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clinical practice

Multiple lymphomatous polyposis with diffuse involvement of the gastrointestinal tract. Case report

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SUMMARY: **Multiple lymphomatous polyposis with diffuse involvement of the gastrointestinal tract. Case report.**

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The gastrointestinal tract is the predominant site of extranodal non-Hodgkin lymphomas. Multiple lynphomatous polyposis is a type of appearance of mantle cell lymphoma. It is characterized by multiple polypoid lesions involving long gastrointestinal tracts and it accounts for only approximately 1-2% of non-Hodgkin lymphomas.

A 78 years old patient was admitted to our Department of General Surgery with rectal bleeding, abdominal pain and weight loss. Multiple lymphomatous polyposis was detected by endoscopy. Endoscopic biopsies confirmed the diagnosis of mantle cell lymphoma. The patient was transferred to the Department of Hematology for cycles of chemotherapy.

KEY WORDS: Multiple lymphomatous lymphoma - Mantle cell lymphoma - Polyposis.

Introduction

Primary non-Hodgkin's lymphoma (NHL) of the gastrointestinal (GI) tract is the most common extranodal NHL and accounts for 4%-20% of all NHL (1). Mantle cell lymphoma (MCL) includes 2.5-7 % of all NHL (2). Multiple lymphomatous polyposis (MLP) is an uncommon disease that is regarded as the gastrointestinal form of MCL (3). Morphological and immunohistochemical studies are essential for diagnosis of MLP. In fact tumor cells, typically, express CD20 or CD5 and cyclin D1 markers in these conditions (4).

Most MLP cases occur in elderly patients, usually over fifty years old, and presenting clinical events like abdominal pain, melena or hematochezia and weight loss. Any part of GI tract may be involved, but diffuse GI involvement of MCL is an uncommon situation (5). Hereby we report and discusse a case of a MLP with a diffuse gastrointestinal involvement, including stomach, ileum, colon, lombo-aortic lymph nodes and liver metastasis.

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Case report

A 78 years-old man presented at our Department of General Surgery with a 1 month-history of rectal bleeding, tenesmus, abdominal pain and recent weight loss (7 kg by 4 months). Physical examination didn't reveal palpable masses in abdomen. Laboratory data included the following: hemoglobin 13.1 g/dl, hematocrit 41.9%, white blood cell count 9300 / microL (neutrophils 71.1%, lymphocytes 20.3%), beta-2-microglobulin 3841 ng/mL. Other oncologic markers, i.e. Alfa-Fetal Protein (AFP), Carcino-Embrional Antigen (CEA), Carbohydrate Antigen 19-9 (CA 19-9) and Prostatic Specific Antigen (PSA) were in normal range of values.

At colonoscopy there were innumerable polyps (from 0.5 to 3 cm) extending from the rectum (Fig. 1) to the cecum (Fig. 2), in spite of a colonoscopy that identified any lesions two years before. Upper GI endoscopy disclosed multiple small, sessile polyps in the stomach and in the duodenum.

Histopathologic study of endoscopic biopsies from the colon and the stomach showed an uniform lymphoid infiltrate. Immunoistochemical evaluation confirmed that the lymphoma was positive for CD20 and cyclin D1, while it was negative for CD10, CD23 and CD5, features compatible with diagnosis of non-Hodgkin mantle lymphoma. Bone narrow biopsy was negative for lymphoid proliferation.

Abdominal and pelvis TC revealed anomalies of the rectum wall, pelvic lymphadenopaty and two liver metastasis. Abdominal and pelvis RM imaging showed lombo-aortic lymphadenopaty and massive thickening of the rectum. Total body PET-TC detected rachis involvement, specifically at D1, L2 and L3 vertebrae, and an intense signal of the whole ileum.

The patient was transferred to Department of Haematology to plan chemotherapeutic strategy.

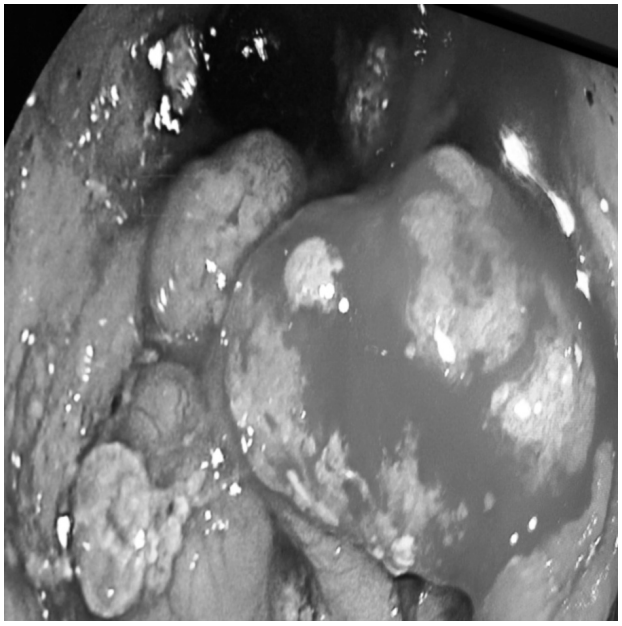

Fig. 1 - Polyposis of the rectum.

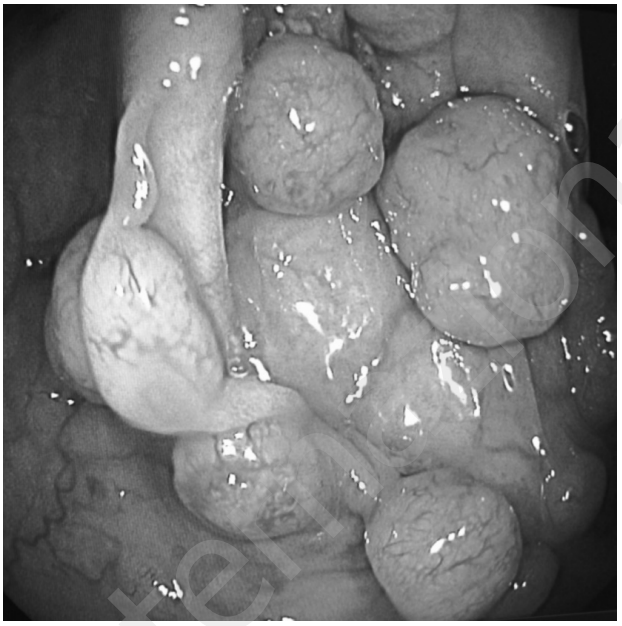

Fig. 2 - Polyposis of the cecum.

Discussion and conclusion

The gut mucosa contains more lymphocytes than the other immune system organs. Nevertheless, only 10% of all lymphomas present in the gut (5). Most of primary GI NHL occur with single lesion, which more frequently involves the stomach and small intestine. Single colorectal lymphomas are relatively rare (6). MCL is an uncommon type of primary GI NHL with particular clinic, morphological and immunophenotypic features. Clinically, patients with a diagnosis of MCL are often elderly adults with a male predominance and present a disease in an advanced stage (7). Abdominal pain, diarrhea, hematochezia and organomegaly are the most common presenting symptoms (8). Liver and spleen involvement are common clinical features too (9). The incidence of GI tract involvement ranged from 10% to 28% in various series (10). The typical endoscopic findings of early MCL is the appearance of MLP with innumerable, small, spherical or hemispherical polyps (11). On the other hand, when an advanced MLP is identified we can classify endoscopic features in three types: 1) elevation type; 2) diffuse infiltration type and 3) ulceration type (12). The diagnosis is finally based on peculiar immunoistochemical findings. MCL cells can express B cell markers including CD19, CD20, CD22, as well as T cell marker such as CD5, and cyclin D1. The classic cytogenetic t (11, 14) translocation with subsequent overexpression of cyclin D1 protein, caused by the translocation of the cyclin D1 gene to the promoter of the immunoglobulin heavy chain, are diagnostic for MCL (13).

In the series of MLP cases by Ruskone-Fourmestraux et al., the colon and the rectum were affected in about 90% of cases, followed by the small bowel, stomach and duodenum (14). The prognosis of GI MCL is poor with a mean survival time of less than three years. COP (Cyclophosphamide, Doxorubicin, Prednisolone), anthracycline-containing regimens and CHOP (Cyclophosphamide, Doxorubicin, Vincristine and Prednisolone) are used as usual chemiotherapies for MCL (15). However, a variety of new molecules, including bortezomib, represent the first drug specifically approved for this type of lymphoma, and others substances such as bendamusine, mTOR inhibitors and lenalidomide have also shown a specific activity against MCL cells (16). Infiltrating lymphoma cells expressed CD20 antigen on their surfaces. Rituximab is a chimeric antibody binds specifically to the CD20 (17). Because this lymphoma occurs in the elderly population, stem cell transplantation is not possible.

In conclusion, GI lymphomatous polyposis is a rare disease, but in all elderly patients with a widespread polyposis of the GI tract a diagnosis of lymphoma must be considered.

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