The incidence of primary colorectal lymphoma is very rare, accounting for 0.2%–0.6% of colorectal malignancies.\(^1\) Primary colorectal lymphomas manifest in a variety of ways, ranging from solitary fungating masses to multiple polyps. In 1968, Friedman et al.\(^2\) reported four cases of “lymphomatous colitis,” a rare form of primary colorectal lymphoma mimicking ulcerative colitis (UC). Since then, a small number of patients with diffuse-type colorectal lymphoma have been reported, in whom a clinical and/or histological diagnosis of colitis, including inflammatory bowel disease (IBD), was made initially but were subsequently discovered to have diffuse lymphoma involvement of the colon within a short period of time.\(^3\) IBD and immunosuppression have been reported as risk factors for primary colorectal lymphoma.\(^4\) Most IBD-related lymphomas develop late in the course of an extensive longstanding disease. In that respect, these cases differ from most reports of primary colorectal lymphoma as a complication of longstanding IBD. We experienced a case of UC-like primary colorectal lymphoma in a 31-year-old woman who presented with profuse hematochezia, was misdiagnosed with UC, and died of a diffuse lymphoma involving the entire colon 12 months after hematochezia first developed.

CASE REPORT

A 31-year-old woman with no prior history of IBD presented with intermittent abdominal pain and mucoid diarrhea. Five months later, she developed hematochezia and was diagnosed with UC on the basis of endoscopic biopsy. She was treated with oral Pentasa and Pentasa enema. However, she continued to be symptomatic with marked weight loss (20 kg in 4 months). Follow-up colonoscopy revealed continuous mucosal changes from the rectum to the cecum, such as diffuse hyperemia, edematous changes, friability, loss of normal vascularity, and multiple variable-sized ulcers, consistent with UC (Fig. 1A). No mass-forming lesion was present throughout. Abdominal and pelvic computed tomography revealed diffuse edematous wall thickening in the whole colon and multiple enlargements of mesenteric lymph nodes. No hepatosplenomegaly was observed. Multiple biopsies were taken from the terminal ileum, cecum, transverse colon, sigmoid colon, and rectum. All biopsy specimens revealed diffuse infiltration of large, atypical lymphoid cells with a high nuclearto-cytoplasmic ratio, occasional prominent nucleoli, and mitotic figures (Fig. 1B, C). They expanded the lamina propria, pushed the crypts apart, and invaded the submucosa. By immunochem-
The tumor cells were diffusely positive for CD20, bcl2, and p53, and negative for CD3, CD5, CD10, and cyclin D1. The Ki-67 labeling index was about 60%. The overall histology and immunophenotype supported a diagnosis of diffuse large B-cell lymphoma. There was no detectable evidence of extraintestinal lymphadenopathy or lymphomatous involvement elsewhere. Bone marrow evaluation was also negative for lymphoma. As no extraintestinal disease was found, we considered it as a primary colorectal lymphoma. The patient died while receiving the second cycle of R-CHOP chemotherapy.

We retrospectively reviewed the colonoscopic biopsy specimens that were initially diagnosed with UC. At low magnification view, the background mucosa displayed crypt architectural distortion and dense inflammatory cell infiltration, resembling chronic colitis. Atypical lymphoid cells were aggregated in the bottom right corner (below the yellow dotted line), which show not only strong expression on CD20 immunostaining (E) but also high Ki-67 labeling index (F).
colitis. However, we found some lymphoma cells, which were admixed with other inflammatory cells or showed focal aggregation in the basal portion of the mucosa (Fig. 1D). Those lymphoma cells were more distinguishable on immunostaining for CD20 and Ki-67 (Fig. 1E, F).

**Ethics statement**

This study was approved by the Institutional Review Board of Inje University Ilsan Paik Hospital with a waiver of informed consent (IRB No. ISPAIK 2019-05-005) and performed in accordance with the principles of the Declaration of Helsinki.

**DISCUSSION**

Twenty cases (including the present one) of "colitis-like" diffuse-type colorectal lymphomas were reported in the English literature.\(^2\)\(^{17}\) We excluded cases of colorectal lymphoma developed in patients with a longstanding IBD or prior established diagnosis of extracolonic lymphoma. The clinical and pathological findings of these patients are summarized in Table 1. There were 10 men, 8 women, and two children. Their mean age at presentation was 53.2 years (range, 6 to 82 years). The main symptoms were diarrhea/hematochezia (90%), loss of weight (50%), abdominal pain (25%), and fever (10%). Endoscopic findings were consistent with UC (55%), Crohn disease (20%), diffuse colitis (20%), and multiple ulcers (5%); no localized mass-like lesion was present. All cases were non-Hodgkin's lymphoma: eight were B-cell type (four mantle cell lymphomas, two mucosa-associated lymphoid tissue lymphomas, one diffuse large B-cell lymphoma, and one follicular lymphoma), seven were T-cell type, one was natural killer cell lymphoma, and four other unclassified lymphoreticular malignancies (one malignant lymphoma, one reticulum cell sarcoma, one lymphosarcoma, and one lymphocytic lymphoma). Of note, there were nine patients in whom colitis was histologically confirmed with multiple biopsy or colectomy but were subsequently found to have lymphoma involvement in a retrospective review of previous slides. Taken together, diffuse type primary colorectal lymphoma is a very rare disease and is easily misdiagnosed, particularly in reliance on endoscopic biopsy examination alone. The possibility of a hidden lymphomatous involvement would have to be considered in patients with medically refractory and rapidly progressive colitis.

**Table 1. Clinical and pathological findings of previously published cases of “colitis-like” diffuse-type colorectal lymphomas in the English literature**

| Year | Author | Sex/Age (yr) | Pathologic diagnosis | Endoscopic findings | Revision of initial diagnosis | Superficial LAP at presentation | Extracolonic involvement |
|------|--------|-------------|---------------------|-------------------|-----------------------------|-------------------------------|------------------------|
| 1968 | Friedman et al.\(^2\) | M/37 | Reticulum cell sarcoma | UC | No | No | Liver |
| 1968 | Friedman et al.\(^2\) | F/54 | Malignant lymphoma | UC | Yes (colectomy) | No | No |
| 1968 | Friedman et al.\(^2\) | M/73 | Lymphosarcoma | UC | Yes (Bx) | No | No |
| 1980 | Weir et al.\(^2\) | F/67 | Lymphocytic lymphoma | CD | Yes (Bx) | Yes, generalized | BM |
| 1992 | McCullough et al.\(^4\) | M/44 | Mantle cell lymphoma | UC | Yes (Bx) | Yes, cervical axillary | Pancreas |
| 1995 | Lenzen et al.\(^5\) | F/53 | MALT lymphoma | UC | No | No | Upper GI tract, BM |
| 1996 | Robert et al.\(^6\) | F/71 | Mantle cell lymphoma | UC | Yes (colectomy) | No | No |
| 1996 | Hirakawa et al.\(^7\) | M/47 | T-cell lymphoma | UC | No | No | Upper GI tract |
| 1997 | Son et al.\(^8\) | F/40 | Peripheral T-cell lymphoma | CD | Yes (Bx) | No | No |
| 2003 | Isomoto et al.\(^4\) | M/47 | Adult T-cell leukemia/lymphoma | UC | No | Yes, generalized | Stomach, skin |
| 2004 | Payne et al.\(^10\) | F/76 | High-grade T-cell lymphoma | Colitis | No | Yes, generalized | No |
| 2004 | Tamura et al.\(^11\) | M/61 | Mantle cell lymphoma | Colitis | No | No | No |
| 2008 | Berkelhammer et al.\(^12\) | F/82 | MALT lymphoma | CD | Yes (Bx) | No | No |
| 2014 | Koksal et al.\(^13\) | M/73 | Mantle cell lymphoma | UC | No | No | Stomach |
| 2015 | Zaheen et al.\(^14\) | M/74 | EBV-negative NK cell lymphoma | Colitis | No | No | BM, pleural effusion |
| 2015 | Wu et al.\(^15\) | M/56 | T-cell lymphoma | Ulcers | Yes (Bx) | Yes, generalized | No |
| 2016 | Cheung et al.\(^16\) | NA/12 | EBV-positive T-cell lymphoma | Colitis | No | No | Hepatosplenomegaly |
| 2016 | Cheung et al.\(^16\) | NA/6 | EBV-positive T-cell lymphoma | CD | No | No | No |
| 2017 | Zenda et al.\(^17\) | M/59 | Follicular lymphoma | UC | No | Yes, cervical inguinal | Spleen, BM |
| Present case | F/32 | Diffuse large B-cell lymphoma | UC | Yes (Bx) | No | No | |

LAP, lymphadenopathy; M, male; UC, ulcerative colitis; F, female; Bx, biopsy; CD, Crohn's disease; BM, bone marrow; MALT, mucosa-associated lymphoid tissue; GI, gastrointestinal; EBV, Epstein-Barr virus; NA, not available.
applied: (1) no palpable superficial lymphadenopathy at initial presentation, (2) a normal chest X-ray with no mediastinal lymphadenopathy, (3) no evidence of leukemia, (4) a predominant mass in the bowel with only local lymphadenopathy, and (5) no hepatosplenomegaly. When the diagnostic criteria of primary colonic lymphoma are strictly applied, only seven of 20 cases of “colitis-like” colorectal lymphoma are eligible for diffuse-type primary colorectal lymphoma: 13 cases were excluded due to synchronous other organ involvement at initial presentation, such as superficial or generalized lymphadenopathy (six cases), upper GI involvement (five cases), bone marrow involvement (four cases), or hepatosplenomegaly (three cases), etc. Consequently, it is a reflection that “colitis-like” diffuse involvement of colorectal lymphoma seems to be more often secondary. Accurate discrimination between primary and secondary colorectal lymphoma is important for proper staging and management. In conclusion, since primary colorectal lymphoma can rarely manifest as “colitis-like” diffuse colonic involvement, awareness of this rare presentation is important to ensure proper diagnosis and treatment.

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**Conflicts of Interest**

The authors declare that they have no potential conflicts of interest.

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