Turk J Gastroenterol 2001; 12 (4): 311-313

Primary lymphoma of rectum with a fistula to ileum

İleuma fistülize olan primer rektum lenfoması

Zeki KARASU[1], Nevra ELMAS[2], Ahmet MUSOĞLU[1]

Ege University School of Medicine, Departments of Gastroenterology[1] and Radiology[2], İzmir

Primary lymphoma of the rectum is uncommon and if the criteria described by Dawson et al. are used for diagnosis, it is a rare tumor. Barium enema, rectosigmoidoscopy, ultrasonography, computed tomography and magnetic resonance imaging have been used in the diagnosis and staging of rectal lymphomas. Since the introduction of per rectal paramagnetic contrast agents, magnetic resonance imaging appears to have superiority over other diagnostic tools. We report a case of primary rectal lymphoma with a fistula to the ileum, which was diagnosed by magnetic resonance imaging using a per rectal paramagnetic contrast agent.

Key words: Rectum, lymphoma, magnetic resonance imaging.

Rektumun primer lenfomaları seyrek görülür. Eğer tanı için Dawson ve arkadaşları tarafından tanımlanan kriterler kullanılırsa primer rektal lenfomaların çok seyrek görülen tümörler olduğu ortaya çıkacaktır. Rektal lenfomaların tanısında ve evrelemesinde baryumlu kolon grafisi, rektosigmoidoskopi, ultrasonografi, bilgisayarlı tomografi ve manyetik rezonans gibi görüntüleme yöntemleri kullanılmaktadır. Rektal yoldan uygulanan paramanyetik kontras ajanların kullanılmaya başlamasından sonra, manyetik rezonans görüntüleme yönteminin diğer yöntemlere üstünlük sağlayacağı düşünülmektedir. Rektal yoldan verilen kontrast ajan kullanılarak, manyetik renzonans görüntüleme yöntemi ile tanısı konmuş olan ileuma fistül yapmış bir primer rektal lenfoma olgusu burada sunulmaktadır.

Anahtar kelimeler: Rektum, lenfoma, manyetik rezonans.

Primary lymphomas of the rectum are rare (1,2). They are often included in the category of primary lymphoma of the gastrointestinal tract and not considered separately as primary rectal malignancies (3,4).

Non-Hodgkin's lymphoma (NHL) of the gastrointestinal (GI) tract accounts for 4% to 20% of all NHLs and is the most common extranodal site of presentation (5). The majority are B-cell lymphomas, although occasional T-cell lymphomas and anaplastic large cell lymphomas have also been described (6,7). A significant proportion of GI lymphomas arise from mucosal-associated lymphoid tissue (MALT). Distinct clinicopathologic entities, such as immunoproliferative small intestinal disease and multiple lymphomatous polyposis have also been described (5).

We report a case with primary B-cell lymphoma of the rectum with a fistula to the proximal ileum, which was demonstrated by MRI, using a per rectal paramagnetic contrast agent.

CASE

An 82-year old female presented with a three-month history of irregular bowel habits, rectal and abdominal pain, and bright red blood per rectum. She reported weight loss of approximately eight kg during this period. Her past medical history included a humerus fracture after an accident the previous year and treatment of peptic ulcer disease 15 years previously. There was no past history of inflammatory bowel disease, lymphoma, celiac disease or bowel irradiation. On physical examination, a 2 cm- to 3 cm palpable mass was noted on the posterior rectal wall 7 cm above the dentate line. On rectosigmoidoscopic examination the tumor appeared as a well-demarcated polypoid mass with eroded mucosa. Barium enema (BE) showed indirect findings of a fistula between the colon and small intestine. Colonoscopy demonstrated no other colonic lesions. Biopsy of the tumor revealed a diffuse large B-cell lymphoma. A CT scan of the thorax and abdomen showed no evidence of metastatic disease and there was no lymphadenopathy, while complete blood count and

Address for correspondence: Dr. Zeki Karasu
Ege Üniversitesi Tıp Fakültesi, Gastroenteroloji Bilim Dalı, İzmir
Tel /Fax: (232) 388 1969
e-mail: zkarasu@hotmail.com

Manuscript received: 15.7.2000 **Accepted:** 2.10.2001

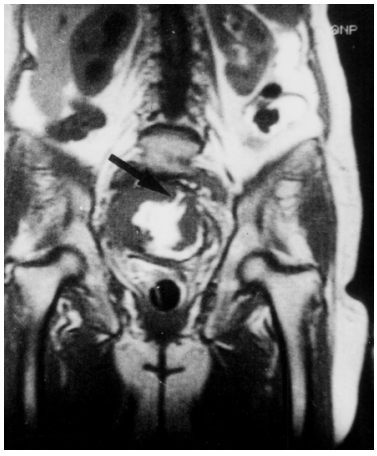

Figure 1. Following per rectal paramagnetic contrast agent (before IV contrast), MRI shows diffuse wall thickening (not narrowing the lumen), and a fistula tract to the ileum. The arrow illustrates the fistula tract.

bone marrow examination were normal. Since it is possible to obtain sagittal and coronal plane images of the lesion, magnetic resonance imaging (MRI) study was performed. With the use of per rectal paramagnetic contrast agent (@ Magnevist Enteral, Schering) MRI (0.5 T GE Vectra) showed diffuse wall thickening without narrowing of the rectal lumen. There was a fistula tract through the thickened wall to the proximal ileal segment (Figure 1).

The patient did not accept an advised surgical procedure and is being treated with a combination of chemotherapy and radiotherapy.

DISCUSSION

Non-Hodgkin lymphomas of the colon can occur as primary tumors or as secondary manifestations resulting from the evolution of other primary extranodal lymphomas (PEL) in other locations (8)

The criteria for the diagnosis of primary rectal lymphoma were described by Dawson et al: in 1961 (4). These criteria include 1) absence of palpable lymphadenopathy; 2) absence of mediastinal node enlargement on chest X-ray; 3) normal white cell count and differential in the evaluation of peripheral blood; 4) a lesion confined mainly to the bowel with only local lymphadenopathy seen at laparatomy; 5) absence of liver or spleen involvement at laparatomy. When such strict criteria are used, primary colorectal lymphomas prove to be rare tumors. Primary lymphomas account for 2 to 4 per cent of all malignant neoplasms of the GI tract. When the colon is considered separately it has been shown that lymphomas comprise less than 1.2 % of all large intestinal malignancies (1-3,9).

The incidence of PEL through the intestinal tract is reported as: small intestine: 42%, ileocecal region: 30 %, colon: 13 %, cecum: 8 %, rectum: 3 %, duodenum:2 %, anorectum:1 %. Multi foci were found in 43 cases (11%) (10).

Primary colorectal lymphomas have been associated with several other conditions, such as ulcerative colitis, Crohn's disease, previous local radiotherapy, AIDS, renal transplantation and ureterosigmoidoscopy (6,11,12). Colorectal lymphomas usually present with rectal bleeding, diarrhea, and abdominal pain and their incidence is higher in the 50-70 year-old age group (6,10-12).

A patient presenting with biopsy proven rectal lymphoma should undergo a complete physical examination to search for associated adenopathy, a chest x-ray to search for mediastinal enlargement and laboratory studies, including a white blood cell count with differential. Colonoscopy, barium enema and MRI or CT scan should be performed to search for synchronous lesions or involvement of other lymphoid regions (liver, spleen, abdominal / thoracic lymph nodes).

The overall five-year survival rate is reported to be 42 %, but using the updated Kiel classification, this survival rate decreases by 11 % for cases with high grade non-Hodgkin lymphoma, while there was a 62 % five-year survival for the cases with low grade lymphoma (1,13). Tumor size, histologic type and grade are prognostic factors, while the macroscopic type and distinction between T- and B-lineage are not (1,13).

The radiological signs observed during barium enemas for non-Hodgkin lymphomas are reported to be as follows: a small nodular pattern, frequently with multiple lesions (46 %), a diffuse or infiltrating pattern (25 %), a filling defect (23 %), endo- and exo-luminal images (18 %), ulcerating patterns (3 %), and a pure mesenteric form (1 %) (12). Associated radiological forms are present in 16 % of cases. The preferential site is the ceacum (53 % of the cases), followed by the rectum (21 %) (14).

Examination by CT and BE are complementary studies in the evaluation of primary lymphoma of the colorectum, and certain relevant findings may

be missed when radiographic evaluation does not include both modalities. Both CT and barium studies are very accurate in detecting bulky endoexoenteric tumor masses, but CT may suggest features which can differentiate primary lymphoma from adenocarcinoma. The real utility of obtaining both a CT and BE examination may lie more in diagnosing complications, such as fistula formation and in the evaluation of subtle infiltrative lesions. While CT is necessary for staging purposes, BE examination may be valuable for detecting subtle mucosal filling defects, which could be missed on CT (15).

Ultrasound, CT, and MRI have all been used for staging and endoexoenteric tumor demonstration (data is nevertheless limited about MRI in the literature). With the introduction of per rectal paramagnetic contrast agent, MRI offers the opportunity to obtain not only axial, but also sagittal and coronal images, making it superior to the other modalities. Moreover, in the case of patients who are surgical candidates, MRI, with its three dimensional images, is a very useful tool for surgeons to plan operation details. Although primary lymphoma of the rectum is reported sporadically in the literature, there is only one paper presenting a rectal lymphoma case with a fistula to the ileum (16). In our case, we demonstrated the fistula tract through the thickened rectal wall to the ileal segment in the axial, sagittal and coronal planes.

In conclusion, it is suggested that MRI may be used as an alternative tool in the diagnosis and staging of rectal tumors as well as in demonstrating complications like fistula. In selected cases, the use of an enteral contrast agent may improve the diagnostic quality of abdominal MRI by causing greater signal intensity, and also by it's functional aspect.

REFERENCES

1. Shepherd NA, Hall PA, Coates PJ, Levison DA. Primary malignant lymphoma of the colon and rectum. A histopathological and immunohistochemical analysis of 45 cases with clinicopathological correlations. Histopathology 1988; 12: 235-52.
2. Devine RM, Beart RW Jr, Wolff BG. Malignant lymphoma of the rectum. Dis Colon rectum 1986; 29:821-4.
3. Richards MA. Lymphoma of the colon and rectum. Postgrad Med J 1986; 62: 615-20.
4. Dawson IMP, Cornes J, Morson BC. Primary malignant tumors of the intestinal tract. Br J Surg 1961; 49: 80-9.
5. Crump M, Gospodarowicz M, Shepherd FA. Lymphoma of the gastrointestinal tract. Semin Oncol 1999; 26: 324-37.
6. Shepherd NA, Hall PA, Williams GT, et al. Primary malignant lymphoma of the large intestine complicating chronic inflammatory bowel disease. Histopathology 1989; 15: 325-37.
7. Gottlieb CA, Meiri E, Maeda KM. Rectal non-Hodgkin's lymphoma: a clinicopathologic study and review. Henry Ford Hosp Med J 1990; 38: 255-8.
8. Diebold J, Audoin J, Le Tourneau A. Anatomical aspects of non-Hodgkin's malignant rectocolic lymphoma. Ann Gastroenterol Hepatol 1984; 20: 325-9.
9. Aragon Sanchez FJ, Garcia Molina FJ, et al. Primary colorectal lymphoma. Rev Esp Enferm Dig 1997; 89: 903-18.
10. Hu RJ. Pathology and prognosis of 378 cases of primary intestinal malignant lymphoma (IML). Chung Hua Chung Liu Tsa Chih 1988; 10: 57-60.
11. Ioachim HL, Weinstein MA, Robbins RD, et al. Primary anorectal lymphoma. A new manifestation of the acquired immune deficiency syndrome (AIDS). Cancer 1987; 60: 1449-53.
12. Habr-Gama A, Campos FG, Ribeiro Jr U, et al. Primary lymphomas of the large intestine. Rev Hosp Clin Fac Med Sao Paulo 1993; 48: 272-7.
13. Iwashita A. Clinicopathologic study concerning primary malignant lymphoma of the large intestine. Stomach and Intestine 1995; 30: 869-86.
14- Bruneton JN, Thyss A, Bourry J, et al. Colonic and rectal lymphomas. A report of six cases and review of the literature. ROFO Fortschr Geb Rontgenstr Nuklearmed 1983; 138: 283-7.
15- Wyatt SH, Fishman EK, Jones B. Primary lymphoma of the colon and rectum: CT and barium enema correlation. Abdom Imaging 1993; 18: 376-80.
16- Bruck B, Dette S, Kaiserling E, Scheurlen M. A fistula between the rectosigmoid junction and the ileum as a complication of highly malignant AIDS-associated lymphoma. Dtsch Med Wochenschr 1991; 116: 1911-4.