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Case Report

A Rare Cause of Constipation and Acute Malnutrition: Primary Intestinal Lymphoma

Nurşah Özdemir[1], Gökhan Tümgör[2], Melikhan Çerçi[1], Ayşe Özkan[3], Gülay Sezgin[3], İbrahim Bayram[3], Mehmet Turgut[1]

ABSTRACT

Constipation is the group of disease that is most frequently consulted with experts in pediatric gastroenterology. Abnormal neurological examination, non-intestinal symptoms or cases unresponsive to treatment should be stimulative and suggestive of another underlying cause to the doctor. Mature B-cell malignant disease (Burkitt lymphoma) forms 35-40 % of all non-Hodgkin lymphoma patients. Burkitt lymphoma can be a mass in gastrointestinal system. In this paper, a four-year-old boy who was hospitalized for 17 days in other centers with a diagnosis of malnutrition and constipation and diagnosed as primary intestinal lymphoma in our clinic is presented. Primary intestinal lymphomas discussed under the light of literature.

Key words: *intestinal lymphoma, constipation, acute malnutrution*

Konstipasyon ve Akut Ağır Malnutrisyonun Nadir Nedeni: Primer İntestinal Lenfoma

ÖZET

Konstipasyon çocuk gastroenteroloji uzmanlarına konsülte edilen en sık hastalık grubudur. Anormal nörolojik muayene, bağırsak dışı semptomlar, tedaviye yanıtsız olgular doktor için uyarıcı olmalı ve altta yatan bir nedeni düşündürmelidir. Lenfomalar, Hodgkin ve non Hodgkin lenfoma olarak ayrılmaktadır. Matür B-hücreli malin hastalık (Burkitt lenfoma (35-40%) tüm non-Hodgkin lenfoma hastalarının 35-40 % ını oluşturur. Bu yazıda, dış merkezlerde malnutrisyon ve konstipasyon teşhisleri ile 17 gün hospitalize edilen ve daha sonra kliniğimizde primer intestinal lenfoma tanısı alan, 4 yaşında erkek hasta sunuldu. Primer intestinal lenfomalar literatür ışığında tartışıldı.

Anahtar kelimeler: *İntestinal lenfoma, konstipasyon, akut malnutrisyon*

INTRODUCTION

Constipation is the group of disease that is most frequently consulted with experts in pediatric gastroenterology. Most cases of constipation have no structural, endocrine or metabolic cause and these cases are called idiopathic or functional constipation. Reasons such as meconium delay, growth retardation, empty rectum, tight anal sphincter are seems to be responsible for constipation that starts in children before 12 months of age. Nevertheless, abnormal neurological examination, non-intestinal symptoms or cases unresponsive to treatment should be stimulative and suggestive of another underlying cause to the doctor (1). Lymphomas are divided as Hodgkin and non-Hodgkin lymphoma (NHL). According to the classification of NHL in childhood that made as a result of the investigations of immunohistochemical, cytogenetic and molecular biologic done in 2008; it is classified as malignant disease of lymphoid precursor (T-lymphoblastic lymphoma (15-20%), B-lymphoplastic lymphoma (3%)), Mature B-cell malignant disease (Burkitt lymphoma (BL) (35-40 %), Diffuse large B-cell lymphoma (15-20%), primary mediastinal B-cell lymphoma (1-2%), pediatric follicular lymphoma (rare), marginal pediatric nodal zone T-cell lymphoma

[1]*Adana Numune Education and Research Hospital, Department of Pediatrics, Adana, Turkey,* [2]*Çukurova University School of Medicine, Department of Pediatric Gastroenterology, Adana, Turkey,* [3]*Çukurova University School of Medicine, Department of Pediatric Oncology, Adana, Turkey*

Correspondence: *Nurşah Özdemir, Adana Numune Education and Research Hospital, Department of Pediatrics, Adana, Turkey*
Telephone Number: +90- 322- 355 01 01
E-mail address: nursahozdemir@yahoo.com

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rare and mature T-cell lymphoma ALK positive (15-20%), peripheral T-cell lymphoma. Diffuse large B-cell lymphoma and Burkitt lymphoma can also be abdominal involvement. In addition, Burkitt lymphoma (BL) can be masses in gastrointestinal system (2). In this paper, 17 days hospitalized in other centers with a diagnosis of malnutrition and constipation and diagnosed primary intestinal lymphoma case is presented in our clinic.

CASE

A four-year-old boy who has no complaints previously consulted with an abdominal pain, weight loss (3 kilograms), vomiting, and constipation continued for a month. Approximately one month ago before cases story began constipation started, defecate started with an enema every 3-4 days, because bilious vomit and inability to defecate, 17 days hospitalized with diagnosis of ileus is followed in a foreign center, discharged with oral lactulose upon improvement of the overall situation and on the reduction of complaints. Constipation and vomiting continued during 15 day period at home. The medical history and family history of the case was unremarkable. In physical examination; Body weight: 13.8 kilograms (10-25 p) Height: 104 cm (50-75 P), body temperature: 36.7oC, pulse rate: 90/min, blood pressure: 100/60 mmHg was determined. General condition of patient was moderate, conscious, pale, weak and cachectic. The eyeballs were sunken and mucous membranes were dry. Left tonsillar was hypertrophy compared to the other and there were bleeding focal over it. The cardiovascular system and respiratory system examination was normal. In the examination of abdomen, abdomen was distended, decreased bowel sounds, liver and spleen was nonpalpable. Neurological system was normal. In laboratory investigation of the case, anemia, leukocytosis, thrombocytosis, hypocalcemia, hypoalbuminemia were detected (Table 1). Also GGT, LDH, amylase and uric acid elevation were detected. In abdominal ultrasound hypoechoic areas was observed that led to diffuse thickening of the intestinal loops, lobulated appearance of the wall. In the upper gastrointestinal system endoscopy, the gastric mucosa was hyperemic and edematous. Gastric rugal folds were enlargement; there were polypoid lesions, ulcers above it, and their diameters ranging between 0.5-1 cm. Bulb and duodenum also had polypoid lesions (Figure 1). When biopsy was taken, tissue felt to be too hard. As a result of endoscopy, the intestinal lymphoma suspected patient was sent to Cukurova University Medical Faculty, department of Pediatric Oncology. The patient's intestinal biopsy was evaluated as BL by department of Pathology. BL was defined as bone marrow involvement in bone marrow aspiration (Figure 2). Bone marrow biopsy was evaluated as lymphomatous bone marrow infiltration by department of Pathology. The patient began che-

Table 1. Hematologic and blood chemical values

Hemoglobin (gr/dl)	6,6
White cell count (/mm3)	15300
Platelet (/mm3)	795000
Calcium (mg/dl)	7,7
Amylase (IU/L)	786
SGPT (IU/L)	47
SGOT (IU/L)	15
Alkaline phosphatase (IU/L)	178
GGT (IU/L)	130
Total Protein (gr/dl)	4,5
Albumin (gr/dl)	2,8
LDH (IU/L)	1062
Urik acid (mg/dl)	13,7

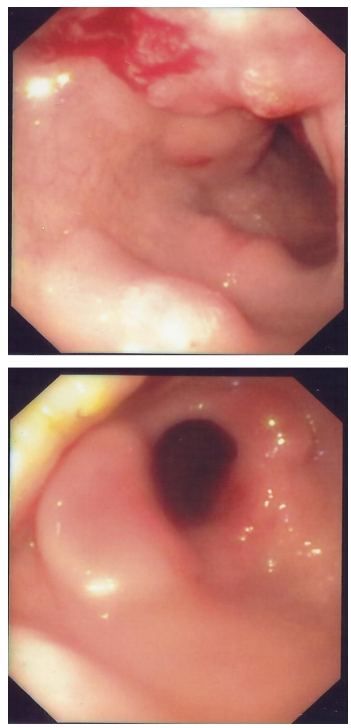

Figure 1. *The gastric mucosa was hyperemic and edematous, ranging from 0.5-1 cm in diameter, polypoid lesions ulcers above it.*

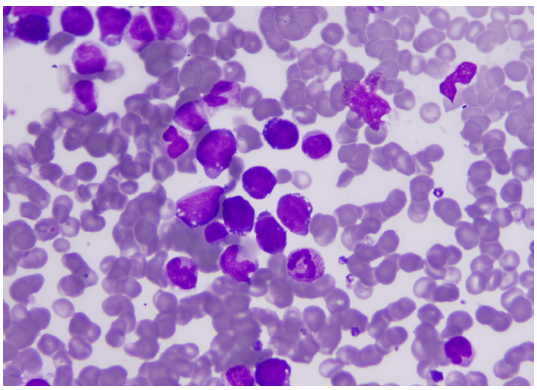

***Figure 2**. Bone marrow involvement in case*

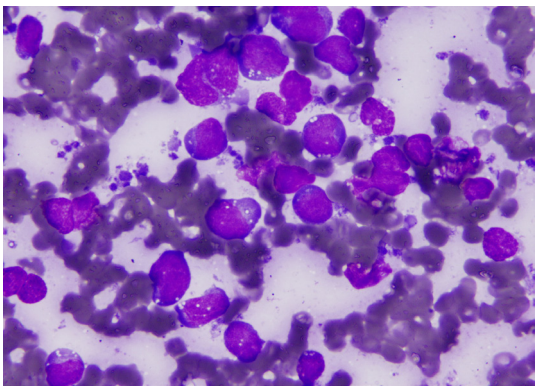

***Figure 4**. The lower jaw, gum infiltration*

motherapy. In the 2nd month of chemotherapy treatment leukemic transformation ALL-L3 development was observed in bone marrow aspiration (figure 3). In the 5th month of chemotherapy jaw involvement (figure 4), blast cells in examination of brain cerebrospinal fluid, 3x2 cm sized lesion and leptomeningeal consistent with lymphoma on posterior wall of the right mastoid sinus on cerebral magnetic resonance imaging and contrast enhancement in pachymeningeal structures was detected. Chemotherapy was continued for patient; however lesion on the jaw of the patient rapidly grew and died in the 6th month of follow-up due to sepsis.

DISCUSSION

Approximately 25% of patients admitted to pediatric gastroenterology is constipation (3). World prevalence of childhood constipation is 0.3-28 % (4,5). Most childhood constipation is functional. However, 5-10% of cases, abnormalities of the colon and rectum, spinal cord abnormalities, gastrointestinal diseases, neuropathic, or constipation may occur due to systemic diseases and drugs (1). Small intestine constitutes 75 % of the entire gastrointestinal tract and 90 % of the mucosal surface. Looking at the place of lymphomas in all the gastrointestinal malignancies, fewer than 2% seen in the small intestine and 35-40 % of these are known to be lymphomas. Endemic and sporadic BL residences are different. While the most common localization of endemic BL is jaw area, abdomen is most commonly seen area of sporadic BL. Many sporadic patients are young males 5-10 aged. Often, they consult with abdominal pain and abdominal distension, nausea and vomiting, and gastrointestinal bleeding, or rarely with intestinal perforation. Lymphomas in children are associated with ileocecal region, approximately between 25-30% of patients with right lower quadrant mass and acute pain with intussception consult. Sometimes it can be confused with acute appendicitis. Sometimes, patients with acid fluid associated with massive disease, abdominal disease, mesenteric, retroperotoneal region, kidneys, ovaries and peritoneal region may come. Other affected regions in Sporadic BL are the head-neck and pharyngeal, nasopharyngeal bölgelar, paranasal sinuses and tonsils. The status associated with jaw is determined at a rate of 10%. Bone marrow involvement rate is 20%. Other rare-held areas were including testicles, breasts, thyroid gland, skin, epidural region, chest, bone and pankreas (2.7). In the literature, the majority of primary gastrointestinal lymphomas are adult patients, and in the published ar-

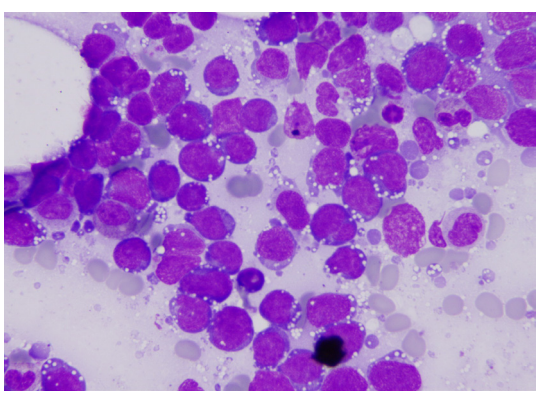

***Figure 3**. Leukemic Transformation (FAB; ALL-L3)*

ticles related to them; Zhan and colleagues (8) studied the clinical features of primary intestinal lymphoma 309 cases and found that the most frequent cases applied was abdominal pain (71%), abdominal mass (14%), vomiting (10%), melena (10%) and ignition (9%) complaints. In this article it is reported that the most common malignant tumors is small bowel disease (40%) and intestinal lymphoma rate is 9 % between malignant tumors. Zinhani and colleagues (9) reported that the most frequent complaint in primary intestinal lymphoma patient is abdominal pain and respectively, nausea, vomiting, weight loss and constipation.

There are a few reports about pediatric patients with primary small bowel lymphoma in the literature (10-12). Watanabe and colleagues (10) reported the treatments of the four children patients with detection of thickening of small bowel wall and mass in the abdomen, but with no obstruction evidence and symptoms. Takahashi and colleagues (11) reported 47 cases of gastrointestinal lymphoma. Two of these cases are in stomach, 17 are in small intestine, 20 are in ileocecal area, 7 are in thick intestine and one case has multifocal settled in small intestine and also in thick intestine are reported. They reported 35 of these cases are reported in BL. Lewin and colleagues (12) when they posted the results of 117 cases with gastrointestinal lymphoma; they reported only nine of these cases were under 16 years of age with histological type and in terms of gender distribution with site of involvement are different with adults. One month before of applying patients to our clinic, in other clinic prolonged constipation, fatigue, vomiting and presented with weight loss and diagnosis of ileus in bilious vomiting deposited for 17 days with discharging of giving treatments for constipation. Cachectic case was admitted have not improved despite treatment of lactulose constipation in our clinic.

They reported diffuse involvement in very little part of primary intestinal cases in literature (11). During the application of cases process was observed being large of the left tonsil. There was no lymphadenopathy in examination of lymphatic system. In examination of the abdominal mass there was not tenderness and liver spleen were not palpated. The stomach and duodenum involvement was observed in upper GIS endoscopy and were confirmed by pathological examination. Bone marrow, central nervous system and also spread to the jaw were detected in returns of ALL-L3 to the follow-up of cases.

Constipation is a common symptom in childhood. If acute onset constipation of a child complaints weight loss and accompanied of the acute malnutrition cases, then primary gastrointestinal lymphoma of a malignancy in the different diagnosis are needed to be considered though being of rare in childhood.

Conflict of interest/Funding

No authors have potential conflicts of interest, including financial interests or relationships and affiliations relevant to the subject of this manuscript.

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