Rectal Lymphoma: Report of a Rare Case and Review of Literature

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Abstract- Colorectal lymphoma is an extremely rare disease, representing less than 0.5% of all primary colorectal neoplasms. The disease is usually diagnosed in the advanced stages because of its primary non-specific symptoms. The most common involved site is cecum followed by rectum and ascending colon. Diffuse large B-cell lymphoma is a more frequent subtype. Although surgical resection is often technically feasible, optimal therapy for a colorectal lymphoma, especially rectal lymphoma, has not yet been identified. The authors describe a patient with the primary rectal lymphoma, high-grade features and complete response to chemotherapy.

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Introduction

Lymphoma occurs as a primary lesion or as part of a generalized malignant process involving the gastrointestinal tract. Differentiation of these two processes is a very important issue, because their treatment and prognosis are different. Primary gastrointestinal lymphomas are defined as those in which the alimentary tract involvement predominates or those with symptoms of gastrointestinal involvement on presentation. More strict guidelines include absence of palpable peripheral lymphadenopathy at time of presentation; absence of mediastinal adenopathy on a chest radiograph; a normal peripheral blood smear; involvement of esophagus found during laparotomy, stomach, bowel or regional lymph nodes (excluding retroperitoneal lymph nodes); and absence of hepatic and splenic involvement except by direct spread of the disease from a contagious focus (1).

Primary malignant lymphoma of the colon is uncommon and accounts for only 0.2-0.4% of all colonic malignancies and 10-15% of all primary lymphomas of the gastrointestinal tract, which themselves account for about 30% of extra nodal lymphomas (2-6).

The most frequent colonic location is the cecum (70%), followed by the rectum and ascending colon (7).

Treatment of colorectal lymphomas remains uncertain. While surgical treatment may be indicated for some localized tumors, many authors consider medical management to be the primary treatment (8). Some studies reported that the primary treatment is to attempt resection when the disease is judged to be resectable because of poor prognosis in patients with residual disease (12-14). Most of knowledge about the rectal lymphoma and its management is harvested from case reports because of the paucity of disease. Therefore, treatment modality of primary lymphoma of the rectum remains uncertain. The authors describe a patient with the primary rectal lymphoma managed medically with excellent result.

Case Report

The patient was a 75-year-old man who had developed rectal bleeding, pain and alteration in bowel habits since 5 years ago. He was known as a case of ulcerative colitis receiving Asacol. The patient had relative remission. His symptoms have gotten worse since about 6 months ago. There were an irregular fungating and the circumferential mass from the dentate line toward upper part on rectal examination. The mass was firm to hard, and fixed to the rectal wall.

Abdominopelvic CT scan showed extensive, bulky rectal tumor (Figure 1). A colonoscopy showed a 10 cm length tumor in the rectum from two cm of the anal verge (Figure 2). A hypoechoic lesion was reported at 2-

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19 cm from the anal verge in the patient's endosonogeraphy. The lesion invaded beyond muscularis properia. Regional Lymph nodes and iliac nodes were present but major pelvic organs were spare of tumor. At upper anal canal, internal anal sphincter was invaded by the tumor. Biopsy specimens were taken for histological diagnosis. The lesion was diagnosed as large B-cell lymphoma (Figure 3). Results of immunohistochemical staining were CD 45: Positive, CD 20: Positive, CD 3: Negative, Cytokeratin: Negative. The patient was referred to an oncologist for chemotherapy (eight sessions) and radiotherapy (28). The patient had no clinical symptoms, signs and lesion in colonoscopy after one year.

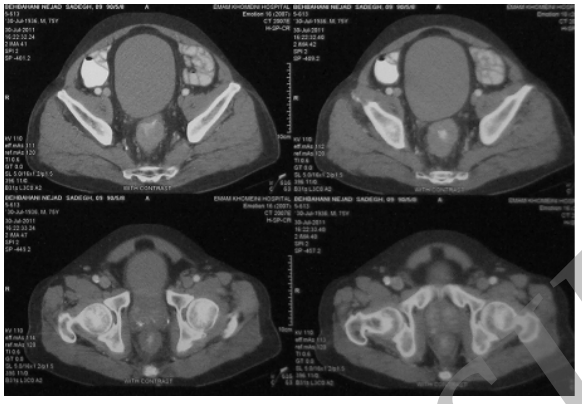

Figure1. Bulky rectal tumor in Spiral Pelvic CT-Scan

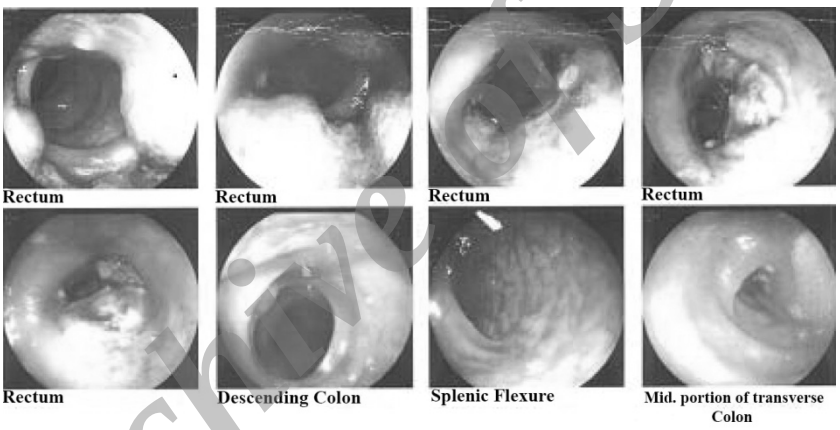

Figure 2. Colonoscopy demonstrating large fungating rectal tumor

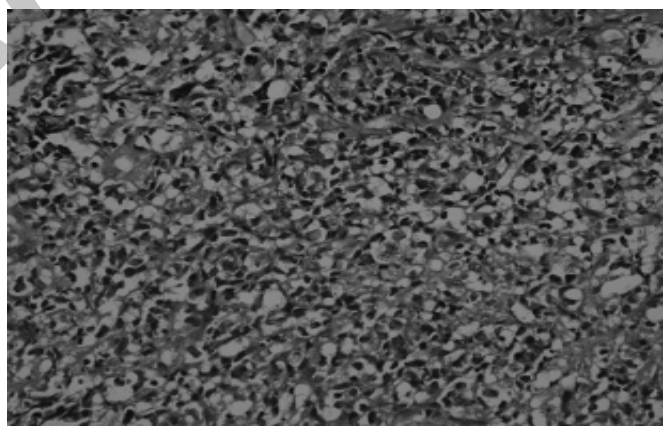

Figure 3. Pathologic examination reveals Large B-Cell Lymphoma

Discussion

Lymphoma involving the colon and rectum is rare. The cecum is most often involved, probably as a result of spread from the terminal ileum (9).

Rectal lymphoma usually presents with signs and symptoms suggestive of primary rectal carcinoma. Patients with a rectal lymphoma usually seek treatment because of rectal bleeding or an alteration in bowel habits. It is reported that the majority of patients are symptomatic complaining of abdominal pain, nausea, vomiting, fever, and weight loss (3-6).

The gross appearance may be a circumferential or polypoid mass, ulceration, or a diffuse infiltration with stricturing and bowel wall thickening. 86 per cent of the lesions are solitary, but they can be multiple and diffuse in nature. The intestinal lymphomas may be classified into B-cell lymphomas (85%) and T cell lymphomas (15%). Among the B cell lymphomas, mantle cell lymphoma has a worse prognosis, whereas mucosa-associated lymphoid tissue (MALT) lymphomas have a better prognosis than other B-cell tumors (7).

Abdominal CT scan and endoscopy with biopsy are the most useful diagnostic tests. When CT scan revealed a combination of a focally or diffusely infiltrative process of the colon and extensive abdominal and/or pelvic adenopathy, lymphoma should be the primary consideration in the differential diagnosis and must be excluded by endoscopic biopsy. However, if adenopathy was not associated with a primary colorectal lymphoma, it might be difficult to distinguish this lesion from a primary adenocarcinoma of the colon by radiologic methods. This difficulty arises predominantly in the settings of solitary mass lesions. Primary colorectal lymphomas manifested as discrete masses tend to have a greater depth of mural invasion than infiltrative lesions (9).

Two risk factors are associated with the development of the primary colorectal lymphoma: inflammatory bowel disease and immunosuppression (post transplant, AIDS, or immune disorder). The aggressive nature of AIDS-related lymphoma could usually result in a disseminated disease at the time of diagnosis (10,11).

Diffuse large B-cell lymphoma of the large bowel is generally treated with an uniform therapeutic approach: aggressive surgery followed by adjuvant chemotherapy. However, most of the studies included patients with primary gastric or small bowel and large bowel in the same analysis, and also patients at different stages, different histology, and different surgical approaches. Lymphoma of the rectum should be considered as a different clinicopathological entity with different behaviors, histology and clinical presentation. So, treatment should be defined based on this special clinical condition (13). The therapeutic experience in this selected group of patients is limited, because the number of patients is few even in large tertiary referral centers. The analysis of those retrospective data regarding surgery, radiation therapy, and chemotherapy also is difficult to interpret because of lack of uniformly accepted regimens.

Lymphoma of the rectum is a very rare entity, and there is still controversy about its treatment. Some authors still believe that medical management should be considered as the primary therapy even in surgically operable localized tumors. This case report shows some promises in treating the colorectal lymphoma by non-surgical management.

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