Primary intestinal diffuse large B-cell lymphoma forming multiple lymphomatous polyposis

Figen BARUT¹, Nilüfer ONAK KANDEMİR¹, Kemal KARAKAYA², Neslihan KÖKTEN¹, Şükrü Oğuz ÖZDAMAR¹

INTRODUCTION

The gastrointestinal system (GIS) is the most commonly encountered localization for extranodal non-Hodgkin lymphomas (1-3). GIS lymphomas, which constitute approximately 5-10% of all GI neoplasms, are commonly observed as solitary lesions (1,4), and the majority are diffuse large B-cell lymphomas (DLBCL) (2,5,6). Other BCLs that tend to arise in the GIS include mantle cell lymphoma, which presents as lymphomatous polyposis, Burkitt’s lymphoma, and BCLs associated with immunodeficiency states (7). Most of these lymphomas are high-grade tumors (6).

CASE REPORT

Multipl lenfomatöz polipozis oluşturan primer intestinal diffüz büyük B hücreli lenfoma


Key words: Multiple lymphomatous polyposis, intestinal involvement, diffuse large B-cell lymphoma

Anahtar kelimeler: Multipl lenfomatöz polipozis, intestinal tutulum, diffüz büyük B hücreli lenfoma

INTRODUCTION

The gastrointestinal system (GIS) is the most commonly encountered localization for extranodal non-Hodgkin lymphomas (1-3). GIS lymphomas, which constitute approximately 5-10% of all GI neoplasms, are commonly observed as solitary lesions (1,4), and the majority are diffuse large B-cell lymphomas (DLBCL) (2,5,6). Other BCLs that tend to arise in the GIS include mantle cell lymphoma, which presents as lymphomatous polyposis, Burkitt’s lymphoma, and BCLs associated with immunodeficiency states (7). Most of these lymphomas are high-grade tumors (6). Primary GIS lymphomas, displaying multifocal and skip involvement, represent a distinctive en-
tity defined as multiple lymphomatous polyposis (MLP), which is characterized by multiple polypo-
id lesions affecting the same or different GIS seg-
ments, and it is a quite rarely encountered deve-
lopmental pattern (1,4,8-15). First described by
Cornes in 1961, this developmental pattern ac-
counts for 1-2% of GIS lymphomas (1,4,8,10-12).
MLP is thought to represent mantle cell lympho-
ma of the GI tract (13,16-19). MLP cases associ-
ated with DLBCL are very rare and are also contro-
versial (9).

In the current study, a quite rare primary intesti-
nal DLBCL case, with macroscopic features and
clinical aspects imitating Crohn’s disease and
displaying a cobblestone-like appearance, associa-
ted with MLP is presented, together with the cli-
nical and pathological features.

CASE REPORT

A 25-year-old male patient applied to a university
hospital with nausea-vomiting and weight loss.
After evaluation of the patient with radiological
examinations, the patient underwent an emergent
operation due to intestinal obstruction. Frozen
section of the solid mass obstructing the lumen of
the proximal jejunum demonstrated “malignant
tumor, lymphoma?”, and jejunoileal resection was
performed during the operation. Multiple ulcer-
ated, skip lesions, with patchy polypoid appearance
and infiltrating the serosa, localized 4 cm and 2
cm from the proximal and distal surgical margins,
respectively, and ranging in diameter from 0.5-2.5
cm, shrinking the lumen in one region, were obser-
vied within the resected intestinal specimen, which
displayed patchy, healthy-appearing mucosal are-
as (Figures 1, 2). Microscopic examination of the
cross-sections of the multiple masses, which were
ulcerated and polypoid in appearance, revealed a
tumor infiltrating the intestinal tissue and exhibi-
ting a diffuse pattern (Figures 3, 4). The tumor tis-
sue was found to be comprised of pleomorphic, aty-
tical lymphoid cells with eosinophilic cytoplasm,
marked nucleoli and vesicular nuclei (Figure 5). A
B-cell phenotype immunoreaction was observed
with vimentin, LCA, CD20, and CD79a in those
cells (Figure 6). No immunoreaction was observed
in the tumor tissue with pankeratin, epithelial
membrane antigen (EMA), Ber-Ep4, bcl-2, CD10,
CD30, CD3, CD5, CD23, cyclin D1 or S100 prote-
in.

The diagnosis was DLBCL, and two metastatic
lymph nodes were determined. The patient was

Figure 1. Gross appearance of the jejunoileal resection specimen containing multiple polypoid lesions.

Figure 2. Gross appearance of segmental multiple polypoid lesions with patchy ulcerations and varying from 0.5-2.5 cm in di-

Figure 3. Neoplastic infiltration resembling polypoid appearan-
ce (H&E, x100).
administered six cycles of chemotherapy and is still under follow-up in the Department of Oncology.

**DISCUSSION**

Primary GIS lymphomas are rarely encountered lesions (1,2,4). These lymphomas are usually observed as ulcerative, superficial, polypoid, or diffuse lesions. The areas frequently involved by GIS lymphomas are mainly the stomach, followed by the duodenum; the jejunum, ileum and colon are the other involved areas, as in our case (1-4,12).

Although there has been controversy about whether MLP is a variant of lymphoma with heterogeneous histological features or a distinct clinicopathological disorder, multiple studies recently supported the conclusion that this lesion is a single distinct disease (12,14,17,18). This disease is frequently encountered between 55-64 years of age and cases usually display complaints related with involvement of the GIS, such as abdominal pain, diarrhea, obstruction, and hematochezia (1,4,12,16). Our case applied to a university hospital with signs of intestinal obstruction.

**Figure 4.** Atypical lymphocytes infiltrating the intestinal tissue (H&E, x200).

**Figure 5.** Atypical lymphocytes with hyperchromatic nuclei and eosinophilic cytoplasm (H&E, x400).

**Figure 6.** B-cell phenotype with LCA, CD20, CD79a expression, and without CD5 expression (B-SA peroxidase, DAB, LCA, x400; CD20, x200; CD79a, x400; CD5, x400).
Multiple lymphomatous polyposis (MLP), featuring multiple mucosal polyps, frequently affects a broad area by involving multiple segments of the GIS (4,20). As in our case, MLP is a disorder that involves single or several segments of the intestine, and features smooth and sessile multiple polypoid structures with diameters ranging between 2 mm and a few centimeters. Large polyps are usually located in the ileocecal region and exhibit ulceration. Both radiologic and endoscopic appearances in these cases mimic inflammatory bowel diseases, particularly Crohn’s disease (4,12,15). Besides these benign lesions, similar macroscopic features are observed in malignancies such as mantle cell lymphoma, mucosa-associated lymphoid tissue (MALT) lymphoma, follicular lymphoma, B-cell chronic lymphocytic leukemia, and adult T-cell lymphoma, and those malignancies should be considered in the differential diagnosis as well (4,8,12). Mantle cell lymphoma is a low-grade neoplasm also known as intermediate lymphocytic, mantle zone, centrocytic, and diffuse small cleaved cell lymphoma. The typical tumor cell phenotype (pan-B+, CD5+, cyclin D1+ and CD10-) along with Bcl-1 rearrangement indicates that MLP is the GI counterpart of mantle cell lymphoma (17,19). These features are important in the differential diagnosis of DLBCL (19).

Diffuse large B-cell lymphoma (BCL), besides constituting the largest subgroup of non-Hodgkin lymphomas, is the most frequent type displaying the extranodal involvement. The most common extranodal involvement localizations reported for DLBCL cases are the stomach, bone, skin, small intestine, lung, heart, liver, and the genitourinary system. DLBCL usually forms solitary lesions and rarely displays a MLP pattern (5).

The prognosis of MLP is poor, and most of the cases are diagnosed in advanced stages. Treatment approaches are similar to those for high-grade aggressive lymphomas, and in addition to surgery, various chemotherapy regimens are administered. The mean survival rate has been reported to be approximately 20-30 months after initial diagnosis (2,4,11,12,20).

We presented herein a case with a very rare primary intestinal DLBCL, which is a subgroup of lymphomas, associated with MLP pattern. It is emphasized that, although very rare, this entity must be ruled out in the differential diagnosis of patients presenting with the signs and symptoms of Crohn’s disease.

REFERENCES
15. Isaacson PG, MacLennan KA, Subbuswamy SG. Multiple lymphomatous polyposis of the gastrointestinal tract. Histopathology 1984; 8: 641-56.
