdominal ultrasound signs of invagination were not present but intestinal loops were dilated. Endoscopic duodenal biopsy results confirmed the diagnosis of CD (**Figure 6**). Nasogastric tube was inserted and fluid and antibiotic treatments were ordered. There was a bilious drainage from nasogastric tube. On the 3rd day of his hospitalization, his clinical picture improved and with an insertion of rectal tube, his symptoms reduced. After decompression, SDAG and contrast enhanced colon graphs were normal. With three months of GFD, he gained two kg.

3. Discussion

We have presented three children admitted to the hospital with the invagination symptoms and concomitant diagnosis of CD. Invagination is the prolapse of a part of intestinal loops into the lumen of the adjacent segment.

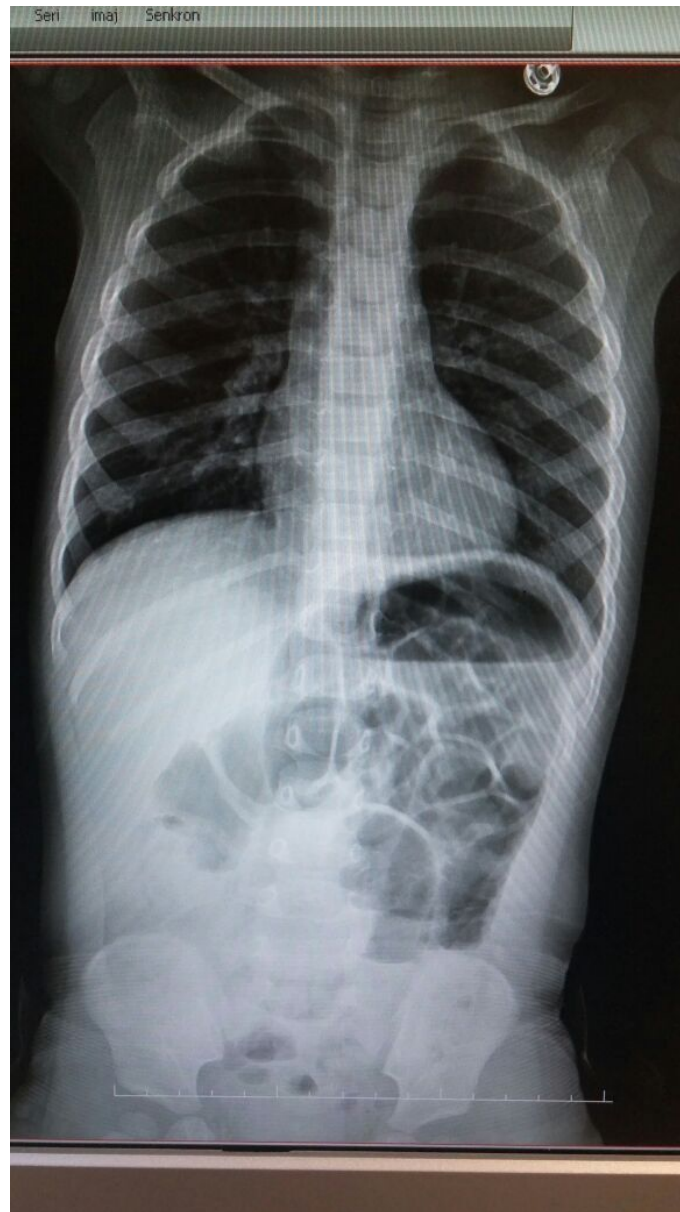


Figure 5. An air fluid level in abdominal X-ray as a sign of intestinal obstruction.

It is generally an idiopathic condition reported more commonly in children [1]. About 90% of the cases with invagination are between six months and two years of age. However, all three cases of us were older than six years of age. Its symptoms include pain, nausea-vomiting and rectal bleeding. Conservative treatment under ultrasound is the preferred method in transient small intestinal invaginations, however, in patients with persistent invagination, atypical radiological findings or deteriorating clinical situation may require prompt surgical exploration [1]. Our 2nd and 3rd cases were followed with conservative treatment, but the first one was operated due to the presence of the signs of perforation.

Gluten sensitive enteropathy, CD, is an autoimmune reaction to the gliadin fraction of gluten present in cereals as barley, rye and wheat [6]. The disease presents with villous atrophy and crypt hyperplasia after ingestion of gluten [3]. It is reported in 1% - 2% of general population [7]. The typical symptoms of CD include chronic diarrhea, anemia and growth retardation. In all 3 cases reported here, growth retardation and anemia were present.

On the other hand, CD patients may present with atypical symptoms such as infertility, abdominal distention

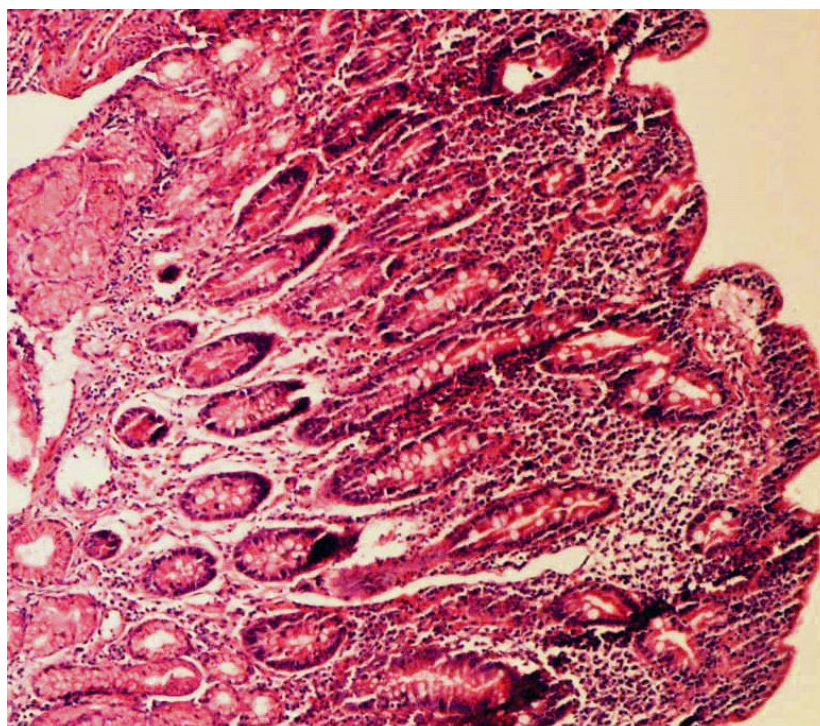


Figure 6. Total villous atrophy, intraepithelial lymphocytosis, crypt hyperplasia, HE, X100.

or constipation. The prevalence of acute abdominal pain in CD patients is 3% and chronic or recurrent abdominal pain may also be the presenting symptom of CD [8]. Although the exact cause of abdominal pain in CD is unknown, autonomic dysfunction in intestinal structures, hypotonic small intestine loops, and deteriorations in neural control of intestinal movements, increase in the thickness of small intestine wall, diffuse inflammation, dilatations in intestinal loops, and excessive peristalsism reported in CD patients may predispose to the development of invagination. Delayed small intestinal transit time and decrease in motility are also accused. Especially in our first case, persistence of abdominal distention made us search for the presence of CD.

The invagination prevalence among CD patients was reported as 1.2 - 18 percent. Hizal *et al.* reported that transient invagination may accompany 20% of CD patients and Reilly *et al.* reported that in children with CD risk of invagination increased 10 - 20 times [4] [5]. Similarly, Cornelius *et al.* suggested checking for CD in patients with invagination though they were asymptomatic [9]. Moreover, Gonda *et al.* reported that invagination and abdominal pain may be the presenting symptom in as high as 57% of the patients with CD and additionally in patients with recurrent invagination on Gluten Free Diet (GFD) any recurrence was not determined [10]. Similarly, recurrence was not observed in our cases after GFD.

In conclusion, children with pseudo-obstruction and invagination without an underlying etiology should be evaluated for the presence of CD especially if they have accompanying growth retardation or anemia and if they are at an unusual age for invagination. Further studies are warranted to elucidate the exact relationship of invagination with CD.

Written informed consent was obtained from the patients's legal guardians for publication of this case report and any accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal.

Competing Interests Section

The authors declare that they have no competing interests

Author's Contribution Section

Sevgi Buyukbese Sarsu designed the study and acquired the data; drafted the article and revised it critically for

important intellectual content. Mustafa Demirci made radiologic examination.

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