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Case Report

MALT Lymphoma of the Rectum: Report of a Case Treated with Chemotherapy

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ABSTRACT

The gastrointestinal tract is the most frequently involved extranodal location for MALT* lymphomas, but MALT lymphomas of the large intestine are rarely observed. A treatment for colorectal MALT lymphoma has not yet been established. In colonic MALT lymphomas, the literature suggests that surgical resection of localized lesion may be the best choice. In the present case, combination of multi-agent chemotherapy and radiotherapy was effective, though a long-term follow-up is definitively needed. In this report, a 56 year-old man with MALT lymphoma manifesting in colonoscopy as multiple mucosal discolorations and some localized granularity of the rectum mucosa is presented.

Keywords: MALT Lymphomas, Gastrointestinal Tract, Rectum

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INTRODUCTION

The term mucosa-associated lymphoid tissue (MALT) lymphoma was first introduced by Isaacson and Wright in 1983.(1), The most frequently involved extranodal location for MALT lymphomas is gastrointestinal (GI) tract and the stomach is the most common GI site. MALT lymphomas of the large intestine are very rarely observed. (2, 3), MALT-type lymphomas of the gastrointestinal tract are low-grade lymphomas derived from this specialized lymphoid tissue. (1, 4), A treatment for colonic MALT lymphoma has not yet been established. Here, we report a case of MALT lymphoma manifesting in colonoscopy as multiple mucosal discolorations and some localized granularity of the rectal mucosa in a 56 year-old man. Complete remission was achieved with combination chemotherapy.

* Mucosa Associated Lymphoid Tissue

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CASE REPORT

A 56 year-old man from Saveh-Iran was referred for investigation of five months history of generalized abdominal pain, fresh rectal bleeding and constipation. Since five months prior to the referral time, his pain was sustained with no radiation to any other area, and was neither positional nor related to feeding and activity. He had about 8kg weight loss, fever, night sweating and decreased appetite before admission to the hospital. During the prior years, he had no history of diabetes mellitus, hypertension, ischemic heart disease, tuberculosis, hyperlipidemia and malignancy. The patient had a history of occasional dyspepsia and was opium addicted and cigarette smoker (20 pack-year), but no drug

history was present. There were no risk factors for HIV infection and there was no preceding history of inflammatory bowel disease. At the time of presentation, his general health was good and appeared normal without any remarkable findings except mild to moderate splenomegaly and foot finger clubbing on physical examination. No lymphadenopathy was detected. Rectal examination was normal. There was no perianal disease. Laboratory findings on presentation are shown in table 1.

Table 1: Laboratory data

WBC (C/µL)	10100	AST (IU/L)	5 (up to 38)
Hgb (g/dL)	14	ALT (IU/L)	12 (up to 38)
Platelet (C/µL)	250/000	ALP (IU/L)	175 (up to 306)
FBS (mg/dL)	111	Total Bilirubin (mg/dl)	0.5 (0.1-1.2)
Cr (mg/dL)	0.9	Direct Bilirubin (mg/dl)	0.1 (0-0.3)
PT(Sec)	13	LDH (U/L)	138 (up to 400)
		ESR (mm/1h)	26

The patient underwent colonoscopy which resulted in rectal bleeding; rectum had diffuse erythematous and friable appearance without spontaneous bleeding and pseudopolyps in rectum (Figure 1). Other parts of colon, from sigmoid colon to cecum, were normal. In view of his dyspepsia, the upper gastrointestinal endoscopic examination revealed a diffuse patchy erythema in antrum and prepyloric areas. The mucosa had a patchy atrophic appearance in body and fundus of the stomach. A rapid urease test performed on gastric biopsy (incisura) was positive. CT scans of the abdomen and pelvis confirmed the presence of splenomegaly and retroperitoneal and mesenteric lymphadenopathies. The rectum had thick tumoral wall. All findings were in favor of lymphoma.

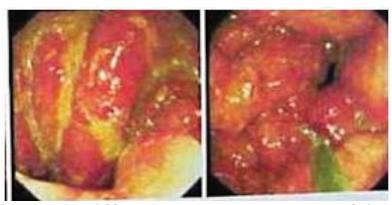

Figure 1.Diffuse erythematous and friable appearance without bleeding and pseudopolyps in rectum.

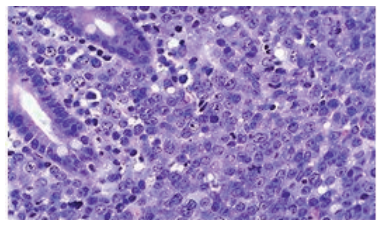

Figure 2.Fragments of colonic mucosa infiltrated by small lymphoid cells with regular nuclei destructing glands.

In histology report, microscopic examination of the rectal mucosal lesions showed fragments of colonic mucosa infiltrated by small lymphoid cells with regular nuclei destructing glands, showing invasion to crypt epithelium in some areas (Figure 2). Immunohistochemistry showed that the neoplastic cells were positive for CD20 and Bcl-2 but negative for CD5, CD3, and CD23.

Finally, management and follow-up of patient led to the diagnosis of low grade marginal B-cell lymphoma of MALT type presenting as diffuse multiple lymphomatous polyposis. The patient's good general health and the diffuse nature of the disease led to chemotherapy (CHOP: cyclophosphamide, doxorubicin, vincristine and prednisolone) after referring to oncology clinic. His presenting symptoms, including rectal bleeding and abdominal pain, anorexia and night sweating subsided after treatment. Three months after treatment, a follow-up colonoscopy revealed no abnormal mucosa.

DISCUSSION

Primary lymphomas of the gastrointestinal tract account for 30% of extranodal lymphomas.(5) The stomach and small bowel are the commonest sites to be affected. Approximately 10–20% of the lesions involve the colon.(6-8) Lymphomas of the colon constitute less than 0.4% of colonic neoplasms and lymphoma of the rectum is very rare.(9, 10), Invariably, these tumors are of the non-Hodgkin's type and are

staged using a modified Ann Arbor system.(11)

The tumor may present as polypoid or ulcerating multiple small polyps affecting any region from the stomach to the rectum, often in a discontinuous pattern with normal intervening intestine. Larger ulcerated lesions also occur. In all cases, small lymphoid cells of B-cell lineage were found to be infiltrating the deep mucosa and submucosa. Bcl-2 overexpression was seen in this case, as it has been previously reported in the gastric MALT-type lymphomas.(12) Bcl-2 gene, which encodes a constitutively expressed protein, is found in different types of neoplasms including MALT-type lymphomas. (13), Antigenic stimulation is vital to the growth of MALT type lymphomas. A causative relationship exists between H. pylori infection and gastric MALT lymphomas. (14, 15), When disease is at an early stage, successful eradication of the bacterium results in apparent cure in a significant number of cases.(16), We were unable to detect H. pylori after careful histological examination of multiple biopsies and a rapid urease test. There is an association between colonic lymphoma and ulcerative colitis. (17), Colonic lymphoma and adenocarcinoma can also coexist.(18), Our patient did not have a history of either of them. Colonic lymphomas carry a worse prognosis than adenocarcinomas. (12), In general, those patients who undergo resection with or without adjuvant chemotherapy or radiotherapy have the best prognosis.(11), A variety of combination chemotherapy regimens have been used, including agents such as cyclophosphamide, chlorambucil, vincristine, and prednisone. Given the rarity of the condition, little consensus exists on the optimal regimen. Three-year survival remains at approximately 50%.(19), Our patient had shown a gratifying short term response to chemotherapy, with no significant side effects. Prognosis, however, must remain guarded. In colonic MALT lymphomas, the literature suggests surgical resection of localized lesion as the best choice.(20), In the present case, combination of multi-agent chemotherapy and radiotherapy was effective, though a long-term follow-up is definitively mandatory. As mentioned above, we think that mucosal biopsy of the discolored but not protruding and/or ulcerative mucosa is necessary to rule out the lymphoma.

CONCLUSION

MALT lymphoma develops in diverse anatomic locations such as stomach, salivary gland, thyroid, lung, and breast;(21), however, colorectal involvement is extremely rare.(22, 23) Colonic MALT lymphoma has not been well investigated as compared to stomach MALT lymphoma. The metastatic ability and sensitivity against chemotherapy of the colonic MALT lymphoma is not known. Because colonic MALT lymphoma presents with various features, meticulous observation is mandatory in colonoscopic evaluations. In general, those patients who undergo resection with or without adjuvant chemotherapy or radiotherapy have the best prognosis.

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