Unusual Presentation of Celiac Disease in a Toddler from a Pediatric Surgery Unit

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Abstract Celiac disease is an intestinal autoimmune disease induced by the ingestion of gluten proteins (wheat, rye, barley) in genetically susceptible individuals. Celiac disease can present with gastrointestinal or extraintestinal manifestations (malabsorption, diarrhea, constipation, liver, skin or bones disorders). We present the case of a 14-months-old girl admitted for altered general condition and symptoms of intestinal occlusion (severe abdominal pain, absence of feces, abdominal distension), with associated severe malnutrition. History revealed chronic diarrhea and a lactose intolerance diagnosed 3 months prior to admission. The biochemical and imaging investigations excluded any acute surgical pathology. The serological tests showed high values of anti-tissue transglutaminase antibodies. The intestinal biopsy confirmed celiac disease. In children with suspicion of intestinal obstruction associated with a history of diarrhea or constipation and failure to thrive, testing for celiac disease should be considered.

Keywords: celiac disease, children, anti-tissue transglutaminase antibodies, Pediatric Surgery Unit


1. Introduction

Celiac disease (CD) is an intestinal autoimmune disease induced by the ingestion of gluten proteins (wheat, rye, barley) in genetically susceptible individuals [1]. CD may present with a large variety of symptoms, including typical gastrointestinal symptoms (diarrhea, steatorrhea, weight loss, bloating, flatulence, abdominal pain) and also non-gastrointestinal abnormalities (abnormal liver function tests, iron deficiency anemia, bone disease, skin disorders) [2,3]. Due to the wide range of symptomatology, an accurate diagnosis is sometimes difficult [4].

The diagnose of CD is based on serological antibodies upon gluten diet, such as anti-tissue transglutaminase antibodies, anti-gliadin peptide antibodies or anti-endomysium antibodies, together with the endoscopic biopsy of the small bowel [5]. The classic form presents clinical symptoms of malabsorption, positive serology and a histological aspect with crypt hyperplasia and villous atrophy [6].

Treatment of CD consist in a strict life-long gluten-free diet, which usually offers a protective effect against complications of malabsorption and extraintestinal involvement [7]. Wheat, rye, and barley contain the gluten protein, which is toxic for the intestinal cell, and their consumption should be completely eliminated.

2. Case Report

We present a case of a 14-months-old girl, admitted to the Pediatric Surgery Unit of “St. Mary” Children’s Emergency Hospital for acute abdominal pain and suspicion of an intestinal occlusive syndrome. The medical history of the patient revealed lactose intolerance, diagnosed 3 months earlier based on chronic diarrhea, and recurrent pulmonary infections. Family history was negative for chronic or autoimmune diseases. Physical examination at admission showed altered condition, malnutrition (W=8.5kg), poor subcutaneous tissue, with pallor of the skin. The respiratory and cardiac systems were normal, excepting a tachycardia with a heart rate of 120/min. The examination of the digestive system showed an important abdominal distension with tympanic sounds on percussion and diffuse sensitivity at palpation. The appetite was significantly reduced and the intestinal transit was present only for gases, with no stool emission in the last 5 days. There was no history of fever, vomiting, hematochezia or melena.

Initial laboratory tests showed iron deficiency anemia, absence of inflammatory syndrome. Serum liver tests showed moderate-to-severe cytolysis. Hepatic viral serology (anti-HCV antibodies, HBsAg, anti-Epstein Barr virus, anti-Cytomegalic virus) was negative. Bilirubin, renal function tests and electrolytes were within normal
ranges. Stool examination was negative for intestinal infections (negative culture for Salmonella spp., Shigella spp., Yersinia enterocolitica) and parasitosis. Biochemical analysis of the stool suggested an intestinal malabsorption. Urine analysis was within the normal range.

Abdominal ultrasound showed normal structure of the liver, kidneys, spleen and pancreas, distended bowel loops with air content, normal peristaltis, no liquid in the peritoneum. Thoracic-abdominal X-Ray showed the presence of multiple air-fluid levels in the colon and mesogastrium (Figure 1 A and B).

Figure 1. Thoracic-abdominal radiography with multiple air-fluid levels in the colon and mesogastrium. (A: anterior-posterior incidence, B: profile)

Figure 2. Histology of the duodenal mucosa-villous atrophy, crypt hyperplasia, intraepithelial lymphocytes infiltrate

The clinical course was favorable, and a surgical condition was ruled out as a cause of the digestive symptoms. Considering the intestinal malabsorption, the poor weight and the history of recurrent respiratory infections, the diagnosis was oriented towards cystic fibrosis or CD. An iotophoresis test was performed, and the normal values (27 mmol/L NaCl) ruled out the possibility of cystic fibrosis. Further serological tests were conducted highlighting IgA and IgG anti-tissue transglutaminase antibodies (Table 1). The intestinal biopsy showing villous atrophy of the mucosa, crypt hyperplasia and intraepithelial lymphocytes infiltrate confirmed the CD diagnosis (Figure 2). The elevated liver enzymes were considered in the context of CD.

<table>
<thead>
<tr>
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<th>Patient's values</th>
<th>References range</th>
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<tr>
<td>HBG</td>
<td>9.8 g/dL</td>
<td>11.3-14.1 g/dL</td>
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<tr>
<td>MCV</td>
<td>60.8 fl</td>
<td>70-84 fl</td>
</tr>
<tr>
<td>MCH</td>
<td>19.3 pg</td>
<td>22-30 pg</td>
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<tr>
<td>Serum iron</td>
<td>26 µg/dL</td>
<td>63-150 µg/dL</td>
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<tr>
<td>CRP</td>
<td>0.24 mg/L</td>
<td>0-5 mg/L</td>
</tr>
<tr>
<td>ESR</td>
<td>3 mm/h</td>
<td>0-12 mm/h</td>
</tr>
<tr>
<td>Serum total proteins</td>
<td>57 g/L</td>
<td>46-68 g/L</td>
</tr>
<tr>
<td>Albumin</td>
<td>35.9 g/L</td>
<td>30-46.5 g/L</td>
</tr>
<tr>
<td>ASAT</td>
<td>124 U/L</td>
<td>5-35 U/L</td>
</tr>
<tr>
<td>ALAT</td>
<td>273 U/L</td>
<td>5-38 U/L</td>
</tr>
<tr>
<td>IgA anti-tissue transglutaminase antibodies</td>
<td>&gt;320 IU/dl</td>
<td>&lt;20: negative 20-25: equivocal &gt;25: positive</td>
</tr>
<tr>
<td>IgG anti-tissue transglutaminase antibodies</td>
<td>&gt;160 IU/dl</td>
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A gluten-free diet was started together with iron supplementation. Clinical and biological follow-up after 6 months of diet revealed an amendment of symptomatology, with improvement of general condition and ascending weight curve (W=11kg), associated with normal liver enzymes (ASAT 34 U/L, ALAT 38U/L). After other 6 months a second intestinal biopsy was performed, showing normal aspect of the intestinal mucosa.

Table 1. Patient’s laboratory data
3. Discussion

CD is a chronic mucosal inflammation and villous atrophy of small bowel, responsible for symptoms such as malabsorption, steatorrhea, weight loss, vitamin deficiency [8]. Although the disease affects mostly the small bowel, the symptomatology of CD is remarkably various, and the disease may involve many extraintestinal organs, including the liver. The hepatic damage in CD can manifest either as asymptomatic liver enzyme elevations or nonspecific reactive hepatitis, or even as chronic liver disease [9]. Hypertransaminasemia is a frequent diagnostic finding in a significant proportion of children with CD, younger patients being more likely to be affected [10,11]. Our patient had moderate to severe cytolysis that we ascertained as an extraintestinal symptom of the CD, since all hepatic serology was negative and no other signs of liver impairment were present.

Intestinal occlusive syndrome as a presenting symptom of pediatric CD has been rarely reported [12], although, transient intussusception was seen in 20% of patients with proven CD, especially after the increased use of barium studies as investigation toll of malabsorption syndromes [13]. Literature data show that intussusception was the initial manifestation of CD in 57% of the cases, being associated with abdominal pain [14]. There is also evidence that intestinal occlusion was more frequent among children with untreated CD than in the general pediatric population [12,15]. Our patient presented with impaired generally condition, important abdominal distention and absent intestinal transit, suggesting a surgical emergency. The diagnosis of CD should be considered even if there is only a suspicion of intestinal obstruction, especially when a history of failure to thrive is present.

The most effective treatment for CD is the gluten-free diet. Excluding from the diet all products containing wheat, barley and rye results in clinical remission within a few weeks. Normalization of the intestinal mucosa and negativity of anti-transglutaminase antibodies are obtained in several months or even years [16]. Also, abnormal liver tests return to normal in the majority of patients after 1 year of gluten-free diet [10,11]. Our patient presented a favorable evolution once the dietary recommendations were initiated, as well as a normalization of liver enzymes. Gluten-free diet is useful to obtain clinical assessment, but also to prevent long-term complications of CD.

4. Conclusion

Our case report is of interest because the particularity of rare onset of the CD, with acute abdominal pain suggesting a surgical emergency. Children with less common gastrointestinal signs like acute abdominal pain, constipation, iron deficiency anemia should be tested for CD.

References