

A rare case of extranodal marginal zone lymphoma of mucosa-associated lymphoid tissue of the descending colon

Qian Li, Li Yang, Honglin Ju, Siqi Cheng

Department of Gastroenterology, The First Affiliated Hospital of Hunan Normal University, China

Abstract

Mucosa-associated lymphoid tissue (MALT) lymphoma is pathologically characterized by lymphoepithelial lesions, and MALT lymphoma of the colon is relatively rarer than that of the stomach or small intestine. Endoscopically, colonic MALT lymphoma usually presents as a protruding polypoid mass with no clitoris and an ulcer in the cecum or rectum. In this paper, we analyzed a case of MALT lymphoma misdiagnosed as descending colonic polyp in Hunan Provincial People's Hospital, and reviewed and discussed the related literature to deepen the understanding of this disease.

Keywords

Colon, Lymphoma, Mucosa-associated lymphoid tissue, Colonoscopy.

1. Introduction

Marginal zone B-cell malignant lymphoma is a low-grade malignant non-Hodgkin's lymphoma that develops in mucosa-associated lymphoid tissue (MALT). MALT lymphoma is a relatively rare disease, but it is the third most common type of lymphoma, accounting for approximately 7% to 8% of all non-Hodgkin's lymphomas [1]. The most common site of MALT lymphoma involves the stomach (70%), although almost all other organs may be affected, including the lungs, salivary glands, ocular adnexa, skin, and thyroid gland [2]. Primary colonic MALT lymphoma is a rare malignant tumor with atypical clinical manifestations, mostly manifested as abdominal pain, blood in stool, localized swelling and change of fecal habit, which can cause intestinal obstruction in severe cases, similar to the manifestations of inflammatory bowel disease (such as ulcerative colitis, Crohn's disease) and intestinal malignancy [3], with high misdiagnosis rate and unclear prognosis, so the clinicians should pay attention to them.

2. Case presentation

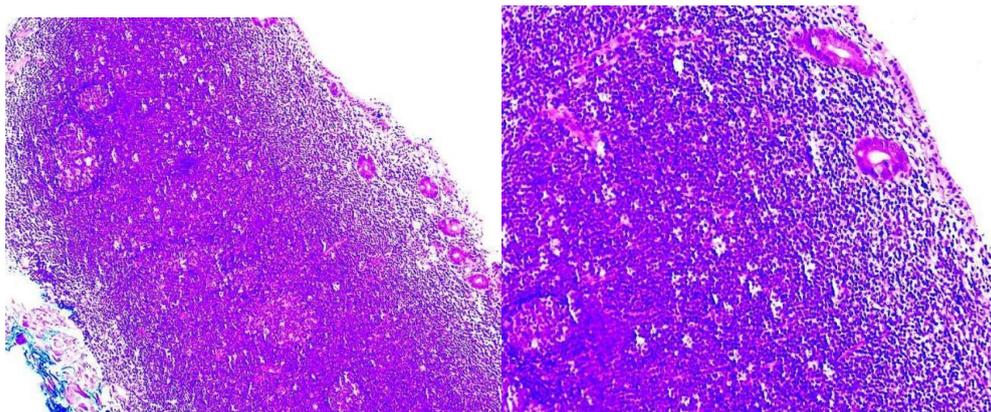
The patient was a 61-year-old male, admitted to the hospital for "epigastric pain for 4 days". The patient had paroxysmal hidden pain under the raphe 4 days ago without any obvious triggers, which could be tolerated without radiating elsewhere, and could be relieved by pressure, accompanied by abdominal distension, belching, and malaise, without any fear of cold, fever, vomiting, blood, black stools and other discomforts, and had received treatment in the local community hospital because of the symptoms that could be tolerated (specific diagnosis and medication were unknown). Because the symptoms were tolerable, the patient had been treated in the local community hospital (the specific diagnosis and medication were unknown), but the effect was unsatisfactory, and then he came to our hospital for further treatment. The patient was admitted to our hospital with the following symptoms: paroxysmal vague pain under the raphe, which was tolerable and did not radiate elsewhere and could be relieved by pressure, accompanied by abdominal distension, belching, fatigue, shortness of breath after activity, paroxysmal cough, white sputum, no fever, vomiting, vomiting of blood, black stools and other discomforts, poor mental health, poor sleep, poor appetite, unresolved stools for the

time being, normal urination, and no significant change in body weight. Physical examination: flat abdomen, no abdominal wall varicose veins, no gastrointestinal type and peristaltic wave, soft abdominal wall, subxiphoid pressure pain, no muscle tension and rebound pain, negative mobile turbidities, normal bowel sounds on auscultation. After admission, she completed the relevant examinations: laboratory tests of the three major routines, liver and kidney function, tumor markers were not abnormal. Electrocardiogram was normal. Enhanced CT of the whole abdomen: 1. gastric retention is possible, high density foci in the gastric pouch is considered gastric contents, please combine with the clinic; 2. left renal cyst is the same as before; 3. calcified foci of spleen is the same as before; 4. prostatic hyperplasia and calcified foci formation is the same as before. Gastroscopy: 1. Atrophic gastritis (C3); 2. Gastric polyp. Colonoscopy: LST of descending colon (EMR). Postoperative pathology: naked eye: descending colon polyp: a piece of mucosal tissue, size 2*1.5*0.5cm, mucosal surface texture is not uniform, around the cutting edge and the base of the ink coating (see Figure 1, 2).



(Figs. 1 and 2) The colonoscopic image of the descending colon showed a flat bulge of about 1.0 cm*1.5 cm with a reddish surface, and NBI showed that the lesion had clear borders, and black dots of CO were visible on the surface.

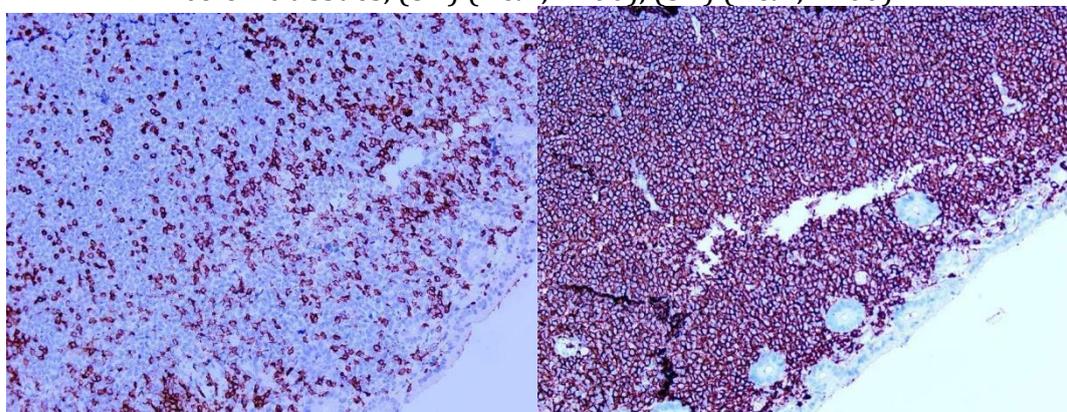
Postoperative pathology diagnosis: (descending colon polyp) lymphocyte hyperplasia, focal atypical hyperplasia, lymphoid follicle formation, combined with immunohistochemistry of low-grade lymphoma: Mucosa-associated lymphoid tissue lymphoma in the extra-nodal marginal zone (MALT lymphoma) can not be excluded, and it is recommended that B-cell gene rearrangement be done for further diagnosis (see Figure 2-3). Immunohistochemistry: 2340909-A07#: CK (pan) (epithelial +), CD5 (scattered T cells +), CD3 (scattered T cells +), CD21 (+), CD20 (++++), CD10 (germinal centers +), CD23 (FDC network +), Ki67 (+, outside the germinal centers of about 5%), Bcl-6 (germinal centers +), Bcl-2 (germinal centers +), and Ki67 (+, outside the germinal centers of about 5%). -2 (germinal center -), MUM1 (+), Pax-5 (++++), CD30 (-), EBER (in situ hybridization -), Kappa (-), Lambda (-), CyclinD1 (-).



(3A)

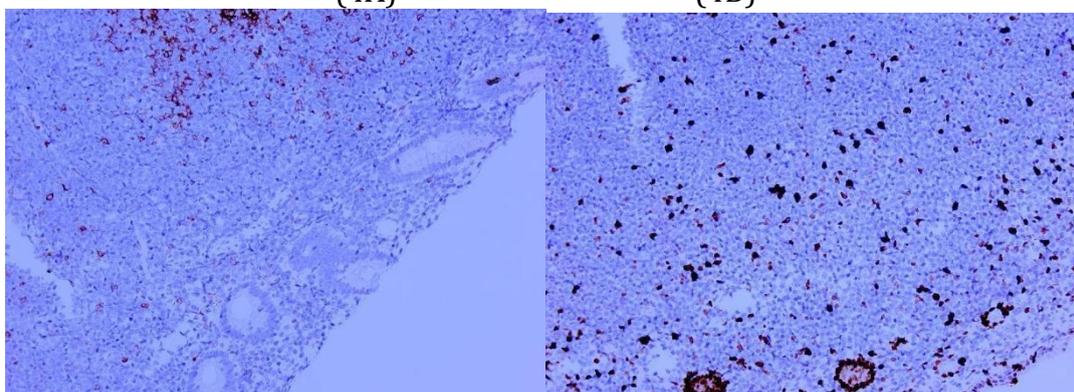
(3B)

(Fig. 3) Histopathological examination (hematoxylin-eosin staining) showed exuberant lymphocytic hyperplasia with focal atypical hyperplasia and lymphoid follicle formation in the colonic tissues, (3A) (H&E, $\times 100$), (3B) (H&E, $\times 200$).



(4A)

(4B)



(4C)

(4D)

(Figure 4) Immunohistochemical assay, (4A) Immunohistochemical staining of CD3 showed diffuse reaction in cell membrane ($\times 200$), (4B) Immunohistochemical staining of CD20 showed diffuse reaction in cells ($\times 200$), (4C) Immunohistochemical staining of CD23, (4D) showed diffuse reaction in cells, and immunohistochemical staining of KI-67 showed diffuse reaction in cells. diffuse reaction in the cells.

3. Discussion

MALT lymphoma was first described by Isaacson and Wright in 1983, and most colorectal lymphomas occur in the cecum or ascending colon, with $>70\%$ occurring near the hepatic flexure [4]. Gastric MALT lymphomas have been reported to be associated with *Helicobacter pylori* infection, whereas non-gastric MALT lymphomas have been associated with *Borrelia*

burgdorferi, Chlamydia psittaci, Hepatitis C virus, Campylobacter jejuni, and autoimmune diseases [5]. Clinical manifestations range from asymptomatic and detected only by screening colonoscopy, to manifestations of nonspecific abdominal discomfort, to more dramatic manifestations including gastrointestinal bleeding, problems with bowel movements, or pain, sometimes associated with intussusception or obstruction. On colonoscopy, the majority of MALT lymphomas present as a single mass with protrusion, ulceration, or absence of a pedicle or infiltrate, most commonly as a single polypoid lesion (70%), but also as multiple polypoid lesions (30%) [6].

Gastric MALT lymphoma is mainly associated with Helicobacter pylori infection, the main treatment for limited gastric MALT lymphoma is antibiotic therapy against Helicobacter pylori infection [7,8,9], and it has been reported that complete remission can be achieved in 70%-80% of the cases, and in case of poor efficacy or recurrence, gastric radiotherapy alone or in combination with chemotherapy and surgery is required [10]. The treatment options for colonic MALT lymph nodes are inconclusive, with surgery or chemotherapy being used as the first-line treatment in most cases [11], and the use of nitrogen mustard phenylbutyrate or rituximab in combination with cyclophosphamide, vincristine, and prednisolone for the treatment of colonic MALT lymphomas has also been reported in the literature [12].

Due to the rarity of the disease and the multiple features of colonic MALT lymphoma, the imaging manifestations of the disease have been described in only a very small number of cases in the past; it is poorly recognized clinically, and the rate of preoperative misdiagnosis is high. Therefore, we believe that mucosal biopsy of discolored but not prominent and ulcerated mucosa is also needed to exclude lymphoma.

References

- [1] Dong, Shuilin et al. "Primary hepatic extranodal marginal zone B-cell lymphoma of mucosa-associated lymphoid tissue type: A case report and literature review." *Medicine* vol. 96,13 (2017): e6305.
- [2] Schreuder, Max I et al. "Novel developments in the pathogenesis and diagnosis of extranodal marginal zone lymphoma." *Journal of hematology* vol. 10,3-4 91-107. 25 Sep. 2017,
- [3] Di Rocco, Alice et al. "Extranodal Marginal Zone Lymphoma: Pathogenesis, Diagnosis and Treatment." *Cancers* vol. 14,7 1742. 29 Mar. 2022,
- [4] Won, Jae Hee et al. "Clinical features, treatment and outcomes of colorectal mucosa-associated lymphoid tissue (MALT) lymphoma: literature reviews published in English between 1993 and 2017." *Cancer management and research* vol. 11 8577-8587. 20 Sep. 2019,
- [5] Zucca E, Conconi A, Pedrinis E, et al. Nongastric marginal zone B-cell lymphoma of mucosa-associated lymphoid tissue. *Blood*. 2003;101:2489-2495.
- [6] Kim, Myung Hwan et al. "A case of mucosa-associated lymphoid tissue lymphoma of the sigmoid colon presenting as a semipedunculated polyp." *Clinical endoscopy* vol. 47,2 (2014): 192-6.
- [7] Zucca E, Copie-Bergman C, Ricardi U, et al. Gastric marginal zone lymphoma of MALT type: ESMO Clinical Practice Guidelines for diagnosis, treatment and follow-up. *Ann Oncol*. 2013;24(Suppl. 6):vi144-48.
- [8] Ruskone-Fourmestreaux A, Delmer A, Lavergne A, et al. Multiple lymphomatous polyposis of the gastrointestinal tract: Prospective clinicopathologic study of 31 cases. *Groupe D'étude des Lymphomes Digestifs. Gastroenterology*. 1997;112:7-16.