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Primary Colon Lymphoma. Case Report

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ABSTRACT
Introduction: *Primary colon lymphoma is a rare disease, which represents between 0.2 and 0.6% of colorectal cancers. Most extranodal lymphomas involve the gastrointestinal tract. These are usually non-Hodgkin's lymphomas.*
Objective: *To evaluate a rare pathology of the colon from a clinical case.*
Case: *A 53-year-old man presented to the emergency room complaining of acute fever, abdominal pain and diarrhea. A palpable mass on the right flank was noted on physical examination. A right colectomy was performed due to perforation of a cecal tumor. Pathological diagnosis: large B-cell non-Hodgkin lymphoma.*
Conclusion: *It is a rare entity, which mainly affects the colon proximal to the hepatic flexure. The combination of surgery and chemotherapy is the treatment of choice*

Keywords: *Lymphoma; Colon*

INTRODUCTION

Primary colon lymphoma, a rare disease described by Billroth in 1871, represents 0.2-0.6% of colorectal cancers.[1,2] Extranodal non-Hodgkin´s lymphomas more frequently involve the gastrointestinal tract (30-40% of cases). The predominant location is the stomach (50-60%), followed by the small intestine (20-30%) and finally the colon and rectum (10-20%).[3] Inflammatory bowel disease (IBD) and immunosuppression represent risk factors. The symptoms are not pathognomonic, so the diagnosis is frequently made in advanced stages. Treatment involves a multidisciplinary approach combining surgery, chemotherapy, and radiation therapy in selected cases.[2] The objective of this work is to evaluate this rare disease of the colon.

CASE

A 53-year-old male patient, HIV + and with a history of tuberculosis, presented to the emergency room complaining of a four-day history of abdominal pain and diarrhea. On physical examination, he presented fever and a distended abdomen, with guarding and rebound, and a palpable abdominal mass at the right flank. WBC: 16000 mm^3.

Computed tomography showed a 98 x 88 mm, lobulated, irregular, heterogeneous mass, with gas bubbles, in the region of the right flank in close relationship with the ascending colon. Other findings were adjacent peritoneal fat stranding, lymphadenopathies, parietal thickening of the loops of the adjacent ileum and dilated small bowel loops (Fig. 1).

Exploratory laparoscopy was indicated with a presumptive diagnosis of perforated ascending colon tumor. During surgery, a large, perforated, cecal tumor was found, with supra and inframesocolic purulent fluid, for which it was converted to open surgery. A right colectomy with end ileostomy and mucosal fistula was performed, with lavage and drainage of the peritoneal cavity (Fig. 2). The outcome was favorable, and the patient was discharge after 10 days.

Histopathology reported a high-grade non-Hodgkin´s lymphoproliferative-appearing process, with peri-intestinal lymph nodes free of neoplasia (Fig. 3). Immunohistochemistry: morphological characteristics and immunological profile corresponding to a mature or peripheral B-cell non-Hodgkin´s lymphoma diffuse large cell lymphoma subtype. CD 20 (L26) (Pan-B): Positive; CD 3 (Pan T): Negative; DC 10: Negative; CD 30: Negative; CD 5: Negative; Ki 67: 70% labeled nuclei; Cyclin D1: Negative (Fig. 4).

The Oncohematology Service decided to perform adjuvant treatment with the RDAEPOCH scheme (Rituximab, Doxorubicin, Vincristine, Etoposide, Cyclophosphamide, Meprednisone).

DISCUSSION

Lymphoma ranks third among colorectal cancers, after adenocarcinoma and neuroendocrine tumors.[1] The highest incidence is described between the fifth and seventh decades of life, being 1.5 times more frequent in men.[3] Most colorectal lymphomas are found proximal to the hepatic flexure and the cecum is the predominant site (57%)

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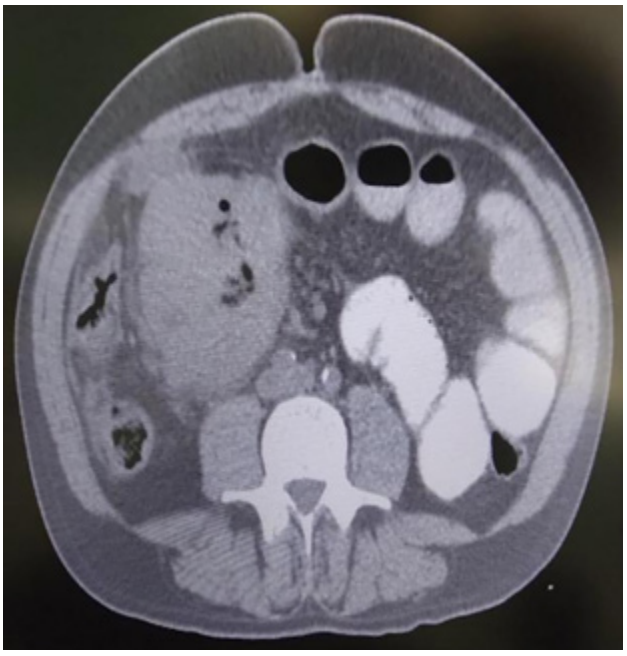

Figure 1: Computed tomography showing a 98 x 88 cm lobulated mass with air bubbles on the right flank, and enlarged lymph nodes.

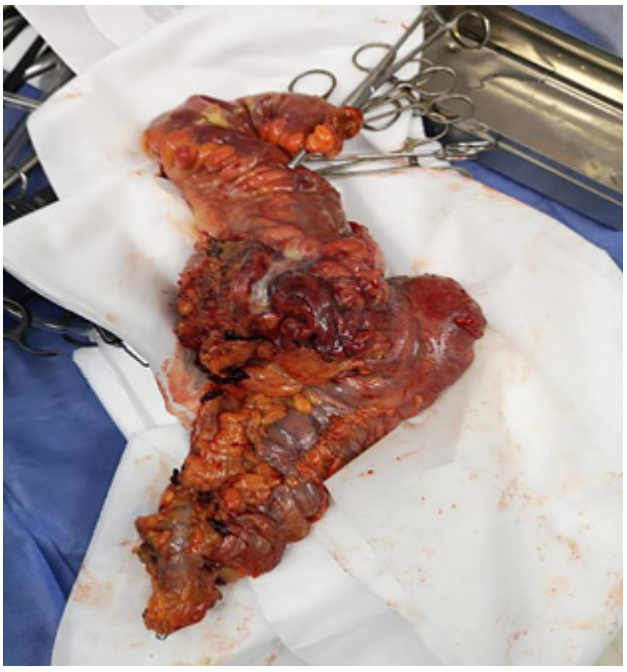

Figure 2: Specimen of the perforated tumor of the distal ileum and cecum. It measures 9 x 8 cm, has 100 % circumferential involvement and infiltration of the pericolonic fat.

due to its high density of lymphoid tissue, followed by the ascending colon (18%), transverse colon (10%), rectum and sigmoid colon (10%) and descending colon (5%).[3]

Gastrointestinal lymphomas can be primary or secondary, which determines the prognosis and treatment. Dawson in 1961 described the criteria to define a primary lymphoma:

1. Absence of peripheral lymphadenopathy and no evidence of mediastinal lymphadenopathy on the chest X-ray.
2. Normal blood tests and bone marrow biopsy.
3. At laparotomy, only regional nodes should be involved.
4. The liver and spleen must be free of tumor involvement.[1-4]

IBD and immunosuppression (HIV +, transplantation, chronic Epstein Barr virus infection) have classically been considered risk factors. HIV+ patients have a higher incidence of anorectal lymphomas.[23] Clinically, it lacks pathognomonic symptoms, the most frequent being abdominal pain, weight loss, and changes in bowel habits.[4] Rectum involvement can manifest as lower gastrointestinal bleeding. In 25% of cases, lymphoma presents as an intestinal obstruction, due to stenosing tumor or intussusception. Perforation is rare.[5] Some cases are diagnosed incidentally. On physical examination, it is common to palpate an abdominal mass where the lesion is located (41%).[3]

Primary colon lymphoma has several histological types, the most common the diffuse large B-cell lymphoma (60%), followed by mantle cell lymphoma (MALT), Burkitt lymphoma, and follicular lymphoma.[3] Large B lymphocytes non-Hodgkin's lymphomas have a worse prognosis, with relapses of around 70 % at 5 years. The vast

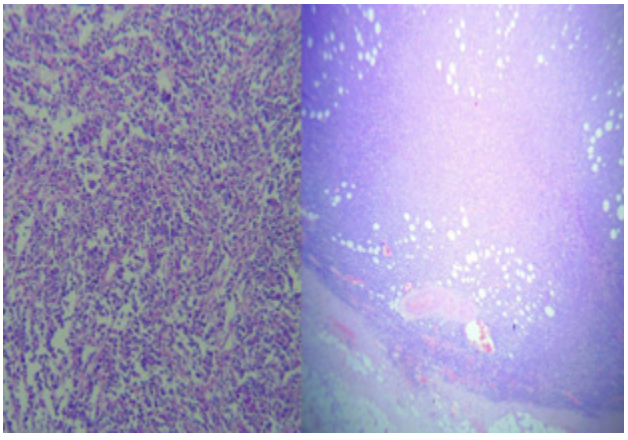

Figure 3: Solid atypical neoplastic proliferation made up of cells with a lymphoid appearance of medium to large size, figures of mitosis and foci of necrosis. The neoplasm infiltrates and ulcers the mucosa, compromising the ileocolic wall up to the adjacent fatty tissue (H&E).

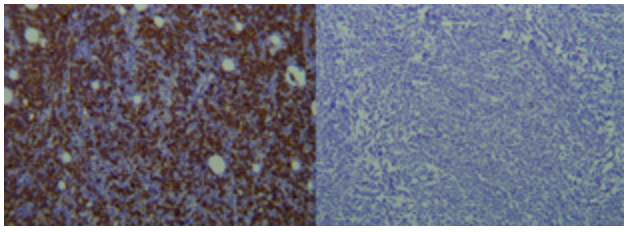

Figure 4: Immunohistochemical study CD 20 (L26) (Pan-B): Positive (left), CD 3 (Pan-T): Negative (right).

majority of these patients evolve with disseminated disease. Preoperative diagnosis is rare due to the absence of specific symptoms. Computed tomography is one of the common diagnostic methods. It can show localized or diffuse involvement of the colon, with or without regional lymphadenopathies. The primary lesion may manifest as a polypoid mass, a circumferential cavitary lesion, focal nodularity of the mucosa, as well as diffuse ulcerated or nodular lesions. Tomographic findings are nonspecific and may be similar to adenocarcinoma, IBD, and familial adenomatous polyposis.[3] Colonoscopy with biopsy is the preoperative diagnostic method of choice. Endoscopic findings are classified into three categories: mucous, polypoid, and massive lymphoma. Mucosal lesions are characterized by superficial erosions or deep ulcerations, with congestive and edematous borders. Polypoid lesions have a broad, sessile or pedunculated base, with a smooth or granular surface. The massive lesions are classified as lobulated neoplastic lesions that protrude intraluminally, with or without superficial ulcers.[3]

There are different classification systems for the staging of the disease, one of the most used at present is the Ann Arbor staging system modified by Musshoff (Table 1). This classification differentiates the nodes involved into: regional, non-contiguous regional and distant. It provides more information on overall and disease-free survival, as well as discrimination between localized and disseminated disease.[3]

Treatment of colorectal lymphoma usually includes surgery and chemotherapy, without reaching a consensus, due to the low incidence of the disease. Surgery is indicated for complications (obstruction, perforation and/or bleeding). As the only treatment, it should be limited to specific, localized, low-grade cases. In most cases there is a spread outside the local region, in which case the basis of treatment should be chemotherapy associated or not with surgery.[2] Radiation therapy is used in selected cases, due to its intestinal side effects. It may be indicated in the case of incomplete resection or in for recurrence in soft tissues or retroperitoneum.

TABLE 1. ANN ARBOR STAGING SYSTEM MODIFIED BY MUSSHOFF.

Stage	Description
I	Involvement of a single nodal group or single extranodal site (IE).
II	Involvement of more than one nodal group on the same side of the diaphragm or single extranodal site and adjacent lymph nodes (IIE).
III	Involvement of multiple nodal sites on both sides of the diaphragm, including extranodal sites (IIIE) or spleen (IIIS).
IV	Bone marrow, central nervous system, or diffuse visceral involvement.

The chemotherapy regimen known as CHOP (cyclophosphamide, doxorubicin, vincristine, and prednisone) is the first-line treatment for primary lymphomas of the colon and rectum. Recently, several studies have shown that the association of the monoclonal antibody Rituximab with this scheme increases the number of responses and survival.[2-4]

CONCLUSION

Primary lymphoma of the colon is a rare disease. Because its incidence increases in risk groups related to this disease (IBD, immunosuppression) it should be taken into account among the differential diagnoses of colorectal disease.

The combination of surgery and chemotherapy is the treatment of choice in most cases. Monoclonal antibodies, such as Rituximab, associated with chemotherapy, could improve the survival rate.

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