CASE REPORT

TJG
The Turkish Journal of Gastroenterology

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

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Multifocal and skip involvement is quite a rare developmental pattern for primary gastrointestinal lymphomas. A 25-year-old male patient with diffuse large B-cell lymphoma of the small intestine, with macroscopic features and clinical aspects imitating Crohn's disease and attracting attention with cobblestone-like appearance, is presented herein together with the clinical and pathological features.Multiple ulcerated lesions were also observed infiltrating the serosa with polypoid appearance, 2.5 cm in largest diameter, within the resected jejunoileal specimen, which displayed patchy, healthy-appearing mucosal areas. In microscopic examination, a tumoral infiltration was observed comprised of pleomorphic, atypical lymphoid cells with abundant eosinophilic cytoplasm, marked nucleoli and vesicular nuclei. A B-cell phenotype immunoreaction was observed by vimentin, LCA, CD20, and CD79a in those atypical cells. The diagnosis of the case was diffuse large B-cell lymphoma.The possibility of the presence of this disorder, although rare, is emphasized here for patients applying to the hospital with the signs and symptoms of Crohn's disease.

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

Multipl lenfomatöz polipozis oluşturan primer intestinal diffüz büyük B hücreli lenfoma

Multifokal ve atlamalı tutulum, primer gastrointestinal lenfomalar için oldukça nadir görülen bir gelişim paternidir. Klinik ve makroskopik özellikleriyle Crohn hastalığını taklit eden kaldırım taşı görünümü ile dikkat çeken bir ince barsağın multifokal diffüz büyük B hücreli lenfoma olgusu, klinik ve patolojik özellikleri ile beraber sunulmuştur. 25 yaşındaki erkek olgunun jejunoileal rezeksiyon materyalinde, arada sağlam görünümde mukoza alanları izlenen, en büyüğü 2,5 cm çapında olan, ülsere, serozayı da infiltre eden polipoid görünümde multipl lezyonlar izlendi. Mikroskopik incelemede, belirgin nükleollü, veziküler nükleuslu, geniş eozinofilik sitoplazmalı, pleomorfik, atipik karakterde lenfoid hücrelerden oluşmuş tümöral infiltrasyon gözlendi. Bu atipik hücrelerde vimentin, LCA, CD20, CD79a ile B hücre fenotipinde immün reaksiyon izlendi. Olguya, diffüz büyük B hücreli lenfoma tanısı verildi. Crohn hastalığı belirti ve bulguları ile başvuran hastalarda nadir de olsa bu antitenin olabileceği vurgulanmıştır.

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-

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Manuscript received: 21.01.2010 **Accepted:** 22.03.2010

Turk J Gastroenterol 2011; 22 (3): 324-328
doi: 10.4318/tjg.2011.0220

This manuscript was presented at the 19th National Pathology Congress.

tity defined as multiple lymphomatous polyposis (MLP), which is characterized by multiple polypoid lesions affecting the same or different GIS segments, and it is a quite rarely encountered developmental pattern (1,4,8-15). First described by Cornes in 1961, this developmental pattern accounts for 1-2% of GIS lymphomas (1,4,8,10-12). MLP is thought to represent mantle cell lymphoma of the GI tract (13,16-19). MLP cases associated with DLBCL are very rare and are also controversial (9).

In the current study, a quite rare primary intestinal DLBCL case, with macroscopic features and clinical aspects imitating Crohn's disease and displaying a cobblestone-like appearance, associated with MLP is presented, together with the clinical 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 ulcerated, 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 observed within the resected intestinal specimen, which displayed patchy, healthy-appearing mucosal areas (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 exhibiting a diffuse pattern (Figures 3, 4). The tumor tissue was found to be comprised of pleomorphic, atypical 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 protein.

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

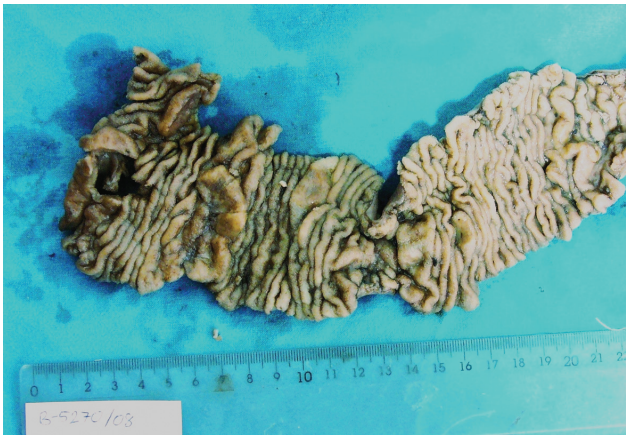

Figure 1. Gross appearance of the jejunoileal resection specimen containing multiple polypoid lesions.

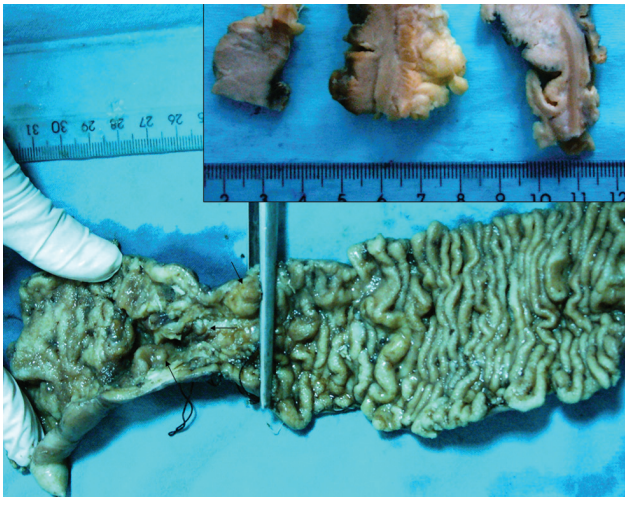

Figure 2. Gross appearance of segmental multiple polypoid lesions with patchy ulcerations and varying from 0.5-2.5 cm in diameter.

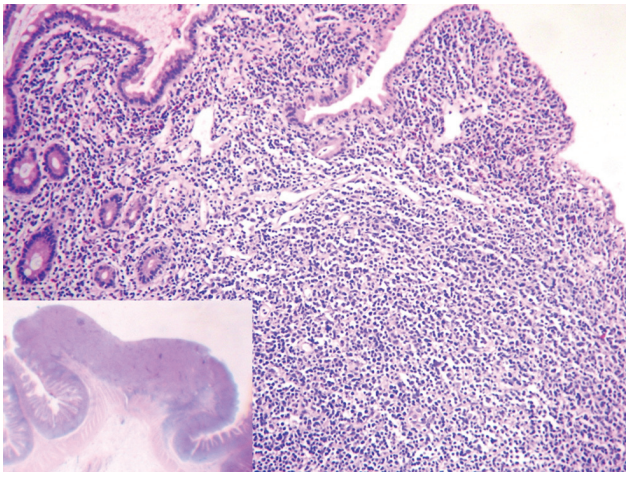

Figure 3. Neoplastic infiltration resembling polypoid appearance (H&E, x100).

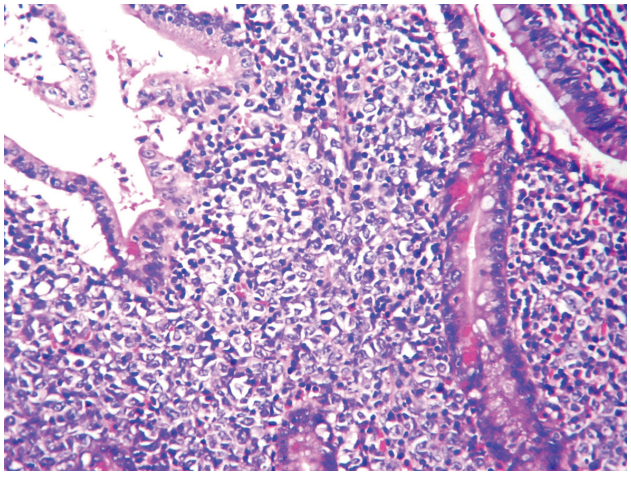

Figure 4. Atypical lymphocytes infiltrating the intestinal tissue (H&E, x200).

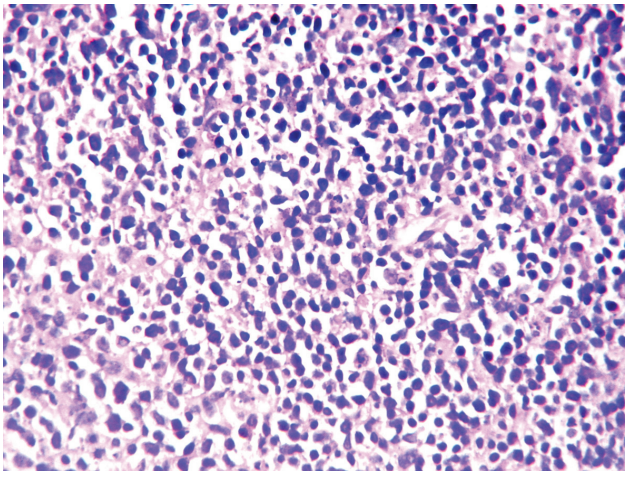

Figure 5. Atypical lymphocytes with hyperchromatic nuclei and eosinophilic cytoplasm (H&E, x400).

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.

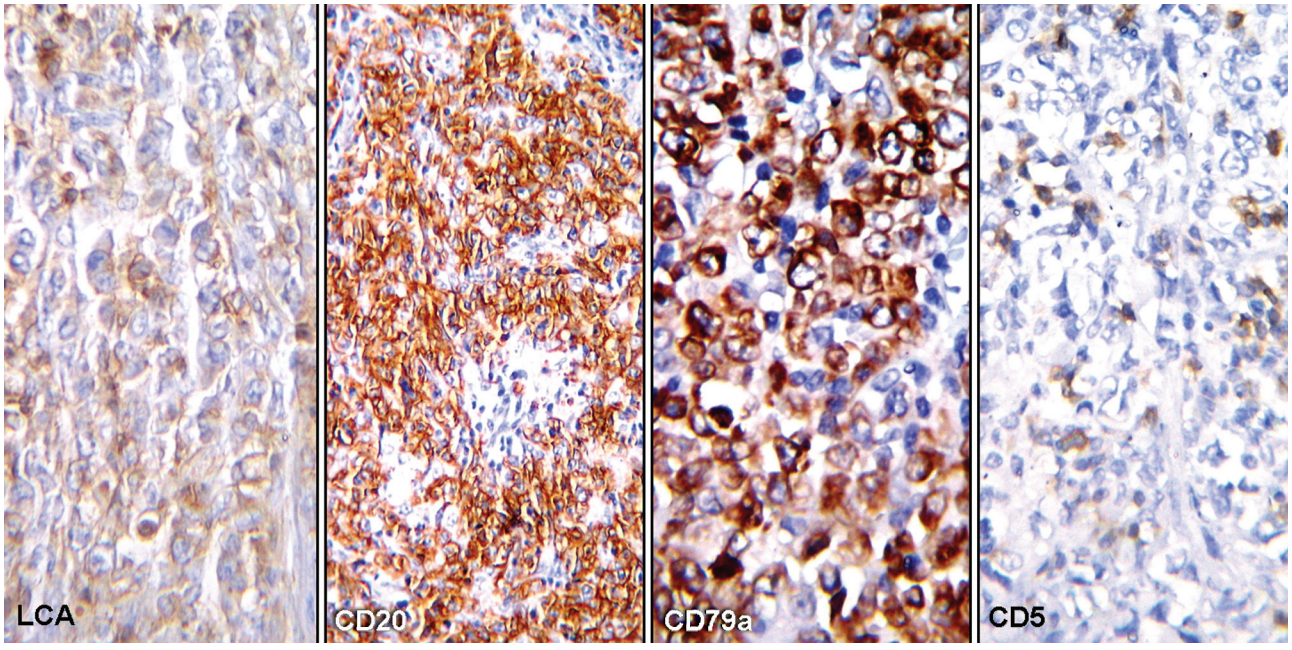

Figure 6. B-cell phenotype with LCA, CD20, CD79a expression, and without CD5 expression (B-SA peroxidase, DAB, LCA, x400; CD20, x200; CD79a, x400; CD5, x400).

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 mast be ruled out in the differential diagnosis of patients presenting with the signs and symptoms of Crohn's disease.

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