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Acute abdomen in adult Celiac disease: An intestinal intussusception case

Erişkin Çölyak hastalığında akut batın: Bir entero-enteral intussussepsiyon olgusu

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It is well known that half of the cases admitted to hospital emergency services complain of abdominal pain and that nearly half of these cases are diagnosed with nonspecific abdominal pain. The population of patients with celiac sprue is rarely encountered at the emergency room. Although acute abdominal pain is rarely seen in adult celiac sprue, it should be added to the differential diagnosis. It should also be remembered that acute abdominal pain in these patients could be originating from perforation, intussusceptions and/or intestinal lymphoma. Herein we report a case of adult celiac sprue where successful surgical exploration was carried out because of entero-enteral intussusception.

Key words: Celiac disease, acute abdomen, intussusception, intestinal necrosis

Acil servise başvuran olguların yaklaşık yarısı karın ağrısından yakınmaktadır. Bu hastaların da yarıya yakını nonspesifik karın ağrısı tanısı almaktadır. Çocukluk döneminde veya erişkin çağda karın ağrısı ile başvurabilecek oldukça ender bir hasta grubunu çölyak hastaları oluşturmaktadır. Bu hastalarda akut ağrıya neden olabilecek fizyopatolojik faktör olarak gastrointestinal traktın dönüşümlü otonomik disfonksiyonu hipotezi ileri sürülmektedir. Çölyak hastalığında akut karın ağrısı nadir görülse de ayırıcı tanıda mutlaka değerlendirilmelidir. Bu hastalarda akut karın ağrısının perforasyon, intussepsiyon ve/veya intestinal lenfoma sonucu olabileceği unutulmamalıdır.

Anahtar kelimeler: Çölyak hastalığı, akut batın, intussussepsiyon, nekroz

INTRODUCTION

Acute abdominal pain is one of the most often encountered acute surgical cases in surgical practice (1). Although the vast majority of the cases consist of nonspecific abdominal pain, differential diagnosis is essential. It has been published that patients with Celiac sprue can be admitted to emergency services because of acute abdominal pain. One of the hypotheses as to why the adult Celiac patient applies for medical care is rotating autonomic dysfunction of the gastrointestinal tract (2). Thus, Celiac sprue should be added to the differential diagnosis when evaluating these kinds of cases. In addition, perforation and lymphoma, which could be seen during the chronic phase of the disease, are possible reasons for the acute abdominal pain. In this report, we present a case of adult Celiac sprue where surgical exploration was carried out because of entero-enteral intussusception.

CASE REPORT

Our case is a 57-year-old female who was diagnosed with Celiac sprue at a gastroenterology clinic when she was 40 years old. The history obtained from the patient revealed that she was not following her Celiac diet properly and did not respect her medical therapy. The patient was admitted to the emergency department complaining of abdominal pain, bloating, nausea and vomiting for 48 hours. The pain was diffuse and colic in character. In the physical examination the patient was cachectic. The vital signs were normal except tachycardia and diffuse edema. Auscultation of the abdomen revealed two mechanical intestinal sounds per minute, and palpation revealed tenderness, defense and rebound. Biochemical laboratory studies showed hypoalbuminemia and leukocytosis. Assays for immunoglobulin A anti-gliadin antibody (AGA) (Euroimmun, Lubeck, Germany) and

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antitransglutaminase antibody (Euroimmun, Lubeck, Germany) were positive at >100 RU/ml and >200 RU/ml, respectively (normal = <25 RU/ml and <20 RU/ml, respectively) measured by enzyme linked immunosorbent assay (ELISA). Abdominal X-ray performed in the erect and supine positions revealed air-fluid differential heights suggesting mechanical intestinal obstruction of the small intestine. Abdominal ultrasound revealed intestinal invagination with a bowel-within-bowel appearance that appeared as a target sign on transverse scans and a sandwich sign on longitudinal scans.

An explorative laparotomy was carried out. Surgical exploration revealed an intussusception at the level of the terminal ileum (Figures 1, 2) together with 25 cm of necrosis at this level. There were lymph nodes as large as 2 cm at the mesenterium draining this segment. Since intestinal necrosis was detected, manual reduction to the invaginated segment was not performed. No other intestinal pathology was encountered and partial intestinal resection and an end-to-end anastomosis were performed. The patient was followed at the intensive care unit for two days. During the early postoperative period the patient had two diarrhea attacks (12 times a day, each 150cc in quantity, watery and light brown in color). Liquid and electrolyte treatment was given properly and the patient's medical and dietary regimen was prescribed after consultation with the gastroenterologist and dietician. The patient was sent home on the 15th day postoperatively with no other complications. The specimen sent to the pathology laboratory was examined and revealed hemorrhagic necrosis and ischemic changes, as well as reactive sinus histiocytosis in the 23 lymph nodes that were dissected from the specimen. There were no signs of villous atrophy or cryptic tissue hyperplasia.

Figure 1. Intussusception in the terminal ileum during surgical exploration

Figure 2. Intussusception in the terminal ileum II (proximal part)

DISCUSSION

Celiac sprue is a rarely encountered disease, with a reported incidence of <1% (3), though there are papers from different regions reporting incidences higher than 1%. Malekzadeh et al. (4) reported an incidence of 3-20% in developing countries from the Middle East region, India and North Africa. Tatar et al. (5) investigated tissue transglutaminase antibody positivity in healthy Turkish blood donors and found a 1.3% incidence. The disease is also called "gluten sensitive enteropathy" and the pathological mechanism underlying this disease is augmented immunological response to gluten in individuals who have an underlying genetic predisposition (6). These patients can be identified by non-invasive serological tests. Some of the antibodies held responsible for this disease and their sensitivity-specificities are as follows: immunoglobulin G anti-gliadin antibody (AGA) (sensitivity = 30%-50%, specificity = 30%-70%); immunoglobulin A AGA (sensitivity = 50%-70%, specificity = 50%-85%); and endomysial antibody (EMA) (sensitivity = 70%-90%, specificity = 80%-100%) (7). These serological markers have a 95% correct diagnosis rate (6). Although positive antibodies have a high rate of correct diagnosis, exact diagnosis must be confirmed by histopathological examination of the specimen taken from the duodenum by endoscopy. Villous atrophy of the intestinal tissue is the gold standard in the diagnosis (6). These patients usually have gastrointestinal complaints, which in most of the cases are caused by the underlying malabsorption. Although most of the complaints are gastrointestinal, indirect involvement of other organ systems can rarely cause atypical complaints, such as iron deficiency anemia, osteoporosis,

cryptogenic liver failure or neurological symptoms (3, 6, 7).

Acute abdominal pain is rarely encountered in Celiac sprue patients, and it is reported that 3% of the patients diagnosed with non-specific abdominal pain have Celiac sprue (2). Although the underlying mechanism for the abdominal pain is not yet well known, a few hypotheses exist, among them reversible autonomic dysfunction of the intestinal structures (8-10). Other hypotheses consist of pseudo-obstruction due to hypopotassemia, small-bowel obstruction due to lymphoma (11) or ileus due to idiopathic recurrent acute pancreatitis (12). Of the above mentioned hypotheses, the most supported is that of reversible autonomic dysfunction. This hypothesis suggests that Celiac patients suffer from a motor activity problem caused by neuropathy affecting the extrinsic autonomic system (8). More evidence is needed to support this hypothesis.

Although Celiac sprue is one of the most often encountered immunologic gastrointestinal diseases in western countries, there are very few case reports concerning Celiac sprue and surgical abdominal pain. This is probably due to its prevalence, which is as low as <1%. There is only one case report discussing the relation of acute abdominal pain and adult Celiac disease (9). In this review, it was reported that 3% of the patients admitted to emergency services were diagnosed with Celiac sprue. Whatever the cause, the condition leading to acute abdominal pain should be treated properly.

Intussusception in adults is encountered less often than in children and the patients usually present with chronic abdominal pain up to one month instead of acute pain. In cases where spontaneous reduction of intussusception is seen, the medical history of the patient reveals annual repeating abdominal pain intervals. Although intussusception can present in Celiac disease, this relation is not very well known (13). Radiological studies revealed a 20% incidence of small bowel invagination in untreated Celiac sprue patients (14). Although these patients are diagnosed with intussusception radiologically, most of them present with partial obstruction and mild symptoms. There are only a few case reports considering this relation in the literature (15-20). In cases presenting with mechanical bowel obstruction with suspicion of intussusception, intraluminal malignancy, metastasis, diverticular lesions and tumoral incidents such as polyps and lipomas should be considered.

Although rarely seen, Celiac sprue should be considered when evaluating acute abdominal pain. It should also be kept in mind that abdominal pain in Celiac sprue patients can originate from a chronic ulcerative state. The most important point to bear in mind is that abdominal pain in these patients can be caused by intestinal lymphoma, necrosis, perforation and intussusception.

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