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REVIEW ARTICLE

Primary colorectal lymphoma: Case series and literature review

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Summary Primary colorectal lymphoma is a rare disease that accounts for 0.16% of colorectal malignancies. Treatments include surgical intervention with or without chemotherapy. Outcome of this intervention among the Chinese population are lacking. Perforation resulting from chemotherapy may need further exploration. A retrospective review of patient records was performed among those who were diagnosed with colorectal malignancy in a single center from January 1998 to June 2009. Ten patients met Dawson's diagnostic criteria for primary colorectal lymphoma [0.66% (10/1516) of all colorectal malignancies]. The male-to-female ratio was 9:1, and median age at diagnosis was 76 years. The most common site was the cecum ($n = 5$). B-cell lymphoma was present in eight patients. Seven patients underwent surgical intervention. The median follow-up of all patients was 16.5 months. Median survival was 17 months and 13 months in the surgical and chemotherapy group, respectively. Primary colonic lymphoma is a rare disease. Surgical intervention appeared to be superior to chemotherapy alone, but the findings were limited by the small number of patients in this study. Whether surgery or chemotherapy should be offered first remains unknown and requires further research.

1. Introduction

Primary colorectal lymphoma is a rare disease that accounts for only 5.8% of gastrointestinal lymphoma and 0.16% of colorectal malignancy.[1] The gastrointestinal tract is the most common site of extranodal lymphoma, which affects 6–12% of all patients.[2,3] However, most patients

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present late and have nonspecific symptoms such as abdominal pain and weight loss.[1,4]

The number of articles concerning primary colorectal lymphoma in the Chinese population is few. One study from Taiwan showed that the proportion of T-cell lymphomas (42%), known to have a poorer prognosis than B-cell lymphomas, is much higher in this population than in Western populations.[5] We review the incidence of primary colorectal lymphoma in our hospital and compare it with that of other studies.

2. Patients and methods

A retrospective study was performed of all patients who presented with primary colorectal lymphoma from January 1998 to December 2008. Clinical details derived from patients' medical records, including patient characteristics and clinical data, type of treatment, pathologic findings, recurrence rate, mortality, and morbidity, were recorded and analyzed.

All of the patients were diagnosed with primary colorectal lymphoma preoperatively. Tissue diagnosis was obtained by colonoscopy, and all patients underwent a computed tomography (CT) scan to determine the stage of disease and exclude metastases. Patients who had undergone emergency surgery were also excluded.

Our patients fit Dawson's criteria[6] for the diagnosis of primary colorectal lymphoma: histopathologic confirmation of a lymphoproliferative neoplasm confined to the colon and/or rectum with: (1) absence of palpable lymphadenopathy; (2) absence of mediastinal node enlargement; (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 laparotomy; and (5) absence of liver or spleen involvement at laparotomy. Statistical Package for Social Sciences (SPSS) version 15 (SPSS Inc., Chicago, IL, USA) was used for data analysis. Chi-square or Fisher's exact test was used in comparing the categorical variables. The Mann—Whitney U-test was used in comparing the continuous variables in median values.

3. Results

During the captioned period, 1516 cases of colorectal malignancy were identified from our database, and 15 patients were found to have lymphoma involving the colorectal region. However, only 10 patients (0.66%) fit Dawson's criteria. Nine patients were male and one was female. The median age at diagnosis was 76 years (range, 69—92). Seven patients complained of nonspecific abdominal pain, two patients had a change of bowel habit, and one patient complained of anemia. All patients underwent colonoscopy and had a biopsy taken for tissue diagnosis. The most common site was the cecum (50%, $n = 5$), followed by the rectum (40%, $n = 4$) and sigmoid colon (10%, $n = 1$). B-cell lymphoma was most common ($n = 8$), followed by Burkitt-like ($n = 1$) and Mantle cell ($n = 1$). A CT scan of the thorax, abdomen, and pelvis was performed, and none showed signs of distant metastasis.

All of the patients were assessed by physicians preoperatively to confirm the diagnosis and offered surgical intervention if appropriate. Three patients (30%; disease in cecum, sigmoid, and rectum, respectively) refused surgery and received chemotherapy as the sole treatment. The choice of chemotherapeutic agent depended on tumor pathology. There were no complications of bowel perforation during or after chemotherapy.

The remaining seven patients (70%) received surgical intervention. A right hemicolectomy was performed for five patients who suffered from lymphoma in the cecum. Polypectomy was performed on one patient who presented with a rectal polyp. One patient who had an abdominal perineal resection performed died of a cardiac event soon after surgery.

The median follow-up of all patients was 16.5 months. Differences in demographic data of the surgical and nonsurgical groups did not statistically affect outcomes significantly (Table 1). The median survival times were 17 and 13 months in the surgical and nonsurgical groups, respectively.

4. Discussion

Primary colorectal lymphomas usually present between the fourth and seventh decades of life; the average age at diagnosis is approximately 50 years. However, all of our patients were elderly, with a median age of 76 years, significantly older than patients in studies from Western countries who had a median age of 51—55 years. Although the exact incidence and prevalence are not known, the incidence of colonic lymphoma among all colorectal malignancies in our studied population (0.66%) is slightly higher than that in the Caucasian population (0.5%).[1,4,7—12]

Table 1 Comparison of the demographic data between the operative and chemotherapy group.

	Operative (n = 7)	Chemotherapy (n = 3)	p-value
Age (median)	78	74	0.454
Male	85.7% (6)	100% (3)	0.399
Site			
Caecum	71.4% (5)	0	0.073
Sigmoid	0	33.3% (1)	
Rectum	28.6% (2)	66.6% (2)	
Presentation			
Abdominal pain	71.4% (5)	100% (3)	0.644
Change of bowel habit	14.3% (1)	0	
per Rectal bleeding	14.3% (1)	0	
Abdominal mass	0	0	
Intestinal obstruction	0	0	
Cell type			
T-cell	0	0	0.240
B-cell	85.7% (6)	66.7% (2)	
Burkitt-like	0	33.3% (1)	
Mantle	14.3% (1)	0	
Class			
I	14.3% (1)	33.3% (1)	0.728
II	57.1% (4)	33.3% (1)	
III	28.6% (2)	33.3% (1)	
Median survival time (months)	17	13	0.124

It is unknown if the male predominance (9:1) in our study population differs from that of other populations because studies have shown varying results.[13,14]

Like results found in others' reports, our results show that the most common site of primary lymphoma is the cecum, attributable to the presence of relatively abundant lymphoid tissue.[4,12,14] However, our findings that 80% of our patients suffered from B-cell lymphoma differ from those of Wang et al[5] whose Chinese patients had predominantly T-cell lymphoma (42%). It has been shown that the T-cell type is associated with a poorer prognosis than B-cell type.[15,16] We cannot explain the difference.

The rarity of the disease renders clinical trials very difficult, and the optimal treatment remains uncertain. Most authors would offer the same treatment as used for colorectal carcinoma: surgery with or without chemotherapy. However, a review of the literature indicates no superiority of the treatment. Surgical intervention plays an important role in the management of the disease. Surgery with a curative intent can be offered to patients with localized disease; palliative surgical procedures can help in relieving obstruction or other symptoms. However, it is also important to treat patients who present with perforation, bleeding, or fistula. In our series, we did not randomize our patients to either a surgical or chemotherapy arm, and we offered surgical intervention as the initial treatment to all the patients who were fit and operable. The three patients who refused surgical intervention were referred for chemotherapy. We did not observe any difference in survival during a similar follow-up period. However, we cannot draw any conclusions based on the small sample size of this retrospective study. A Korean multicenter trial showed that the outcome of patients receiving chemotherapy alone was poorer than that of patients who received surgery with or without chemotherapy.[13] However, this result might be explained by the fact that most of the patients receiving chemotherapy were suffering from T-cell lymphoma, which has been shown to be associated with a poorer prognosis.[10,13] In one of our patients, we performed a laparoscopic abdominal perineal resection for his rectal lymphoma, but he died soon after surgery from a cardiovascular event. Therefore, whether the surgery offered benefit or imposed further risk to the patient remains unknown. Thus, the best treatment modality for primary colonic lymphoma cannot be concluded at this time.

One of the reported complications of chemotherapy in treating gastrointestinal lymphoma is tumor necrosis with bowel perforation.[17,18] The tumor with possible transmural involvement, which invades or weakens the bowel wall, might undergo tumor necrosis, causing bowel perforation as a result of the chemotherapy.[19] There is still no consensus or evidence to conclude the optimal timing for the combined treatment of surgical intervention and chemotherapy. One recent study reported two patients with colonic lymphoma who presented with a large bowel perforation due to obstruction rather than necrotizing enteropathy resulting from the chemotherapy. The series by Kim et al[13] of 95 patients with colorectal lymphoma demonstrated that none of the patients ($n = 23$; 24.5%) who received chemotherapy alone developed bowel perforation during chemotherapy. A literature search shows that bowel perforation following chemotherapy alone is uncommon; therefore, there is no solid evidence to correlate chemotherapy with bowel perforation.

For primary colorectal lymphoma, the evidence of surgery as the primary treatment is not strong. Chemotherapy should be considered as the primary treatment if the surgical risk is high, if it is the patient's preference, and if the patient understands the remote risk of bowel perforation. A preoperative CT scan can help to identify tumors with features of full-thickness bowel involvement and invasion. We also suggest a further exploration of the tumor pathology (type, size of the tumor, and site of perforation) and level of involvement of the bowel to analyze the relationship between chemotherapy and bowel perforation.

Our study has several limitations. This is a retrospective study with a limited number of patients, and, as a result, there was selection bias. The rarity of the disease rendered a prospective study very difficult. A meta-analysis would have helped in this case.

5. Conclusions

Primary colonic lymphoma is a rare disease. Our series seem to show superiority of the surgical intervention; however, it is limited by the number of patients. Whether surgery or chemotherapy should be offered first remains unknown. Several controversial issues are still unsettled, and further research is required.

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