Mucosa-Associated Lymphoid Tissue (MALT) Lymphoma of the Rectum: Report of Two Cases

Yasuhisa YAMAMOTO, Hiroyuki IMAI, Sueharu IWAMOTO, Yoshiyuki TADAOKA, Kazuhiro YOSHIDA and Tsukasa TSUNODA

Division of Gastroenterological Surgery, Department of Surgery, Kawasaki Medical School, Kurashiki 701-01, Japan

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ABSTRACT. We have treated two patients with low-grade B-cell lymphoma of mucosa-associated lymphoid tissue (MALT) arising in the rectum. A 70-year-old female with anal bleeding underwent local excision of a rectal polypoid mass (3.0 × 2.3 × 1.3 cm) transanally to preserve the sphincter muscle. Pathological findings showing partial invasion to the muscularis propria were indicative for prophylactic combination-chemotherapy (1 × CHOP). A 72-year-old male in whom positive occult blood was detected underwent a low anterior resection for three aphthoid ulcers (1.0-1.5 cm in diameter, submucosal invasion). Radically resected tumors in early stages should require no further treatment. However, cases with tumors in advanced stages should have elective postoperative therapy, such as chemotherapy and/or irradiation. Since recurrences (15%) of initially low-grade MALT lymphoma have been reported, these two cases should undergo periodic examination of the gastrointestinal tract at regular intervals over five years.

Key words: malignant lymphoma — updated Kiel classification — colorectal mucosa-associated lymphoid tissue (MALT) lymphoma

In 1983, Isaacson *et al*[1] reported the first two cases of so-called mucosa-associated lymphoid tissue (MALT) lymphoma of the stomach and small intestine. The characteristics of MALT lymphoma are as follows: 1) Other common sites for the occurrence are the salivary glands, lung and thyroid. 2) The great majority of tumors are of B-cell origin. 3) These lymphomas tend to remain localized for prolonged periods, and 4) the B-cell group responds favorably to local treatment.[2]

Primary malignant lymphomas of the large intestine are rare neoplasms accounting for less than 1% of colorectal malignancies. Twenty-nine cases with colorectal tumors showing cytological polymorphism and composed of centrocyte-like, plasmacytoid and/or immunoblast-like cells were reported by Shepherd *et al*[3] in 1988 as the first MALT lymphoma of the large intestine. In this paper, we describe two patients with a MALT lymphoma arising in the rectum and presenting as a polyp and an aphthoid ulcer, respectively.

山本康久，今井博之，岩本末治，忠岡好之，吉田和弘，角田　司

CASE REPORTS

Case 1

A 70-year-old female presented at our hospital in December 1994 complaining of anal bleeding. Physical and endoscopical examination disclosed a quite movable polypoid lesion (3.0 × 2.3 × 1.3 cm) on the posterior wall of the rectum, situated 6 cm from the anal verge. A colonoscopic biopsy led us to respect a malignant lymphoma. Since further examinations (conventional radiography, endoretal ultrasonography, abdominal ultrasonography, pelvic computed tomography) showed no abnormal findings, she underwent local excision of the rectal mass transanally with 0.5-1.0 cm of surgical margin to preserve anal sphincter function (Fig 1). Pathological

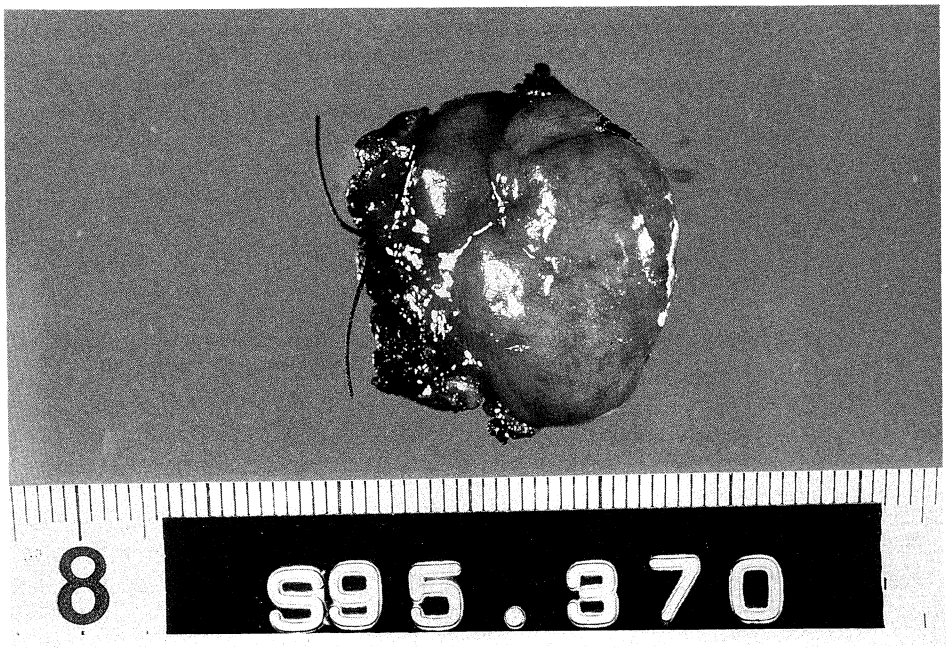

Fig 1. Locally excised specimen of Case 1 shows a polypoid lesion.

findings revealed a low-grade MALT lymphoma of B-cell origin which had partially invaded the muscularis propria (Fig 2). Immunohistochemistry was performed for Case 1 with some antibodies. The tumor cells were positive for Leu 12 (CD 19)+, OKT 8−, Leu 3a−, Leu 4− and Leu 7−. Prophylactic combination-chemotherapy (1 × CHOP=cyclophosphamide, doxorubicin hydrochloride, oncovin and prednisolone) was performed before discharge, and the patient recovered without evidence of recurrence as of Novermber 1995.

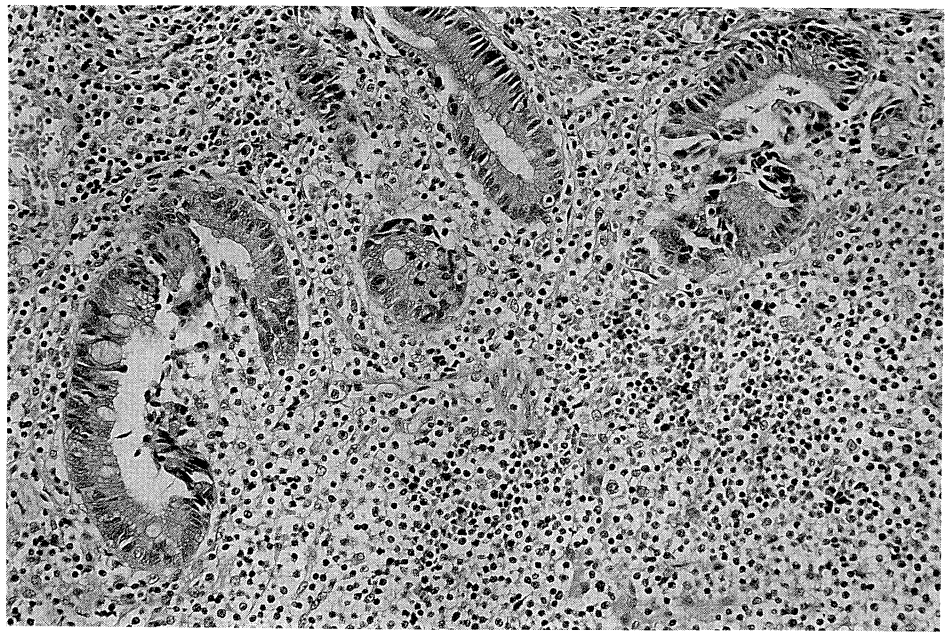

Fig 2. Histological findings reveal infiltration of centrocyte-like cells and plasma cells forming lymph follicles (Case 1. H·E, ×40).

Case 2

A 72-year-old male complained of a four-month history of diarrhea, and positive occult blood was detected from June 1994. Endoscopical examination showed three aphthoid ulcers (1.0-1.5 cm in diameter) on the rectum, situated 10-15 cm from the anal verge. Endorectal ultrasonography and endoscopic mucosal resection (EMR) suggested a MALT lymphoma invading to the mucosa or submucosa. Other investigations (barium-enema studies, abdominal ultrasonography, pelvic computed tomography) disclosed no abnormal findings. Since a complete EMR was thought to be imposssible, he underwent a low anterior resection under intraoperative colonoscopy (Fig 3). Pathological findings of the surgical specimen revealed a low-grade MALT lymphoma of B-cell origin with submucosal invasion and no lymph node involvement (Fig 4). Immmunohistochemical reactivity of Case 2 showed the tumors to be positive for LC+, L 26+, LN 2+, LN 3+, LN 1− and Leu M1−. No prophylactic chemotherapy and/or irradiation were applied, and the patient recovered uneventfully without evidence of tumor recurrence as of November 1995 (Table 1).

Fig 3. Resected specimen of Case 2 shows an aphthoid lesion (black arrow) with two scars after EMR (white arrow).

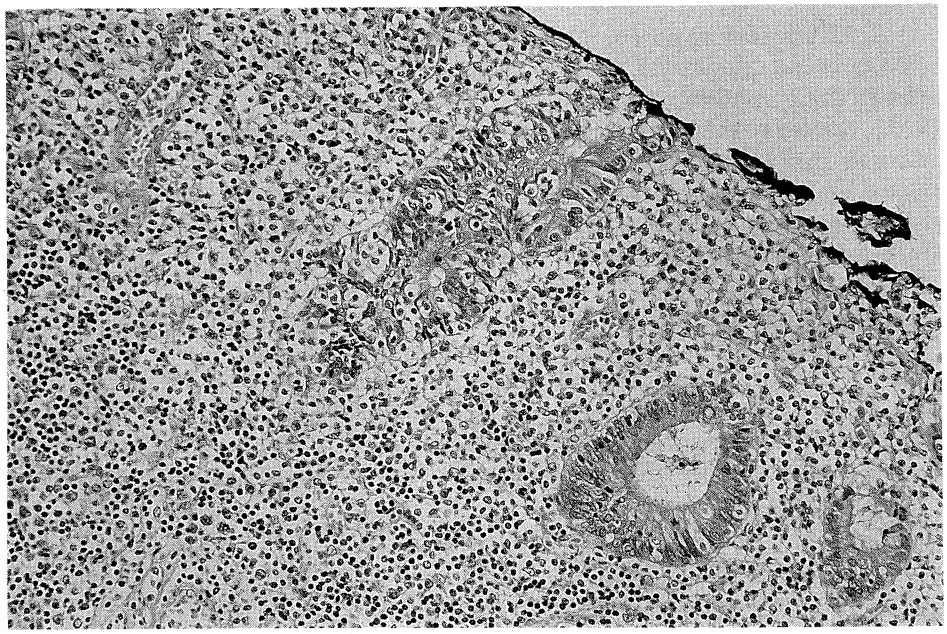

Fig 4. Centrocyte-like cells infiltrate to submucosa forming lymph follicular colonization (Case 2. H•E, x40).

TABLE 1. Clinicopathological features in two case with rectal MALT lymphoma

Cases	Age	Sex	Presenting symptoms	Number of lesion	Size (cm)	Macroscopic type	Histologic type[4]	Depth of invasion	Outcome
1	70	F	anal bleeding	1	3.0x2.3x1.3	polypoid	centrocyte-like	muscularis propria	alive
2	72	M	positive occult blood	3	1.5x1.0 1.0x1.0 1.0x0.9	aphthoid	centrocyte-like	submucosa	alive

DISCUSSION

As some recent reports[5,6] define clearly the term and histological criteria of MALT lymphoma (showing cytological polymorphism and composed of centrocyte-like, plasmacytoid and/or immunoblast-like cells) arising in the large intestine, the problems of initial treatment and following control of this neoplasm are more important than those of diagnosis. Primary colorectal MALT lymphomas have been very rare until recently[7,8] and deciding how to control them may be controversial. As far as we know, there are no macroscopic characteristics of MALT lymphoma.

Local excision and low anterior resection were respectively indicated for our two cases. Case 1, who was treated with local excision, underwent postoperative chemotherapy (1 × CHOP) because the tumor had invaded partially to the muscularis propria. Radaszkiewicz *et al*[6] have recommended that individually adapted resection of lymphomas should be as radical as necessary. Completely resected tumors in early stages (mucosa or submucosa) should require no further treatment. However, cases in advanced stages (invasion through the muscularis propria) should undergo elective postoperative therapy, such as chemotherapy and/or irradiation.

We did not examine the proto-oncogenes of the two patients, however, enhanced bcl-2 expression is important in distinguishing between follicular lymphoma and MALT lymphoma.[9,10]

Although the etiological factors of our two cases are unknown, there are three patterns of recurrence; 1) local recurrence around the site of the removed tumor, 2) gastrointestinal (GI) tract dissemination distant from the primary lesion including Waldeyer's ring and parotis, and 3) generalization involving the bone marrow, central nervous system, lymph nodes or organs unrelated to the GI tract occurred. The characteristics of recurrence after surgery for intestinal MALT lymphoma have been reported to be as follows; 2 of 13 cases (15.3%) with initially low-grade malignancy relapsed and 8 of 50 cases (16.0%) with high-grade malignancy recurred.[6]

Even if the pathological diagnosis reveals low-grade malignancy of MALT lymphoma, periodic examination of the GI tract including radiography, endoscopy, ultrasonography and computed tomography of the abdomen should be performed at regular intervals over five years after initial surgery.

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