

Title:

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Primary colon mantle lymphoma: a case report of a misleading macroscopic appearance!

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AUTHOR CONTRIBUTIONS

Arieira C drafted the manuscript.

Dias de Castro F and Boal Carvalho P revised the manuscript.

Cotter J critically revised the manuscript and approved the final version for submission.

ABSTRACT

Mantle cell lymphoma (MCL) of the colon is a rare entity, usually presenting as lymphomatous polyposis. We report the case of a 43-year-old male with an unusual colonic MCL in the form of a single exophytic and ulcerous lesion with necrotic areas associated with an ileo-cecal intussusception. The endoscopic appearance suggested adenocarcinoma. However, the diagnosis was made by histology and immunohistochemistry studies of the endoscopic biopsies.

Key words: Lymphoma. Mantle-cell. Colonoscopy. Intussusception.

INTRODUCTION

Mantle cell lymphoma (MCL) is a rare and aggressive B-cell non-Hodgkin lymphoma (NHL) defined by the translocation t (11; 14) (q13; q32) that results in the overexpression of protein cyclin D1 (1). It comprises 2-10% of all NHLs and frequently involves extranodal sites, including the spleen, bone marrow and gastrointestinal tract. Gastrointestinal involvement is found in 5-20% of all cases of MCL (2).

Primary gastrointestinal lymphoma is very rare, accounting for only about 1-4% of all gastrointestinal malignancies (3). MCL has one of the worst prognoses of all B-cell NHLs, with a 5-year survival as low as 27% (4). Primary MCL of the colon is a rare entity, with only a few case reports in the literature and the most common endoscopic presentation is lymphomatous polyposis (5). The authors report a case of a colic MCL of the cecum in a 43-year-old male patient with unusual endoscopic features.

CASE REPORT

A 43-year-old male with a previous history of dyslipidemia and medicated with atorvastatin was referred to our unit due to cramping abdominal pain with a 2-month duration. On physical examination, the abdomen was normal and no organomegaly or palpable lymph nodes were noted. The blood count revealed a microcytic and hypochromic anemia (hemoglobin 10.7 g/dl), with no other abnormalities.

During the investigation, the patient underwent an esophagogastroduodenoscopy that was normal and a colonoscopy that demonstrated a bulky exophytic and ulcerous lesion with necrotic areas in the cecum. Underneath the lesion, there was normal ileal mucosa compatible with ileo-colic intussusception (Fig. 1). In addition, a subepithelial lesion was identified at the rectum-sigmoid transition, 18 cm from the anal verge that was covered by 15 mm of normal mucosa (Fig. 2). The remaining colonic segments were normal.

Biopsies were taken from the mass and the subepithelial lesion, which showed atypical lymphocytes infiltrating the lamina propria in both samples. The tumor cells were diffusely positive for CD5, CD20, CD43, CD79a, bcl-2 and cyclin D1 on immunohistochemical staining, which was consistent with the diagnosis of MCL. Subsequently, abdominal CT scan with intravenous contrast administration was performed for staging. This confirmed the presence of a transverse colon invagination with diffuse parietal thickening with multiple satellite

enlarged lymph nodes associated with fat striation, which was compatible with local spread (Fig. 3).

Due to his symptoms, he subsequently underwent a laparoscopic right hemicolectomy and 10 cm enterectomy and the pathologic examination confirmed the diagnosis of lymphomatous polyposis MCL. In addition, a bone marrow core biopsy showed marrow involvement by MCL.

Combining all the findings, the patient was classified as stage IV-a according to the Ann Harbor classification. A treatment regimen of chemotherapy with R-CHOP (rituximab, cyclophosphamide, vincristine and prednisone) and R-DHAP (rituximab-dexamethasone, cisplatin and cytarabine) followed by autologous stem cell transplantation (ASCT) for consolidation was proposed. Thus far, the patient is well and undergoing chemotherapy.

DISCUSSION

MCL was recognized since 1994 as a non-Hodgkin B-cell lymphoma that originated in the mantle region of the lymphoid follicles, characterized by the overexpression of the cyclin-D1 gene as a result of the chromosomal translocation t(11; 14) (q13; q32) (1). On immunohistochemistry, tumor cells are characteristically CD5 and pan B-cell antigen positive (CD19, CD20, CD22) and negative for the expression of CD10 and CD23 (1).

MCL is an uncommon subtype of NHL that comprises 2-10% of NHBCL, usually with an aggressive behavior and a median overall survival of three to five years (6). MCL most commonly affects males (2:1) and usually presents in the fifth or sixth decade of life (1). The usual presentation is generalized lymphadenopathy, although extranodal involvement is common at sites such as the peripheral blood, bone marrow and gastrointestinal tract (2,6). Primary MCL of the colon is a rare entity and the most common symptoms are abdominal pain and weight loss, with a palpable abdominal mass identified on physical examination (7). The most common sites of involvement are the ileocecal region and cecum and the macroscopic appearance varies from a tumoral mass, ulcer and mucosal thickness to multiple polypoid lesions (7). The most common endoscopic feature of primary intestinal MCL is multiple lymphomatous polyposis, which has been described in some reports (1). There are few reports in literature of a single colonic mass as a presentation of primary colic MCL (8,9).

With regard to prognosis, the median duration of remission in most reported trials was 1.5-3 years (1). The Mantle Cell Lymphoma International Prognostic Index (MIPI) score is frequently used to assess prognosis, which includes factors such as age, sex, performance status, LDH level, white blood cell count and the percentage of Ki-67-positive cells determined by immunohistochemistry (1). Patients are stratified into three risk groups: low risk (5-year overall survival rate of 60%), intermediate risk (median survival 51 months) and high risk (median survival 29 months) (1). There is currently no standard of care for patients with newly diagnosed MC due to the rarity of MCL (1). However, therapy usually consists of systemic chemotherapy with CHOP regimen (cyclophosphamide, hydroxydaunomycin, Oncovin® [vincristine] and prednisone), hyper-CVAD (hyperfractionated cyclophosphamide, vincristine and doxorubicin [adriamycin], dexamethasone, methotrexate and cytarabine) with or without rituximab, R-CHOP alternating with R-DHAP (rituximab, cisplatin, cytarabine, dexamethasone), hyper-CVAD with autologous stem cell transplantation and single alkylating agents like chlorambucil (1). Salvage therapies such as bortezomib, lenalidomide and ibrutinib have been used for relapsed and refractory disease. The role of surgery in primary gastrointestinal lymphoma is controversial and is usually restricted to patients presenting with intestinal occlusion, bleeding and perforation, as in the case described here (10).

In conclusion, here we describe a case of primary colonic MCL with an unusual presentation. Although a rare entity, primary colonic lymphomas should be included in the differential diagnosis of single colonic lesions. The awareness of such occurrences is necessary and may help with the differential diagnosis of a colon single mass.

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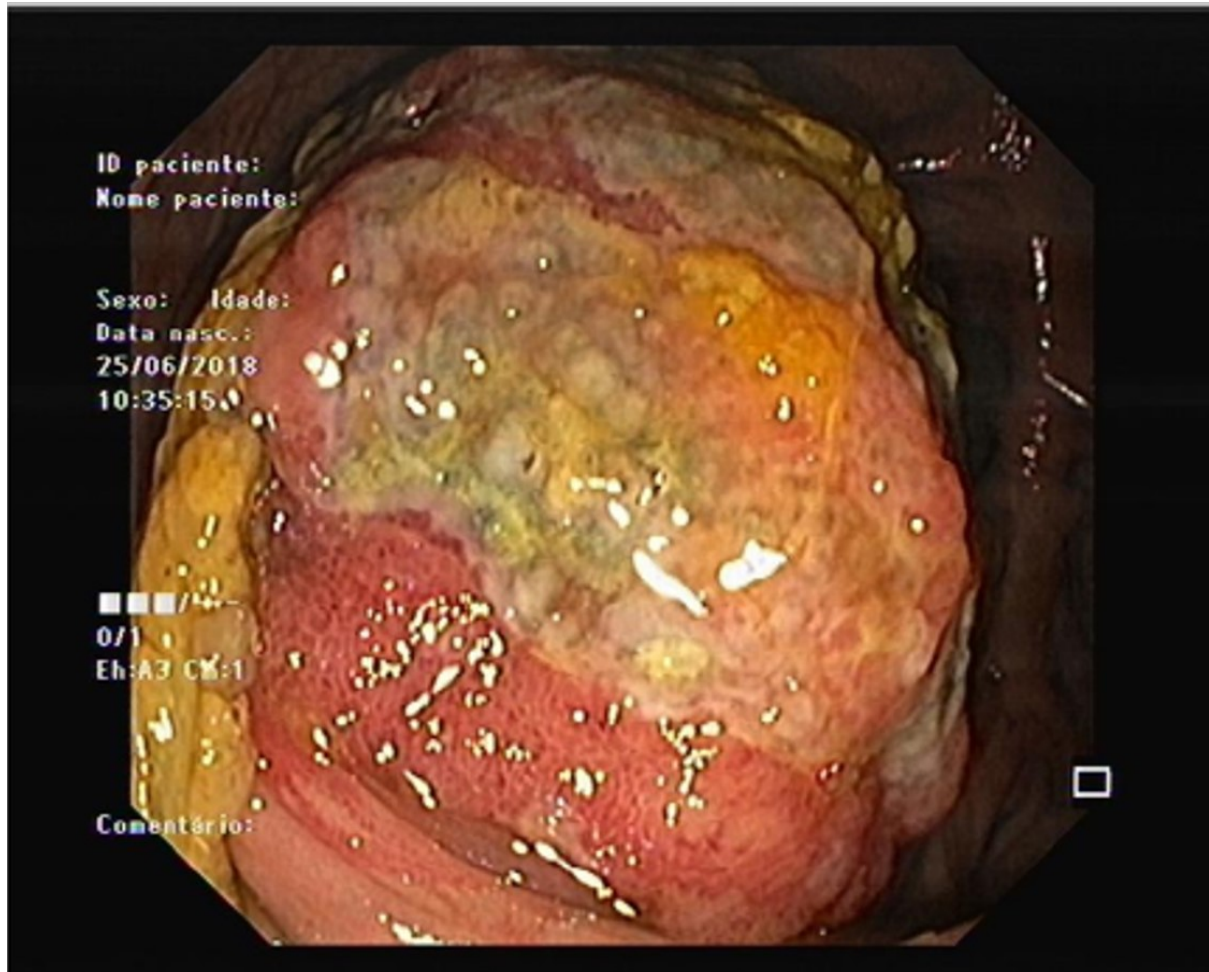


Fig. 1. Exophytic and ulcerous lesion with necrotic areas in the cecum.

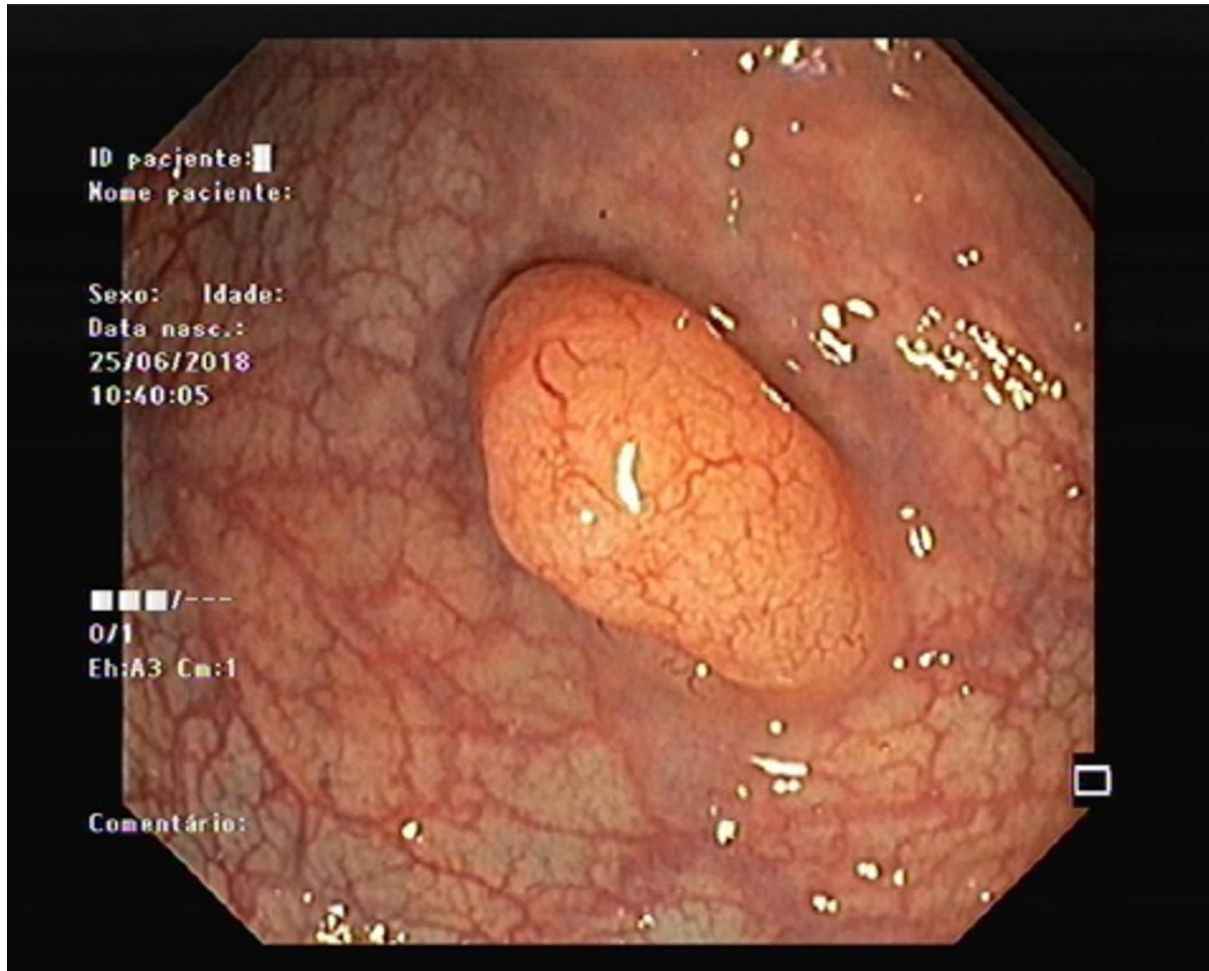


Fig. 2. Subepithelial lesion of the rectum-sigmoid transition.



Fig. 3. Abdominal CT scan with intravenous contrast administration.

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