RECTAL LYMPHOMA: A RARE CASE REPORT WITH REVIEW OF LITERATURE

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Abstract

Introduction: Colorectal lymphoma is an extremely rare disease. The disease is usually diagnosed in the advanced stages because of its primary nonspecific symptoms. Treatment modality of primary lymphoma of the rectum remains uncertain.

Case History: A 45-year-old male presented with rectal bleeding, pain in perianal region and alteration in bowel habits along with fungating and circumferential mass in the rectum. The histopathology and immunohistochemistry confirmed the diagnosis of Plasmablastic Lymphoma. He was planned six cycles of CHOP regime. On follow up the residual disease was managed with second line of chemotherapy (DHAP regimen). The patient was started on Metronomic chemotherapy (Chlorambucil & Etoposide) in view of persistent residual disease.

Conclusion: Optimal treatment for rectal lymphoma needs to be established.

Keywords: Rectal lymphoma, surgery, chemotherapy

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, stomach, bowel or regional lymph nodes (excluding retroperitoneal lymph nodes) found during laparotomy; 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 caecum (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.[9-11]

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. This case report describes a patient with primary rectal lymphoma which was managed medically with chemotherapy.

CASE REPORT

A 45-year-old male presented with the history of rectal bleeding, pain and swelling in perianal region along with alteration in bowel habits. On rectal examination there was an irregular fungating and the circumferential mass from the dentate line toward upper part. The mass was firm to hard, and fixed to the rectal wall.

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On abdominopelvic CT scan, extensive bulky rectal tumor was seen along with enlarged B/L iliac lymph nodes. Colonoscopy showed 11 cm length tumor in the rectum starting 2cm from anal verge. Patient underwent abdomino perineal resection with colostomy. Subsequent histopathological examination showed Poorly differentiated carcinoma. The lesion invaded beyond muscularis propria but serosa was free. Patient was referred to radiotherapy department for further management.

On immunohistochemical staining CD45, CD56, CD138, MUM-1, KAPPA and LAMBDA: positive, Ki-67= 80%, CD20, CD3, Cytokeratin: negative. Final diagnosis established was Plasmablastic lymphoma (Figure-1). PET CT Scan whole body showed post APR status metabolically active ill defined heterogeneously enhancing soft tissue density lesion in right ischiorectal fossa region, bilateral iliac lymph nodes enlarged (Fig.-1).

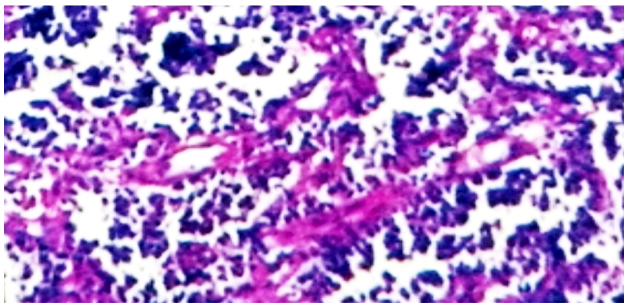

Figure-1: H & E showing Poorly Differentiated Carcinoma

He was delivered 6 cycles of CHOP regimen with complete resolution of clinical signs and symptoms. After 4 months patient developed complaints of lower abdominal pain and irregular episodes of constipation. PET-CT Scan revealed residual disease present and persistence of residual right internal iliac lymph nodes and soft tissue density nodular lesion adjacent to the jejunal loops (Fig.-2).

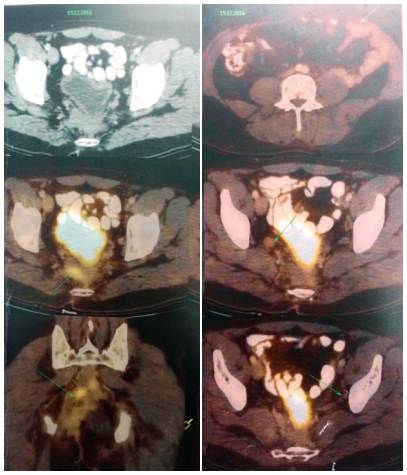

Figure-2: Pre and post chemo PET CT Scan showing residual disease after six cycles of CHOP

He was now started on second line chemotherapy with cisplatin 100mg / m^2 continues infusion, cytarabine 2gm/m^2 on day 2, dexamethasone 40mg on day1, day2 and day3. Patient received 6 cycles and was evaluated again by PET-CT Scan which showed a persistent residual disease. Presently patient is on Metronomic Therapy with Oral Chemotherapy (Chlorambucil and Etoposide). There is no progression in the disease process clinically and radiologically after 6 months..

DISCUSSION

Lymphoma of the rectum is a very rare, and there is still controversy about its treatment. In present case patient had a history of rectal bleeding, pain and alteration in bowel habits. Patients with 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]

On rectal examination, there was an irregular fungating and the circumferential mass from the dentate line toward upper part. Similarly findings were seen in a case reported by Ardakani J V et al[12].

Abdominal CT scan and endoscopy with biopsy are the most useful diagnostic tests. In our case we found that there was extensive bulky rectal growth along with enlarged B/L iliac lymph nodes were seen CT Scan abdomen. 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. According to Wyatt SH et al primary colorectal lymphomas manifested as discrete masses tend to have a greater depth of mural invasion than infiltrative lesions.[13]

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.[14,15] In our case did not have any of these risk factors.

Diffuse large B-cell lymphoma of the large bowel is generally treated with an uniform therapeutic approach: aggressive surgery followed by adjuvant chemotherapy. Some authors still believe that medical management should

be considered as the primary therapy even in surgically operable localized tumors. In our case patient underwent surgery followed by chemotherapy CHOP regimen, then in view of persistence of disease on PET- CT Scan, patient received second line chemotherapy DHAP regimen and presently he is on Metronomic chemotherapy (Chlorambucil and Etoposide) with no signs and symptoms of progression of disease.

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] The prognosis of present case whose histopathology is Plasmablastic Lymphoma is poor and a systematic review of 112 HIV-positive patients with PBL showed a median overall survival (OS) of 15 months and a 3-year OS rate of 25%.[16]

CONCLUSION

Lymphoma of rectum is rare. The therapeutic experience is limited and optimal treatment for rectal lymphoma needs to be established. Whether non surgical management with chemotherapy is sufficient to manage, further needs to be explored.

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