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Cade Report

Coexistence of Celiac Crisis and Food Allergy

Celiac Crisis ve Gıda Alerjisinin Birlikte Varlığı

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ABSTRACT

Celiac disease (CD) may present with different clinical manifestations, and very rarely, it may present with Celiac crisis, which is a fatal complication. An 18-month-old girl was referred to our center due to recurrent diarrheal attacks, inability to gain weight, malaise, fever, and poor general condition. It was learned that the patient, who was followed up in the intensive care unit during his last hospitalization, was examined for the etiology of chronic diarrhea. Celiac disease was suspected, and she was administered a short-term gluten-free diet, but no response was received. The patient, who was in a septic state on admission, had anemia, thrombocytopenia, hypocalcemia, hypokalemia, hypomagnesemia, high acute phase reactants, direct bilirubinemia, coagulopathy and metabolic acidosis in the examinations. When the electrolyte disorder was corrected, total parenteral nutrition was started, and when the general condition stabilized, enteral nutrition was started. Amino acid-based enteral product was preferred in the diet of the patient who had itchy rashes on his body and who had a history of rash after intake of dairy products, and it was observed that the rashes regressed. Celiac serology was positive in the examinations performed for the etiology of chronic diarrhea. Histopathological examination of the biopsy taken from the duodenum was compatible with "Celiac Disease, Marsh Type 3b." Due to celiac disease, severe electrolyte imbalance, coagulopathy and severe malnutrition findings, it was thought that the patient might have celiac crisis. A gluten-free diet was started. After a gluten-free diet and elimination of dairy products, the patient's diarrhea improved, and weight gain occurred. Celiac crisis poses a diagnostic challenge in patients without a previous diagnosis of CD. Celiac crisis, which is a complication of celiac disease with a high mortality, should be considered especially in patients with chronic diarrhea and hyponatremia, hypokalemia, hypoalbuminemia, metabolic acidosis.

Keywords: *Azygos vein, congenital abnormalities, child*

ÖZET

Çölyak hastalığı (ÇH) farklı klinik tablolar ile ortaya çıkabilmekte, çok nadir olarak ölümcül bir komplikasyon olan Çölyak krizi ile karşımıza gelebilmektedir. Onsekiz aylık kız olgu tekrarlayan ishal atakları, kilo alamama, halsizlik, ateş, genel durum bozukluğu nedeniyle merkezimize sevk edildi. Son hastane yatışında yoğun bakım ünitesine takip edilen olgunun kronik ishal etyolojisine yönelik tetkik edildiği, Çölyak hastalığından şüphelenildiği kısa süreli glutensiz beslenme yapıldığı, ancak yanıt alınamadığı öğrenildi. Gelişinde septik tabloda olan hastanın tetkiklerinde anemi, trombositopeni, hipokalsemi, hipopotasemi, hipomagnezemi, akut faz reaktanları yüksekliği, direkt biluribinemi ve koagülopati ve metabolik asidoz mevcuttu. Elektrolit bozukluğu düzeltildiğinde total parenteral beslenme başlandı ve genel durumunun stabilleştiğinde enteral beslenmeye geçildi. Vücudünda kaşıntılı döküntüleri dikkat çeken, öyküsünde süt ürünleri alımı sonrasın–

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da döküntü olan olgunun beslenmesinde aminoasit bazlı enteral ürün tercih edildi ve döküntülerin gerilediği gözlendi. Kronik ishal etyolojisine yönelik yapılan tetkiklerde Çölyak serolojisi pozitif saptandı. Duodenumdan alınan biyopsinin histopatolojik incelemesi "Çölyak Hastalığı, Marsh Tip 3b" ile uyumlu bulundu. Çölyak hastalığı, ciddi elektrolit dengesizliği, koagülopatisi ve ağır malnütrisyon bulguları nedeniyle olguda Çölyak krizi olabileceği düşünüldü. Glutensiz diyet başlandı. Glutensiz diyet ve süt ürünleri eliminasyonu sonrasında olgunun ishal düzeldi, kilo alımı gerçekleşti. Çölyak krizi, önceden ÇH tanısı olmayan hastalarda tanısal zorluk oluşturmaktadir. Özellikle kronik ishali olan ve hiponatremi, hipokalemi, hipoalbüminemi, metabolik asidoz tablosunda gelen hastalarda, Çölyak hastalığına ait mortalitesi yüksek bir komplikasyon olan akut Çölyak krizi düşünülmelidir.

Keywords: *Azigos veni, doğumsal anomaliler, çocuk*

INTRODUCTION

Celiac disease is an autoimmune, chronic disease that occurs after gluten ingestion, affects the small intestine in genetically predisposed individuals, and can lead to multi-organ involvement (1). It is an enteropathy characterized by the diversity of clinical manifestations, the presence of disease-specific antibodies and "Human leucocyte antigen" (HLA)-DQ2/HLA-DQ8 haplotypes (2). Malabsorption occurs as a result of damage to the villi. The disease may occur at any time of life with different clinical manifestations. Celiac crisis, which is a fatal complication very rarely, can be encountered (3,4).

CASE REPORT

An eighteen-month-old girl was admitted with recurrent diarrheal attacks, inability to gain weight, weakness and fever. When we look at the history of the patient who had her first diarrhea attack at the age of ten months, it was determined that she was born 3000 g (10-25p) in the hospital by normal spontaneous vaginal delivery at term. It was learned that complementary feeding was started when she was five months old, and she had a Rotavirus infection and intussusception when she was one year old. In the last hospitalization, hypokalemia, hypocalcemia, hypoalbuminemia, coagulopathy, and resistant metabolic acidosis were found in the examinations, diarrhea did not regress, and the patient was followed up in the intensive care unit because of his general condition disorder. Endoscopic examination was performed for the etiology of chronic diarrhea, duodenal mucosa was normal; It was learned that the colon lumen was significantly dilated and the colonic mucosa was evaluated as normal. It was learned that the tissue transglutaminase IgG and IgA was negative and 16.3 U/mL, respectively, duodenal histopathology was consistent with "Gluten Sensitive Enteropathy Type 3b", and so gluten-free diet was applied and no response was obtained. It was learned that the patient who developed abdominal distension and dilated intestinal loops on abdominal X-ray was evaluated for Hirshprung's disease and Neurointestinal dysplasia, ganglion cells were positive in full-thickness rectal biopsy, diagnostic laparotomy was performed in terms of congenital intestinal anomalies, and no pathological findings were found. The patient whose diarrhea did not regress and whose complaints were fever, restlessness, and general condition disorder was referred to our center.

On physical examination, she was conscious, weak and weak. The patient's body temperature was 38 C, and she was septic; weight: 7500 g (<3p, -2.8 SDS), height: 75 cm (<3p, -2 SDS), head circumference: 43.5 cm (<3p, -2.8 SDS), weight for height: 79%. On examination, she looked pale, her abdomen was distended, defense and rebound were negative, and findings consistent with minimal ascites. There was no organomegaly, a laparotomy scar was observed in the right and left middle quadrants, there were rashes compatible with eczema on both arms, muscle atrophy was noted in the extremities.

Anemia, thrombocytopenia, hypocalcemia, hypokalemia, hypomagnesemia, high acute phase reactants, direct bilirubinemia and coagulation disorder were found in the examinations (Table 1). Sepsis and Disseminated Intravascular Coagulation were considered due to thrombocytopenia, INR length, D-dimer increase and low fibrinogen. Fresh Frozen Plasma was given and vitamin K was made. When the electrolyte disorder was corrected, total parenteral nutrition was started. Enteral nutrition was started on the 4th day when the general condition of the patient stabilized. Amino acid-based enteral product was preferred in the nutrition of the case, which had itchy rashes on her body because of the history of rash after intake of dairy products. His diet was adjusted according to the daily stool amount. Enteral nutrition was gradually increased, while parenteral nutrition was decreased and discontinued.

Celiac serology was positive in the tests sent for the etiology of chronic diarrhea (Table 2). The patient was thought to have Celiac crisis due to Celiac disease,

severe electrolyte imbalance, coagulopathy and severe malnutrition findings. A gluten-free diet was arranged, and table foods were started. Associated endocrinopathies were investigated and the endocrine department was consulted with thyroid hormone values. Subclinical hypothyroidism was considered, and levothyroxine treatment was started. IgG and IgM values were low for his age, IgA was normal, and he was consulted with the pediatric immunology department. It was thought that the patient's humoral and cellular immunity was affected due to malnutrition, and follow-up was recommended. Chronic diarrhea gene panel and immunodeficiency genetic panel were sent and the results were normal. HLA tissue typing tests were requested. HLA DQB1*02, HOMOZYGOT++ resulted.

It was preferred not to perform an upper gastrointestinal system endoscopy immediately in the patient who had significant malnutrition and whose clinical condition took time to stabilize. Biopsy materials taken during the endoscopy and colonoscopy performed two months ago were re-evaluated. The findings observed in duodenal biopsy were compatible with "Marsh Type 3b according to Celiac Disease, Modified Marsh classification." Diarrhea improved and weight gain occurred in the patient who was fed a gluten-free diet. The case was discharged on the 16th day of hospitalization.

DISCUSSION

The term "Celiac crisis" was first introduced in the literature in 1953, when Anderson and di Sant'Agnese reported 35 cases of children with recurrent diarrhea with a 9% mortality rate (5). Celiac crisis is an urgent, severe and life-threatening complication that is challenging to diagnose. Although it is most commonly seen in early childhood, as in our case, it can also occur in adults and older age groups (6). Clinically, patients experience severe diarrhea, dehydration, metabolic disorders, hypokalemia, hypomagnesemia, hypocalcemia, and hypoproteinemia. Factors such as severe malnutrition, infection, hypoproteinemia, intestinal bacterial overgrowth, decreased intestinal motility due to anticholinergic drugs, operations, and pregnancy increase the incidence of celiac crisis (5, 7). Radlovic et al. in their study, in which they described six cases diagnosed with celiac crisis, they reported that their patients were accompanied by rotavirus gastroenteritis, Salmonella enteritis, and severe malnutrition (6). Our case had rotavirus infection, intussusception and severe malnutrition.

The relationship between food allergy and other allergic diseases and celiac disease (CD) has not been clarified yet (8,9). Although there are conflicting results on the relationship between allergic diseases and CD, recent studies have shown that celiac patients have higher allergic symptoms compared to the general population (9); CD was more common in individuals with atopic disease, and atopic diseases were more common in individuals with CD. Similarly, asthma and CD have also been shown to be related (8). Pilon et al. showed that children with severe food allergies (dairy, egg, and wheat allergy) have a five times greater risk of CD compared to controls (9). Our case had clinical findings of food allergy and responded well to the amino acid-based formula.

It is hypothesized that impaired intestinal mucosal permeability may cause increased efflux of dietary antigens from the intestinal mucosa by stimulating food-induced hypersensitivity in celiac patients. In fact, the reverse may also be true, as the impaired intestinal permeability observed in some allergic patients may break tolerance to gluten, promoting CD in genetically predisposed individuals. Allergy and CD may share a similar genetic background (9). However, the pathogenesis of the two diseases is quite different, and there is no apparent common pathway (8).

Table 1. Hemogram, biochemistry and coagulation values

WBC: 11 700 /mm3	Procalsitonin: 3.1 mcg/L
Hgb: 9,5 g/Dl	CRP: 27.8 mg/L
Plt: 80 000 /mm3	Na: 137 mmol/L
Glu: 67 mg/dL	K: 3 mmol/L
Alb: 2.7 g/dL	Ca: 7.6 mg/dL (corrected.ca: 8.6)
Urea:3 mg/dL	P: 2.3 mg/dL
Creatinin: 0.5 mg/dL	Mg: 1.4 mg/dL
Uric acid: 1.1 mg/dL	INR:3.5
ALT: 25 U/L	PT: 22.1 sn
AST: 91 U/L	APTT: 41.3
T.Bil :2 mg/dL	Fibrinogen: 88.4 mg/dL
D.bil: 1,1 mg/dL	D-dimer: 1410

Table 2. Laboratory results to disclose chronic diarrhea etiology

Total IgE: 118 IU/mL	fT4: 0.70 ng/dL (0.5-2.3)
IgA: 1.05 g/L	fT3: 3.92 ng/L (4.3- 6.8)
IgM:0.37 g/L	TSH: 7.3 mIU/L (0.3-5.6)
IgG:4.01 g/L	Anti TPO: 5.1 IU/mL
Nutrition panel 1 ve 2: < 0.1 (class 0)	Anti Tiroglobulin: <0.9 IU/mL
EBV DNA: negative	Cortisol: 9.83 mcg/dL
CMV DNA: negative	Stool culture: negative
Viral serology tests: negative	Stool ph: 6
Brucella agglutination test: negative	Stool reductant material: negative
Tissue transglutaminase IgG: 5 RU/mL	Steatorrhea: negative
Tissue transglutaminase IgA:23 RU/mL	Zinc: 40 mcg/dL
Anti-Endomysium IgG: negative	
Anti-Endomysium IgA: 1/20	
Anti-deamidated gliadin peptide IgG :1/100	
Anti-deamidated gliadin peptide IgA: 1/100	

With the increase in awareness of celiac crisis, the incidence of complications is gradually decreasing thanks to rapid diagnosis and treatment. In the literature, there are studies suggesting that supportive treatment and a gluten-free diet may not respond in severe cases, and systemic steroid treatment should be given to patients. Ciacci et al. showed that using budesonide improved both histology and absorption parameters in CD, possibly by restoring brush border epithelial enzymes and reducing mucosal inflammation (10). A short course of prednisone or budesonide may be considered when standard therapy does not produce rapid improvement. However, the use of corticosteroids may exacerbate electrolyte disturbances when they contribute to the depletion of potassium, magnesium, and phosphate. The only evidence-based intervention in a celiac crisis is a gluten-free diet; however, the use of additional interventions such as corticosteroids and parenteral nutrition has been reported in the literature (5). In our patient, treatment was arranged for accompanying sepsis, DIC, food allergy; TPN support was given, and recovery was observed with a gluten-free diet, and steroid treatment was not required.

Celiac crisis is a diagnostic challenge in patients without a previous diagnosis of CD. Acute Celiac crisis, which is a complication of Celiac Disease with a high mortality, should also be considered, especially in patients with chronic diarrhea who present with hyponatremia, hypokalemia, hypoalbuminemia, metabolic acidosis, and in this respect, investigations and treatment should be arranged quickly. The parents of the patient consent was obtained for this case study.

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The parents' of this patient consent was obtained for this study.

Patient Consent Form / Hasta Onam Formu

The parents' of this patient consent was obtained for this study.

Conflict of Interest / Çıkar Çatışması

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