

Primary Diffuse Large B-Cell Lymphoma of the Colon with Spontaneous Perforation: A Case Report

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ABSTRACT

We report a rare case of primary lymphoma of the colon with spontaneous perforation in A 54-year-old woman.

Keywords

Lymphoma, Perforation, Chemotherapy.

Introduction

Primary colorectal lymphoma is rare, representing only 2-6% of all colorectal malignancies and 10-20% of all gastrointestinal (GI) lymphoma sites [1]. The clinical presentation is commonly nonspecific, leading to diagnosis delays. Some cases of secondary colonic perforation have been reported in the literature, either as the initial presentation or after initiation of chemotherapy. We report a rare case of primary lymphoma of the colon with spontaneous perforation.

Case Report

A 54-year-old woman, with no medical history, presented with acute diffuse abdominal pain and vomiting for 2 days. There were no associated bowel disorders or deterioration of the general condition. Physical examination revealed diffuse abdominal tenderness. Pulse rate was 118 bpm, blood pressure was 100/65 mmHg and body temperature was 37.9°. Laboratory data showed white blood cells at 8.800/mm³, C-reactive protein at 20 mg/l, hemoglobin at 11.g/dl, urea at 6.2 mmol/l and creatinine at 43 µmol/l. Abdominal ultrasound showed free intraperitoneal fluid with caecal thickening. An emergent laparotomy was performed and showed diffuse purulent peritonitis with a ruptured 5 cm caecal tumor. No liver metastasis or peritoneal carcinomatosis were identified. Right hemicolectomy with ileocolostomy was

performed. The postoperative course was uneventful.

Pathological examination revealed a budding tumor mass of 8x7 cm, infiltrating and perforated. Histological examination showed a diffuse and focally nodular lymphoid proliferation infiltrating the colon wall (Figure 1A), reaching the serosa and associated with perforation. Tumor cells are large with centroblastic and immunoblastic features. They had an irregularly nuclei and visible nucleoli. Immunohistochemical analysis showed a diffuse and intense immunostaining with the CD20, confirming the B phenotype of tumour cells (Figure 1B). Tumour cells express diffusely and weakly the CD10. The other antibodies tested are negative (CD5, pancytokeratin, CD23 and cyclin D1). The tumor infiltrates the appendix. Among the 27 lymph nodes examined, 7 were metastatic. The resection limits were free from lymphomatous proliferation. The diagnosis of diffuse large B- cell lymphoma was made. The patient was referred for postoperative adjuvant chemotherapy. She is currently with no signs of recurrence, 7 months after his operation.

Discussion

Gastro-intestinal (GI) non-Hodgkin lymphomas (NHL) are rare tumors representing only 1% of all gastrointestinal tumors. The most common site of involvement is the stomach (50-75%), followed by the small intestine (20-30%) and the colon and rectum (10-20%) [2]. All histological subtypes of NHL can be seen in the GI tract but the most common types are diffuse large B-cell

lymphoma (DLBCL) and mucosa-associated lymphoid tissue lymphoma (MALT) [3,4]. The caecum is the most involved site of colorectal lymphomas, likely due to the presence of a native large lymphoid tissue [5]. Many risk factors and predisposing conditions have been associated with NHL of the GI tract such as autoimmune disease, family history, immunodeficiency, immunosuppression and environmental exposures [6,7]. The clinical presentation of PCL is commonly nonspecific leading to delays in diagnosis. They can manifest as abdominal pain, gastrointestinal bleeding, weight loss and deterioration of general status [8]. Bairey et al. reported that the most common presenting symptoms in their series of 17 patients were abdominal pain, altered bowel habits and weight loss [9]. Auger and Allan in their series of 22 patients with primary ileocecal lymphoma reported the same clinical presentations [10]. In our case, the patient was asymptomatic until she presented in the emergency department with diffuse abdominal pain due to perforation. Despite its rarity, perforation is a serious life-threatening complication of gastrointestinal lymphomas. Although some perforations occur after initiation of chemotherapy, others occur as the initial presentation of lymphoma. Vaidya et al. reported in their cohort of 1062 patients with GI lymphoma, that 9% of patients (92 of 1062) developed a perforation [11]. Out of 100 perforation events, 49 occurred during treatment with chemotherapy and 51 occurred as the initial presentation of lymphoma. The small bowel was the most common site of perforation (59%), followed by large bowel (22%) and the stomach (16%). Diffuse large B-cell lymphoma (DLBCL) was the most common lymphoma associated with perforation (59%, 55 of 92). The frequency of perforation in patients with colorectal DLBCL was 13.2% (12/91), compared with 0% (0/3) in patients with colorectal MALT. Otherwise, the frequency of perforation in patients with gastric DLBCL was 6% (13/215), compared with 0.6% (1/176) in patients with gastric MALT. Compared with indolent B-cell lymphomas, the risk of perforation was higher with aggressive B-cell lymphomas (hazard ratio, HR = 6.31, $P < 0.0001$) or T-cell/other types (HR = 12.40, $P < 0.0001$). These results suggest that the risk of perforation seems to vary by both the site of involvement as well as the type of lymphoma [12]. Bairey et al. reported in their series of 17 patients with colonic lymphomas, that 5 patients with DLBCL presented with acute abdomen due to perforation, requiring emergency surgery. The location was ileocaecal in 2 cases, sigmoid in 2 cases and caecal in 2 cases [9]. Ara et al. reported in their case series of eight patients with bowel perforations from lymphoma, that six of the eight perforations occurred in the small bowel and only 2 occurred in the large bowel [13]. Treatment of DLBCL usually involves chemotherapy, radiation, surgery or a combination approach. Chemotherapy based on cyclophosphamide, doxorubicin, vincristine and prednisolone (CHOP), associated with rituximab (R-CHOP), is now the standard of care in the initial treatment [7,14]. Radiation therapy has been used for regional adjuvant therapy to obtain local control in lymphoma, but its indications are increasingly restricted due to its high risks of complications especially in the small and large intestines [15]. Surgical resection, potentially followed by adjuvant chemotherapy, is the prime mode of therapy for emergent

conditions like obstruction, bleeding or perforation [16]. However, due to the rarity of the disease, no randomized clinical trials have been published yet to establish the optimal therapy of primary colonic lymphoma.

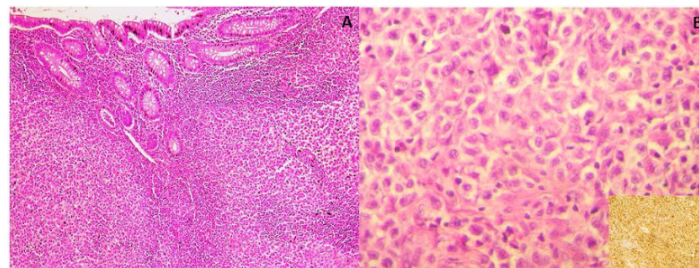


Figure 1: Pathological findings of diffuse and focally nodular lymphoid proliferation infiltrating the colon wall (A). Immunohistochemistry analysis showing a diffuse and intense immunostaining with the CD20 (B).

Conclusion

Primary diffuse large B-cell lymphoma of the colon is a rare case that may present with surgical complications as perforation.

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