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Case Report

Mucosa-associated lymphoid tissue lymphoma of the right colon: a rare presentation

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ABSTRACT

Mucosa-associated lymphoid tissue lymphoma (MALT) lymphoma affects frequently the gastrointestinal tract, most commonly the stomach. The colon is rarely affected. Both clinical presentation and endoscopic findings are variable and the histopathological with immunohistochemical analysis are needed for diagnosis. There are several treatment modalities available, with low remission rates and favorable outcomes. We present the case of an 80-year-old man who, after a positive screening faecal occult blood test, underwent a total colonoscopy which revealed an ulcerated lesion in the hepatic flexure of the colon compatible with lymphoma. After a staging CT scan of the chest, abdomen and pelvis without relevant findings except for a narrowing of the lumen of the hepatic flexure of the colon (where the tumor was located), the case was discussed by a multidisciplinary team, and it was decided to operate the patient. He underwent a laparoscopic right colectomy which was uneventful. Histopathological analysis of the surgical specimen, combined with immunohistochemistry, showed colonic MALT lymphoma. The patient subsequently completed staging with bone marrow aspiration (negative for lymphoma involvement) and respiratory testing for *Helicobacter pylori*, which was also negative. The patient has been followed for four years without recurrence and is completely asymptomatic. Colonic MALT lymphoma is rare and lacks characteristic presentation and diagnostic features. Surgical resection is a good therapeutic option.

Keywords: Colorectal lymphoma, Mucosa-associated lymphoid tissue lymphoma, Treatment, Surgery

INTRODUCTION

Mucosa-associated lymphoid tissue (MALT) lymphoma is a type of non-Hodgkin's lymphoma that originates from B cells.[1] It can occur anywhere outside lymph nodes but has a particular predilection for gastrointestinal tract, with the stomach being the most common site in up to 75% of cases, followed by small intestine, colon and rectum.[2]

Colon MALT lymphomas are rare, accounting for only 1.6% of MALT lymphomas and less than 1% of all colorectal cancers.[1,3] They mainly affect people in their fifth to seventh decade of life, with a slight female prevalence.[4] Presentation can vary from asymptomatic to severe gastrointestinal bleeding or obstruction.[3] There are usually no B symptoms. They have an indolent course and are considered to have a good prognosis.[5]

Because of their rarity, colonic MALT lymphomas are not well studied, and their characteristics, management and outcomes are not fully understood. There's currently no standard treatment or guidelines available for this type of lymphoma. There are several options for their management, ranging from local excision to oncological resection or chemotherapy.[1,5]

CASE REPORT

An 80-year-old man with a medical history of benign prostatic hyperplasia and previous abdominal surgery

(enterectomy for traumatic small bowel perforation) and no family history underwent a screening fecal occult blood test, which was positive. He was completely asymptomatic and had no complaints of weight loss, anorexia, abdominal pain, bloody stools, fever or night sweats. He, therefore, underwent a total colonoscopy, which revealed an ulcerated lesion with raised margins in the hepatic flexure of the colon, which was suspected to be malignant. Pathological exam showed an exuberant inflammatory process with the presence of lymphoid tissue composed mainly of small lymphocytes, compatible with lymphoma. Serum carcinoembryonic antigen level was normal (1.9 ng/ml) and the chest-abdomen-pelvis CT scan showed right-sided colonic thickening, without distant metastases.

After multidisciplinary team discussion, a laparoscopic right colectomy with intracorporeal anastomosis was performed.

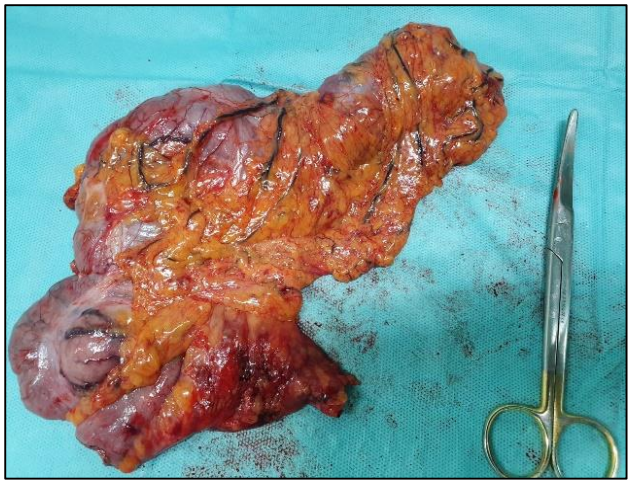

Figure 1: Operatory specimen (right colectomy).

The postoperative period (six days) was uneventful. Pathological exam showed a 5 cm lesion compatible with B-cell non-Hodgkin lymphoma. Immunohistochemistry was consistent with MALT lymphoma. The fourteen lymph nodes removed and surgical margins were both negative. Bone marrow aspiration was negative for lymphoma involvement. Respiratory testing for *Helicobacter pylori* detection was negative. Regarding the age of the patient, the absence of symptoms and the tumor characteristics (low grade), no additional treatments were performed. He is currently in the fourth year of follow-up, with no symptoms nor signs of recurrence.

DISCUSSION

MALT lymphoma is a rare subtype of non-Hodgkin's lymphoma that was first described by Isaacson in the 1980s.[4] It can occur in several organs, but the gastrointestinal tract is the most common extranodal site (50% of cases).[6] When affecting the gastrointestinal tract, this tumor has a predilection for the stomach, but in 10% of cases it can also affect the colon and rectum.[5] Only 2.5% of MALT lymphomas arise in the colon. The average age at diagnosis is 60 years, with some studies reporting a slight female preponderance. One factor known to be associated with gastric MALT lymphoma is chronic infection with *Helicobacter pylori* (Hp), but it's not known whether this is a risk factor for the development of colonic MALT lymphoma.[1] In this case, the patient was tested for Hp, which was negative.

The most common presenting symptom of colonic MALT lymphoma is gastrointestinal bleeding. Systemic B symptoms are usually absent. Other symptoms include abdominal pain, bowel obstruction and intussusception. In some cases, patients are asymptomatic, and the diagnosis is made on routine colonoscopy.[1-3] The endoscopic features can vary from a single polypoid lesion up to 5 cm in size to multiple infracentimeter lesions or a submucosal tumor. Sometimes the lesion is ulcerated, but this is not as common as in gastric MALT lymphoma.[4] In this patient, the endoscopic finding was a single ulcerated lesion measuring 5 cm.

Immunohistochemistry is essential for the diagnosis of MALT lymphoma. These tumors usually express CD19, CD20 and CD79a B-cell markers, and are negative for CD5, CD10 and bcl-2.[2-4]

Synchronous lesions are possible, so patients should have an upper gastrointestinal endoscopy, bone marrow biopsy, and thorax-abdomen-pelvis CT in addition to a total colonoscopy for proper staging.[2]

There are no guidelines or standardized treatment for colon MALT lymphoma. Most of the studies we can find in the literature are case reports and small retrospective series. The Japanese practical guidelines for hematological malignancies describe several therapeutic options for localized extragastric MALT lymphoma, such as observation without intervention, surgical or endoscopic resection, chemotherapy or radiotherapy.[1,5,7] The most used therapeutic approaches are surgical resection and chemotherapy, both with good results and high remission rates (more than 90%). In our case, the patient had an ulcerated lesion with reduced colonic caliber, which was already affecting the surrounding fat, with a risk of occlusion or bleeding in a short period of time, so the preferred therapeutic option was surgical resection. Despite the patient's age, the anesthetic and surgical risk were low.

Colonic MALT lymphoma is a low-grade tumor with a very good prognosis and a very low recurrence rate, as we could see in this case.[4]

CONCLUSION

MALT lymphoma is a low-grade B-cell lymphoma that rarely develops in the colon. Presentation can vary from

no symptoms to life-threatening bleeding or bowel obstruction. The diagnosis is made by colonoscopy and immunohistochemistry. There are several therapeutic options, the most used being surgical resection or chemotherapy, both with good results. Unfortunately, there are few cases in the literature which prevent the development of guidelines for the management of these tumors. In this case, the preferred treatment modality was surgical resection, and the long-term results are very good. These are indolent tumors with an excellent prognosis and a high remission rate. Studies are still needed to standardize the diagnosis and treatment of this rare disease.

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