Mantle Cell Lymphoma Presenting as Multiple Lymphomatous Polyposis of the Colon

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Abstract

Mantle cell lymphoma (MCL) is a subset of B-cell non-Hodgkin's lymphoma with a tendency to involve the gastrointestinal (GI) tract and presents as multiple lymphomatous polyposis of one or multiple segments of the GI tract. Here, we report an unusual case of a 58-year-old female presenting with chief complaints of constipation, bleed per rectum, and unintentional weight loss of 15 kg over 6 months. Colonoscopy revealed the entire colon to be studded with polypoidal lesions of varying sizes which were biopsied. Histopathological examination and immunohistochemistry confirmed the polypoidal lesions to represent MCL. Computed tomography scan showed numerous variable-sized polypoidal lesions scattered throughout the entire colon along with paraaortic and mesenteric lymphadenopathy. Thus far, she has undergone three cycles of chemotherapy with R-CHOP resulting in significant tumor reduction and symptomatic improvement in her well-being.

Keywords
- mantle cell lymphoma
- multiple lymphomatous polyposis
- non-Hodgkin's lymphoma

Introduction

Mantle cell lymphoma (MCL) is a subtype of B-cell non-Hodgkin's lymphoma (NHL) and comprises 2 to 7% of all NHL characterized by chromosomal translocation t (11;14) (q13; q33) with resultant overexpression of cyclin D1. The gastrointestinal (GI) tract is one of the most common extranodal sites and accounts for 5 to 20% of all extranodal involvement[1] and typically manifests as multiple lymphomatous polyposis (MLP).[2,3] MLP most commonly occurs in the ascending colon and the small bowel, particularly in the ileum and ileocecal region and is characterized by multiple sessile polypoidal lesions, sometimes the entire GI tract being carpeted with numerous polyps.

Case Report

A 58-year-old female with no pervious comorbidities presented with complaints of abdominal pain of 6-month duration with unintentional weight loss of around 10 kg, bleed per rectum, and night sweats. On physical examination, she had pallor. Per abdomen exam showed a nontender mass of about 10 × 8 cm in the left iliac fossa. Her spleen was palpable, and per rectal exam showed a firm nodularity.

Her laboratory workup was significant for microcytic hypochromic anemia with a hemoglobin of 8.9 g/dL and an erythrocyte sedimentation rate of 96. Her lactate dehydrogenase was elevated at 389 U/L. Ultrasound of the abdomen/pelvis showed a large lobulated hypoechoic lesion (12.5 × 13 × 8 cm) in the suprapubic region and left iliac region extending into the pelvis. Also noted were multiple paraaortic and paracaval lymph nodes with a spleen measuring 14 cm. The findings on computed tomography abdomen and pelvis were consistent with lymphoma involving terminal ileum with multiple polyps of the large bowel loops, omental lesions, and wall thickening of ileal loops (►**Fig. 1**).

Colonoscopy (►**Fig. 2**) demonstrated the entire colon to be carpeted with multiple polypoidal lesions of varying sizes (0.5–2 cm). Histopathology of the colonic polyps showed diffuse infiltration of the colon by small- to medium-sized lymphocytes (►**Fig. 3**). Immunohistochemistry (IHC) showed the lymphoid cells to be uniformly positive for B-cell marker CD20

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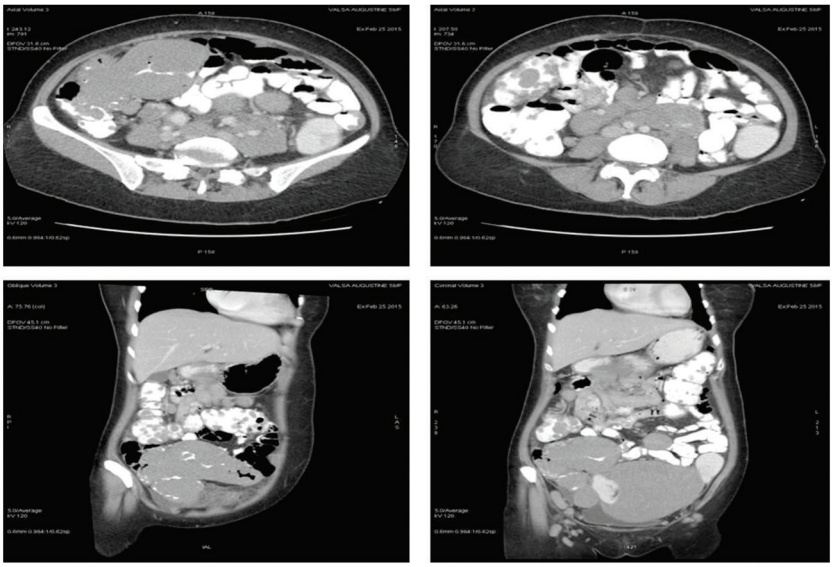

Fig. 1 CT abdomen and pelvis showing circumferential irregular soft tissue mass involving long segments of the distal ileum with aneurysmal dilatation of the lumen. Numerous variable sized polypoidal lesions noted diffusely scattered in the right colon, transverse colon, and proximal descending colon. Also, hepatomegaly and borderline splenomegaly with extensive discrete paraaortic and mesenteric lymph nodes noted.

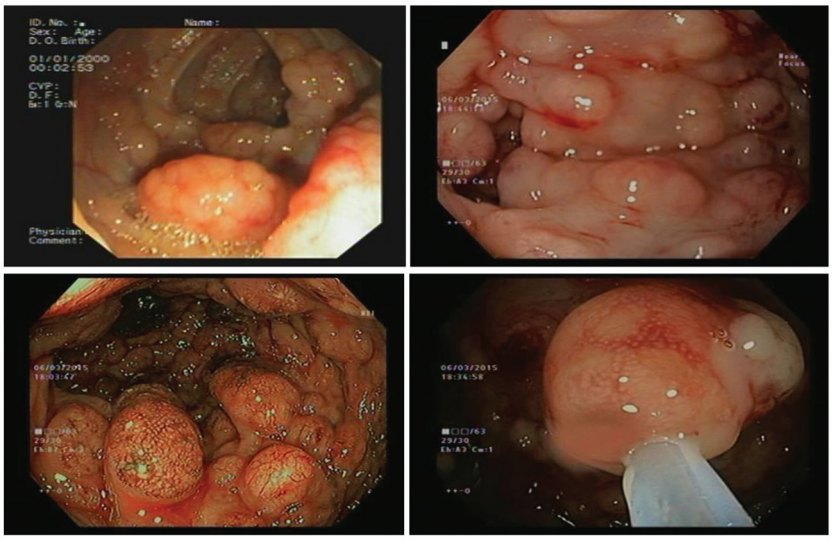

Fig. 2 Colonoscopy showed the entire large bowel to be carpetted with multiple polypoidal lesions of varying sizes. On snare polypectomy, cheesy white material was noted exuding from the polyps.

and for Bcl2 and cyclin D1 which confirmed the diagnosis of MCL. Ki67 index, which is a proliferative index, was 20 to 25% (►**Fig. 4**).

The patient was initiated on chemotherapy consisting of R-CHOP regimen (rituximab, cyclophosphamide, doxorubicin, vincristine, and prednisone) which resulted in remarkable tumor regression and amelioration of her symptoms.

Discussion

Primary GI lymphomas are rare and can involve any segment of the GI tract from the oropharynx to the rectum. MCL is a subtype of B-cell lymphoma typically manifesting as MLP. MLP, first described by Cornes in 1961,[2] is characterized by multiple polypoidal lesions consisting of uniform infiltrates of malignant B cells. Patients with MLP typically present with abdominal pain, hematochezia, diarrhea, weight loss, and night sweats.[4] Diagnosis of this condition requires a combination of endoscopic findings with histopathological analysis and IHC. The typical endoscopic features are nodular or polypoid lesions ranging from 2 to 10 mm in diameter. Immunophenotypic analysis is essential to distinguish MCL from other lymphomas and shows positivity for cyclin D1, CD20, CD79a, and CD5 and negativity for BCL16, CD23, and CD10. In general, patients with MCL tend to have a poorer outcome than those with

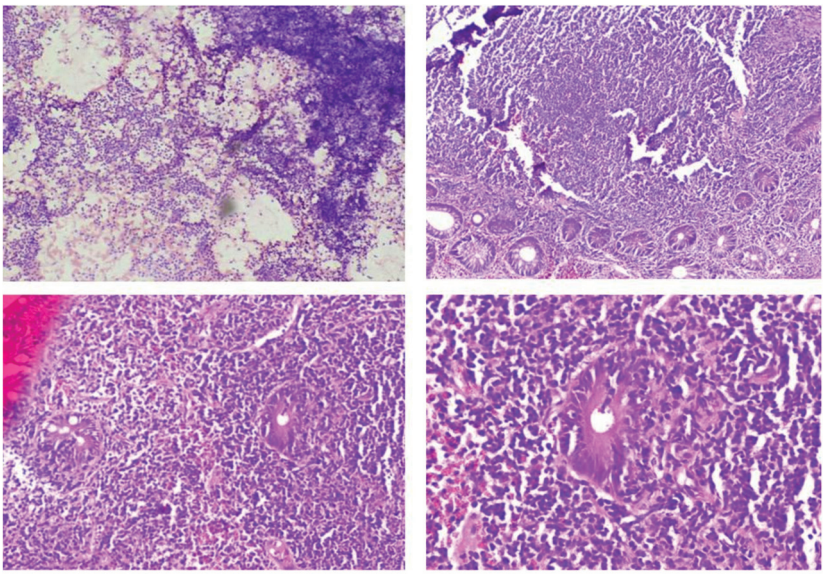

Fig. 3 Patchy eosinophil rich inflammation with widely spaced crypts intervened by sheets of lymphoid cells with several lymphoepithelial lesions.

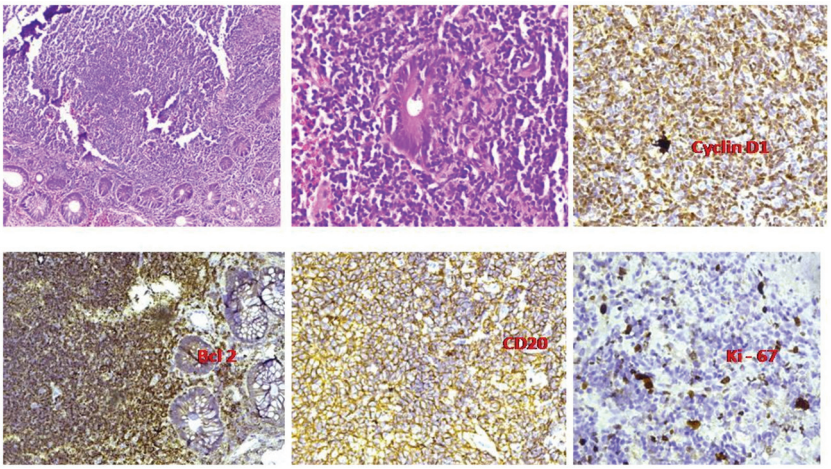

Fig. 4 Immunohistochemistry of the tumor cells showing cyclin D1, CD20, and Bcl2 positivity.

other low-grade NHL with a 5-year survival rate of around 11%.[5] The current treatment regimen recommended for MCL is R-CHOP induction regimen and high dose of cytarabine followed by high-dose consolidation and autologous stem cell transplant.[6]

In conclusion, although GI lymphomas are rare entities, MCL should be included in the differential diagnosis of patients presenting with endoscopic findings of multiple polypoidal lesions. A combination of endoscopic, imaging, and histopathologic examination with IHC clinches the diagnosis of MCL.

Authors' Contributions

All the authors were equally involved in the conception and design; analysis and interpretation of the data; drafting of the article; critical revision of the article for important intellectual content; and final approval of the article.

Conflict of Interest

None declared.

Acknowledgments

None.

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