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Primary lymphoma of the colon: report of a case and review of the literature

Linfoma primario de colon. Reporte de un caso y revisión de la literatura

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ABSTRACT

Primary lymphomas of the colon (PLC) are infrequent neoplasias of the gastrointestinal tract; their incidence is 0.2 a 0.5% among all primary colon neoplasias. They appear mostly in adulthood. The most frequent variety is non-Hodgkin's lymphoma. Multiple lymphomatous polyposis is an infrequent presentation of B-cell non-Hodgkin lymphoma of the colon. The most frequent site of development is the stomach, followed by the small intestine and in third place, the colon. In the colon, the most compromised sites are the cecum and rectum. The treatment of choice of primary lymphomas of the colon is surgical, with adjuvant chemotherapy and in some cases, radiation therapy. We herein report the case of a 62-year old male referred to our outpatient clinic with a three-month history of asthenia, fatigue, hyporexia and lower gastrointestinal bleeding. The diagnosis of B-cell primary non-Hodgkin lymphoma of the colon was established, presenting as diffuse lymphomatous polyposis of the ascending colon. The patient was successfully treated surgically followed by adjuvant chemotherapy.

RESUMEN

Los linfomas primarios del colon (LPC) son neoplasias poco frecuentes del tracto gastrointestinal; su incidencia es de 0.2 a 0.5% de todas las neoplasias primarias del colon. Se presentan sobre todo en la edad adulta. La variedad más frecuente es el linfoma no Hodgkin. La poliposis linfomatosa múltiple es un tipo infrecuente de presentación de linfoma de colon no Hodgkin de células B. El sitio más frecuente de aparición en el aparato digestivo es en estómago, seguido de intestino delgado y, en tercer lugar, colon. Los sitios más frecuentes de presentación en el colon son el ciego y el recto. El tratamiento de elección en los linfomas primarios del colon es quirúrgico, con quimioterapia adyuvante y, en algunos casos, radioterapia. Informamos el caso de un paciente de sexo masculino de 62 años de edad referido a nuestra consulta externa por un cuadro de evolución de tres meses caracterizado por astenia, adinamia, hiporexia y sangrado de tubo digestivo bajo. Se realizó el diagnóstico de linfoma primario de colon no Hodgkin variedad de células B, con la presentación de poliposis linfomatosa difusa en colon ascendente. El paciente fue tratado con éxito mediante cirugía y, posteriormente, sesión de quimioterapia adyuvante.

INTRODUCTION

Primary lymphoma of the gastrointestinal tract was initially described by Billroth in 1871; it represents from 1 to 4% of all malignant tumors of the gastrointestinal tract.[1] In order of frequency, the affected digestive tract organs are as follows: stomach (60%), small intestine (25%) and colon (14%).[1] The etiology of gastrointestinal lymphomas is unknown, but most are associated to the mucosa (mucosa-associated lymphoid tissue, or MALT).[1,2] To establish the diagnosis of a primary lymphoma of the gastrointestinal tract, the criterion is morphologic; immunophenotype and immunogenetic analyses should be included. The criteria used to determine whether a lymphoma is primary of the digestive tract are those described by Dawson *(Table 1)*.[1]

Primary lymphoma of the colon (PLC) is an infrequent neoplasia, with an incidence of 0.2 to 0.5%; the B immunophenotype includes

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Table 1: Dawson criteria for labeling primary gastrointestinal lymphoma.

1. Gastrointestinal tumor histologically confirmed to be a lymphoma
2. No liver or spleen enlargement
3. No peripheral lymphadenopathy
4. No evidence of lymphoma in the chest by CT scan or chest X-ray
5. Blood work and bone marrow biopsy reported as normal

mantle cell lymphomas presenting as multiple lymphomatous polyposis and the Burkitt and non-Burkitt lymphomas.[2,3]

Certain conditions have been associated with an increased incidence of PLC, such as non-specific chronic ulcerative colitis, Crohn's disease, HIV-positive immunosuppressed patients, and posttransplant recipients.[4]

In 5% of patients, there are synchronous tumors or diffuse multiple polyposis in several or all colon segments.[4] Multiple lymphomatous polyposis (MLP) is an unusual variant of B-cell non-Hodgkin gastrointestinal lymphoma presenting with several lymphomatous polyps in one or more long segments of the gastrointestinal tract. Its clinical and morphological features were described by Cornes in 1961; since then, no more than 100 cases have been reported.[4] The treatment of choice for primary lymphoma of the colon is surgical resection.[4-6] In some patients, an increased survival has been observed when adding adjuvant chemotherapy and/or radiotherapy.[7]

The following case of PLC is among the few that have been documented in our hospital and in Mexico. There are no national statistics on extranodal lymphomas.

CLINICAL CASE

A 62-year old male with no relevant medical history was referred to the Department of General Surgery with a three-month history of lower gastrointestinal bleeding, presenting with melena, asthenia, fatigue, abdominal pain (particularly in the right lower quadrant) and weight loss of approximately 10 kg over the previous two months. Laboratory tests only revealed anemia (hemoglobin: 10.1 g/dL). An abdominal and pelvic contrast CT scan *(Figure 1)* reported a hypodense image in the cecum and ascending colon measuring 12 × 12 cm, consistent with a colon tumor; no enlarged lymph nodes or evidence of metastatic lesions were reported. A colonoscopy was performed *(Figure 2)*, revealing thickening of the cecum and ascending colon wall and multiple polypoid lesions with a diameter between 1 and 2 cm extending to the hepatic flexure; there was also diffuse thickening of the ascending colon and a 50% stenosis of the lumen. The final report was polyposis syndrome with an associated

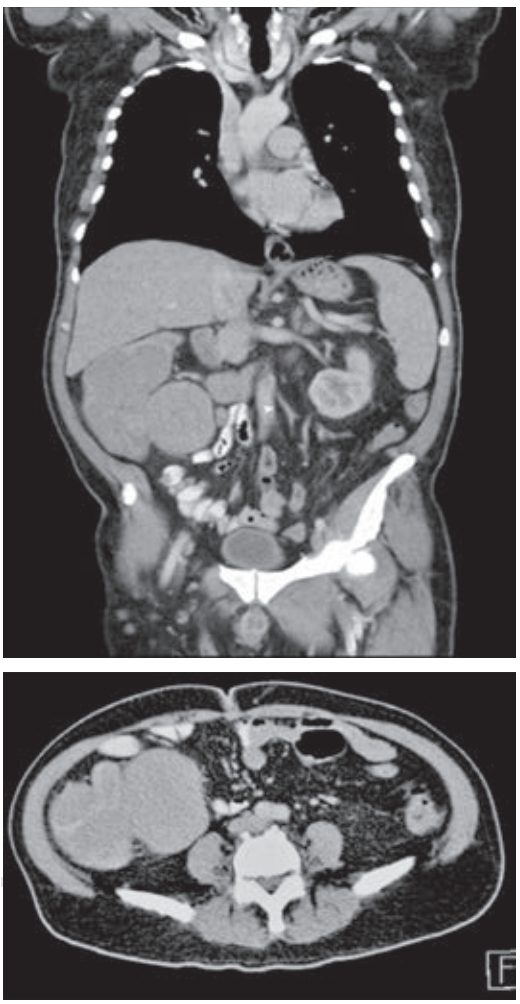

Figure 1: *Contrast-enhanced abdominopelvic CT scan showing hypodense 12 x 12-cm image in the cecum and ascending colon.*

neoplasia of the colon. Multiple biopsies of the tumor and the largest polyps were obtained. The definitive histopathological diagnosis was CD20 positive Bcell lymphoma of the colon with a component of multiple lymphomatous polyposis *(Figure 3)*. The patient was scheduled for surgery. The patient underwent an open extended right hemicolectomy, and an ileotransverse laterolateral anastomosis with a linear stapler was performed. The patient's course was satisfactory and he was discharged on the sixth postoperative day. He was followed in the Oncology Clinic.

DISCUSSION

The World Health Organization (WHO) defines MALT lymphoma as an is an extranodal lymphoma comprising morphologically heterogeneous small B-cells including cells from the marginal zone (centrocyte-like), cells resembling monocytoid cells, small lymphocytes, and scattered immunoblasts and centroblast-like cells.[1] The most common site is the stomach. MALT lymphomas of the colon are extremely rare, and their presentation as multiple lymphomatous polyposis (MLP) is even more unusual.

Multiple lymphomatous polyposis was first described by Cornes in 1961.[3] Several cases of a form of MLP in mantle cell lymphoma have been reported, and the largest series described the followup of 31 patients with MLP.[4] Its frequency is estimated at 9% of all primary gastrointestinal lymphomas; it is more frequent in males, and the average age at diagnosis is 63 years. The involvement of many segments of the gastrointestinal tract with no clinical symptoms is unusual. The main sign reported is gastrointestinal bleeding.[4,5] In the largest series published (31 cases), by RuskonéFourmestraux et al., all cases had digestive signs or symptoms. The main initial symptoms are abdominal pain, diarrhea and lower gastrointestinal bleeding. Obstructive tumors leading to surgery are uncommon. Involvement of the colon and small intestine is highly prevalent (90% of cases), followed by the rectum (60% of cases) and the stomach (50% of cases).[6] Esophageal involvement is exceptional. Invasion of the mesenteric lymph nodes is frequently found.[6]

The radiographic and endoscopic images of patients with MLP reveal characteristic smooth sessile polyps, from 2 mm to several centimeters in diameter and located in one or more intestinal segments.[5,6]

Colonoscopy plays an essential role in the detection of polyps in the colon, as well as for obtaining biopsies. Differentiating lymphomatous polyposis from adenomatous or hamartomatous polyposis based only on colonoscopic and radiological findings is difficult, and they can be confused with one another. Histopathologic analysis is the gold standard in these cases. Consequently, the biopsy-over-biopsy technique during colonoscopy is always preferable and recommended. Histomorphologic evaluation of the biopsy should be performed by an expert pathologist.[7] Histological similarities may be found between MLP and MALT lymphoma, particularly when there is diffuse cellular proliferation, so immunohistochemical analysis is required.[8] Tumor cells in MLP express B cell markers (such as CD19, CD20, and CD22) and the T cell marker CD5.[9] This antigen is normally

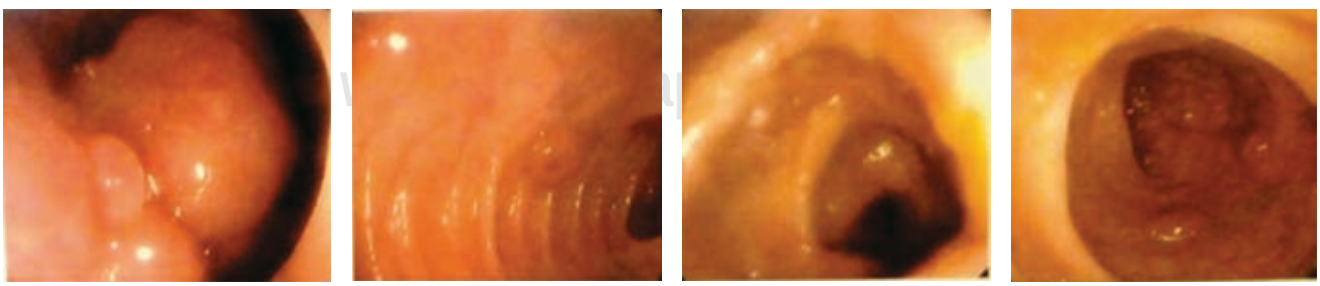

Figure 2: *Colonoscopy reporting thickened wall in the cecum and ascending colon, with multiple polypoid lesions.*

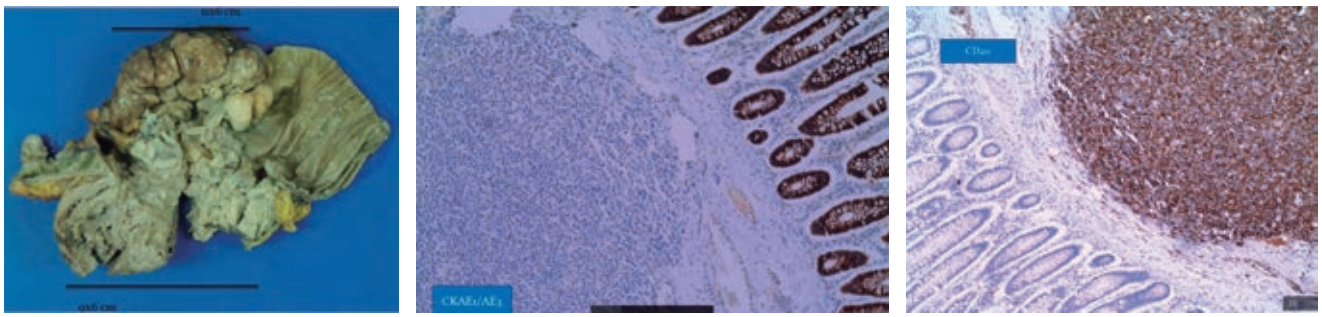

Figure 3: *Biopsies of the tumor and largest polyps: CD20-positive B-cell lymphoma of the colon with a component of multiple lymphomatous polyposis.*

expressed by a minor subpopulation of B cells in the adult's follicular mantle of lymph nodes.[3,10] The absence of T cell markers such as CD3 is common in associated MLP. These morphologic and immunohistochemical properties suggest that MLP originates in the mantle cell zone.[11]

CONCLUSION

Primary lymphoma of the colon (PLC) is an infrequent cause of lymphoma of the colon. Furthermore, the MLP variety as a subtype of B-cell lymphoma of the colon —as in the clinical case reported here— is even more unusual (0.1-0.5%). This is a clinical kind of lymphoma with welldefined histological characteristics. The technique of biopsy over biopsy during endoscopy is important in order to obtain an adequate tissue sample for histopathological differentiation.

In patients with primary lymphoma of the colon of the MLP variety, the prognosis is poor. As in this case, a precise diagnosis is essential before the patient is taken to surgery. Surgical treatment of MLP is mandatory. Considering the location and distribution throughout the colon, segmental resection and anastomosis of the colon can be planned; in some cases of disseminated polyposis, total colectomy should be performed, as well as proctocolectomy if there are polyps in the rectum. Adjuvant chemotherapy is recommended in most cases. It includes regimens with cyclophosphamide, vincristine and prednisone or doxorubicin, teniposide and cyclophosphamide. Most patients with mantle cell lymphoma respond well to chemotherapy initially, but some may become refractory.

Mean survival depends on the tumor's stage, degree of invasion and extension: on average, it ranges between 20 months and 5 years. Cases of complete cure have been reported, if treated in the early stages of the disease.[3,11]

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