

# Multiple Lymphomatous Polyposis Associated with Small Lymphocytic Lymphoma: a Unique Presentation

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## Abstract

Multiple lymphomatous polyposis (MLP) is a rare extra-nodal manifestation of lymphoma. In most cases, MLP is associated with mantle cell lymphoma (MCL). We report a 66-year-old male diagnosed with small lymphocytic lymphoma (SLL)/chronic lymphocytic lymphoma (CLL), who showed evidence of rectal bleeding. A CT-scan of the abdomen and pelvis showed an enlarged spleen, multiple paraaortic and mesenteric lymph nodes, and some diverticular pouching along the antimesenteric border of the pelvic colon. A colonoscopy revealed the presence of multiple polypoid lesions, biopsies of which showed diffuse lymphoid infiltrate without any identifiable follicles. Immunohistochemical analysis combined with a Fluorescence In-Situ Hybridization (FISH) study excluded the diagnosis of MCL. A bone marrow aspiration biopsy demonstrated diffuse infiltration of the bone marrow with low grade lymphocytes that expressed CD 20, CD5 and CD23, with negative BCL-1, t(11; 14) and cyclin D1. A diagnosis of B-cell CLL with kappa light chain restriction was made. Multiple lymphomatous polyposis is considered to be a digestive counterpart to MCL and can therefore be considered as a presentation of MCL. In our case, the polypoid lesions failed to show the characteristic features of MCL. The patient's bone marrow revealed a B-cell lymphoma of CLL/SLL phenotype, which to our knowledge has not been linked to MLP in previously reported cases.

## Key words

Multiple lymphomatous polyposis – mantle cell lymphoma - chronic lymphocytic lymphoma.

## Introduction

Multiple lymphomatous polyposis (MLP) is an extra-nodal involvement of lymphoma, which is characterized by multiple polypoid tumors which affect several parts of the gastrointestinal system, as described for the first time by Cornes in 1961 [1]. In most of the cases, malignant cells of MLP display mantle cell characteristics and are thus recognized as digestive counterparts to mantle cell lymphoma (MCL), which accounts for 20% of all MCL cases [2]. In rare cases MLP can be the only presentation of MCL as we reported before [3]. In this report we are presenting a case of MLP that is associated with small lymphocytic lymphoma (SLL).

## Case report

A 66 year old male, of Mediterranean origin with a medical history of diet controlled hypertension, was referred to us due to abnormal findings in a colonoscopy. Initially, the patient had complained of having progressive symptoms of rectal bleeding. Physical examination of the patient revealed no lymphadenopathy, palpable spleen or any other significant findings. Laboratory testing which was performed showed mild anemia (Hb 12.2 g/dL). A peripheral smear illustrated normal morphology of RBCs, WBCs and platelets. A CT-scan of the abdomen and pelvic regions showed a mildly enlarged spleen and multiple variably sized enlarged discrete paraaortic and mesenteric lymph nodes, as well as few small sized diverticular pouching along the anti-mesenteric border of the pelvic colon. Consequently, the patient was referred for a colonoscopy that revealed multiple polypoid lesions at 6, 10 and 15 cm from the anus. These polypoid lesions varied in size from 0.8 to 1.2 cm; some were very friable and bleeding.

Multiple biopsies were taken, and pathological examination revealed diffuse lymphoid infiltrate without identifiable follicles. The lymphocytes were predominantly small, with rare large cells that had centrally located nucleoli (Fig. 1). Immunohistochemical analysis showed predominant B-cells with co-expression of CD5, CD43, CD20, CD45

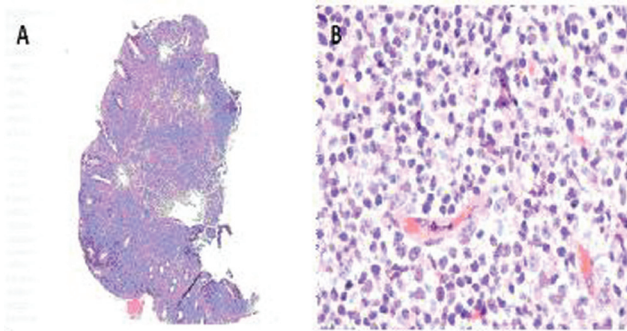
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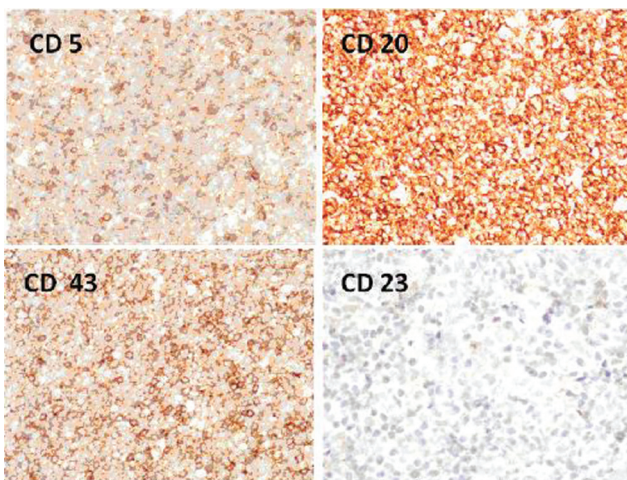
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**Fig 1.** Pathologic examination of a polyp: A) low power; B) high power.



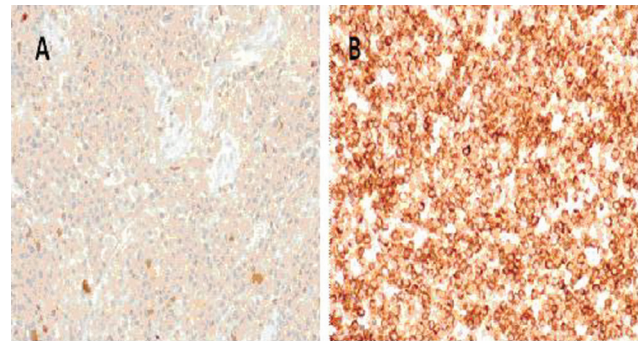
**Fig 2.** Immunohistochemical staining.

and CD79a. CD23 (Fig. 2) was positive in a small subset of the B-cells and was reported as negative. Staining for BCL-1 was negative while staining was positive for BCL-2 (Fig. 3). Fluorescence In-Situ Hybridization (FISH) study for t(11; 14) showed no evidence of t(11; 14) or trisomy 11. PET/CT demonstrated a mild enlargement of spleen (not palpable clinically) with normal FDG distribution, numerous mesenteric lymph nodes on CT component that were not FDG avid on PET component, and no evidence of retroperitoneal or pelvic lymphadenopathy.

The patient also underwent a bone marrow aspiration and biopsy which were subsequently analyzed by flow cytometry. Diffuse infiltration of the bone marrow with low grade CD 20, CD5 positive and CD23 positive lymphocytes was evident along with negative BCL-1, cyclin D1 and t(11;14) with immunoglobulin on the surface of the cells. A suggested diagnosis of B-cell chronic lymphocytic lymphoma with kappa light chain restriction was made based on the result of the bone marrow. The patient was therefore offered Fludarabine containing chemotherapy; however, he refused therapy at that time and decided to follow up for possible chemotherapy later.

## Discussion

Extranodal involvement of the gastrointestinal system with non-Hodgkin lymphoma is not uncommon. As reported



**Fig 3.** A) BCL-1; B) BCL-2

earlier by Freeman [4], about 35% of extranodal involvement takes place in the gastrointestinal system, mostly in the stomach followed by the small intestine and to a lesser extent the colorectal area (less than 6%). Multiple lymphomatous polyposis is considered to be a rare condition and is thought to be a digestive counterpart to MCL due to similarities found in the pathological and histological features of both conditions [1, 2, 5, 6]. In fact, we had reported earlier a case of MLP as a sole presentation of mantle cell lymphoma [3]. In another article, Weisenburger and Armitage indicate that multiple lymphomatous polyposis of the intestine should suggest a diagnosis of MCL [7]. However, other authors report instances in which MLP is associated with other forms of lymphoma such as MALT or follicular lymphoma [8, 9].

In our unique case, the polypoid lesions, present mostly in the rectosigmoid region failed to show the characteristic features of MCL [this was excluded by the negative t(11:14) and negative BCL-1; however cyclin D1 was not available on the polypoid tissue but was negative on the bone marrow biopsy]. Additionally, the bone marrow revealed a B-cell lymphoma of chronic lymphocytic lymphoma (CLL)/small lymphocytic lymphoma (SLL) phenotype, which to our knowledge has not been linked to MLP in previously reported cases. CD23 demonstrated strong positivity in bone marrow aspirate, and a loss in its positivity was noted in the polypoid malignant cells. This unique presentation coupled with data from previous reports indicates that certain markers need to be present in order to clearly identify the manifestation of gastrointestinal lymphoma. At this time more information is required for any conclusive statement to be made.

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