

CASE REPORT

Mantle Cell Lymphoma of Intestine Presenting as Multiple Lymphomatous Polyposis with Intussusception

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Abstract:

Mantle Cell Lymphoma (MCL) is a distinct clinicopathological subtype of B-cell non-Hodgkin's lymphoma (NHL) accounting for 2-10% of all NHL cases. Gastrointestinal tract (GIT) is the predominant site of extranodal MCL which commonly presents as Multiple Lymphomatous Polyposis (MLP). A 60 year old male presented with pain abdomen, diarrhea and weight loss of two months duration. On colonoscopy multiple polyps were found in the entire colon and rectum. Computed tomography revealed ileo-colic intussusception with nodularity in the lead point. Histopathology suggested features of MCL. On immunohistochemistry, the tumor cells were positive for CD20, CD5, Cyclin D1, negative for CD3, CD10, CD23, and CD45 RO.

Keywords: Intussusception, Mantle Cell Lymphoma, Polyposis.

Introduction:

The gastrointestinal tract (GIT) is the most common site of extranodal non-Hodgkin's lymphoma (NHL) [1, 2]. It accounts for 4-20% of all NHL cases [3]. Mantle cell lymphoma (MCL) is the subtype of B-cell NHL comprising 2-10% of NHL cases [4]. The GIT involvement in MCL in various studies is 10-28% [5]. MCL in GIT may occur as primary or as part of generalized presentation. It commonly manifests in the form of primary lymphoma as multiple lymphomatous polyposis (MLP) [3, 5].

The exact incidence of tumour or multiple polyps causing intussusception is not known. There are very few cases of MCL in the literature presenting as MLP with intussusception [6].

Case Report:

A 60 year old male presented with pain abdomen, diarrhea and weight loss of two months duration. He had similar complaints one year back and was diagnosed as NHL suggestive of MCL on colonic biopsy. Patient was not put on any treatment. Physical examination showed pallor, clubbing of fingers and enlarged right axillary lymph node measuring 2x1cm. Oral cavity was unremarkable. Per abdominal examination showed a firm mass in the right hypochondrium moving with respiration measuring 14x6cm. There was no hepatosplenomegaly.

Laboratory investigations revealed normal blood cell counts and microcytic hypochromic anemia without any atypical lymphocytes. Chest X-ray was suggestive of chronic obstructive pulmonary disease. Colonoscopy showed multiple polyps in the entire colon and rectum. Upper endoscopy was unremarkable. Computed tomography of abdomen revealed diffuse circumferential wall thickening of ascending colon and ileo-colic intussusception with nodularity in the lead point (Fig.1).

At operation ileo-colic intussusception was seen with pre aortic, para aortic and mesenteric lymphadenopathy. Intussusception was reduced. Total colectomy with ileo-rectal anastomosis was done along with excision of right axillary lymphnode. The total colectomy was sent in two segments, the larger colonic and smaller ileal, totally measuring 79cm in length. Both segments showed dominant polyps each measuring 8x7cm and 5x4cm respectively along with multiple

sessile polyps varying in size from 0.2 to 2cm (Fig. 2). Mesocolon showed 15 enlarged lymphnodes. Microscopy of intestinal polyps showed monotonous population of atypical lymphocytes in diffuse and nodular pattern extending from mucosa to muscularis propria. These cells were ovoid with scanty cytoplasm. The nuclei were indented showed condensed chromatin with indistinct nucleoli. Mitotic count was 5 to 7 per 10 HPF. Macrophages were seen at places. Blood vessels were thickened. Occasional large cells were also seen. There were no lymphoepithelial lesions. Sections from mesenteric and axillary lymph nodes showed effacement of architecture by nodules of lymphocytes that were morphologically similar to those of intestine (Fig 3).

On immunohistochemistry the cells showed immunopositivity for CD20, bcl-2, CD5, Cyclin D1 and were negative for CD68 (Fig 4).

Focal positivity for CD23 in residual germinal centers was noted. They were negative for CD45RO, CD3 and CD10 (Fig 5). The axillary lymphnode also showed similar immunohistochemical findings.

Bone marrow aspiration showed no infiltration by lymphoma cells. A diagnosis of MCL of intestine with ileo-colic intussusception due to MLP and involvement of mesenteric and axillary lymphnodes stage IV (Ann Arbor System) was made. The patient was put on combined chemotherapy but he expired due to sepsis and pneumonia 20 days after operation.

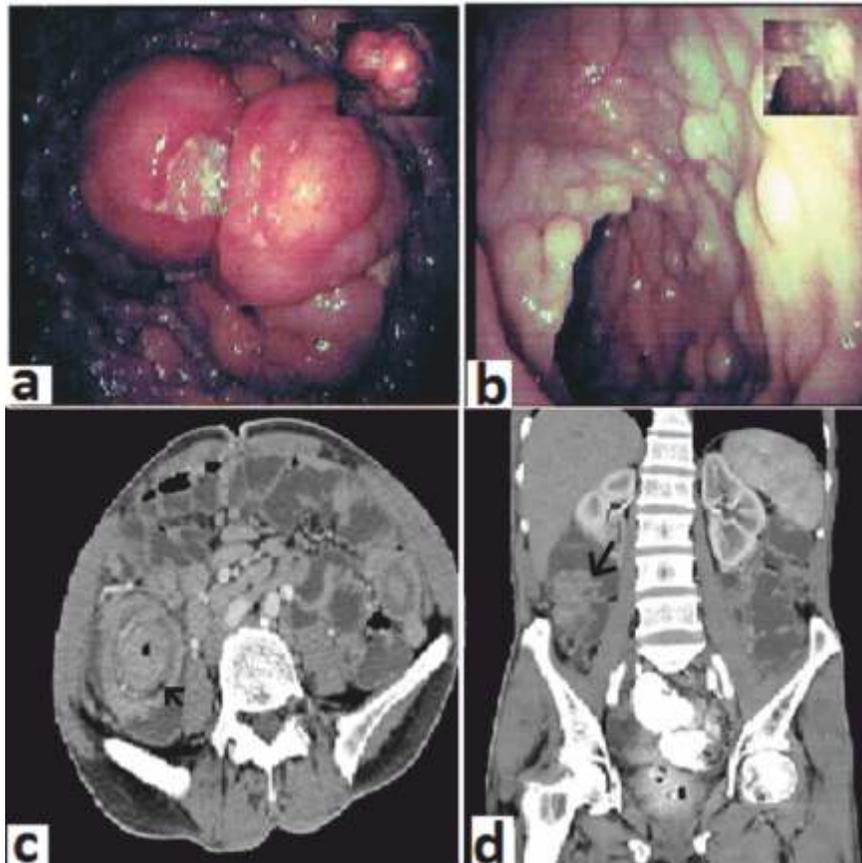


Fig. 1: Colonoscopic View of Polyps in Ascending (a) Descending Colon (b). CT Abdomen Showing Intussusception (Arrow) In Axial View (c) and Nodularity in the Lead Point (Arrow) In Coronal View (d)



Fig. 2: Specimen Showing Polyps in Colon (a) and Terminal Ileum (b).

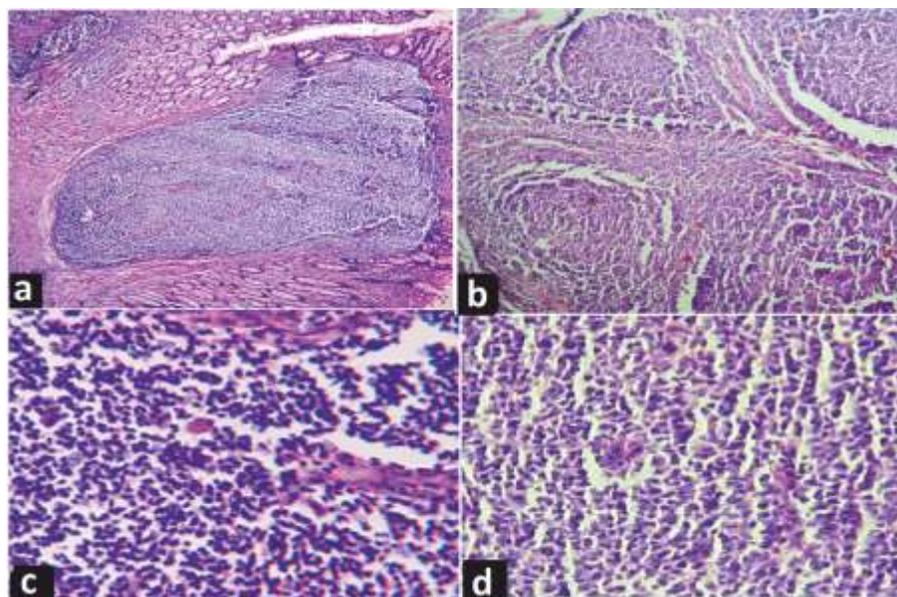


Fig. 3: Microphotograph Showing Tumor Cells in Colonic Mucosa (a) and Axillary Lymph Node (b) (H & E, X 100). Monotonous Population of Atypical Lymphocytes in Colon (c) and Lymph Node (d) (H & E, X400)

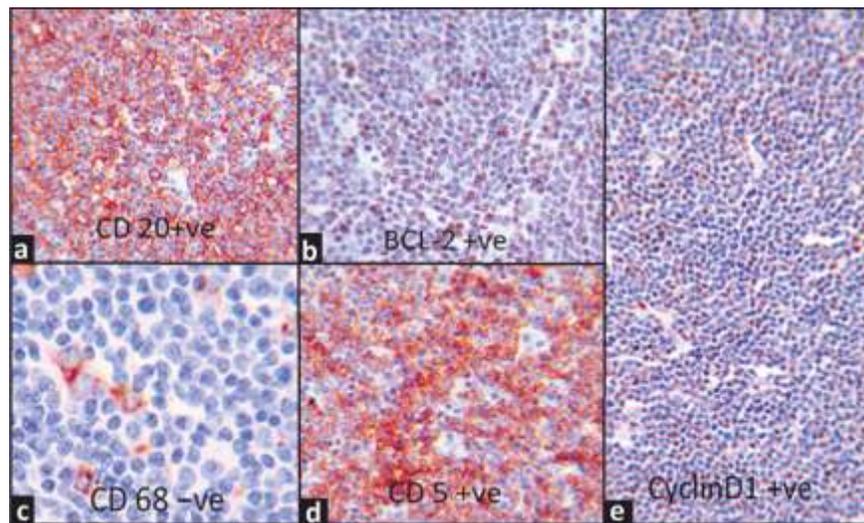


Fig. 4: Immunohistochemistry Showing Lymphoma Cells in Colon Positive for CD20 (a), BCL-2 (b), Negative for CD68 (c), Positive for CD5 (d) and Cyclin D1 (e) (X400)

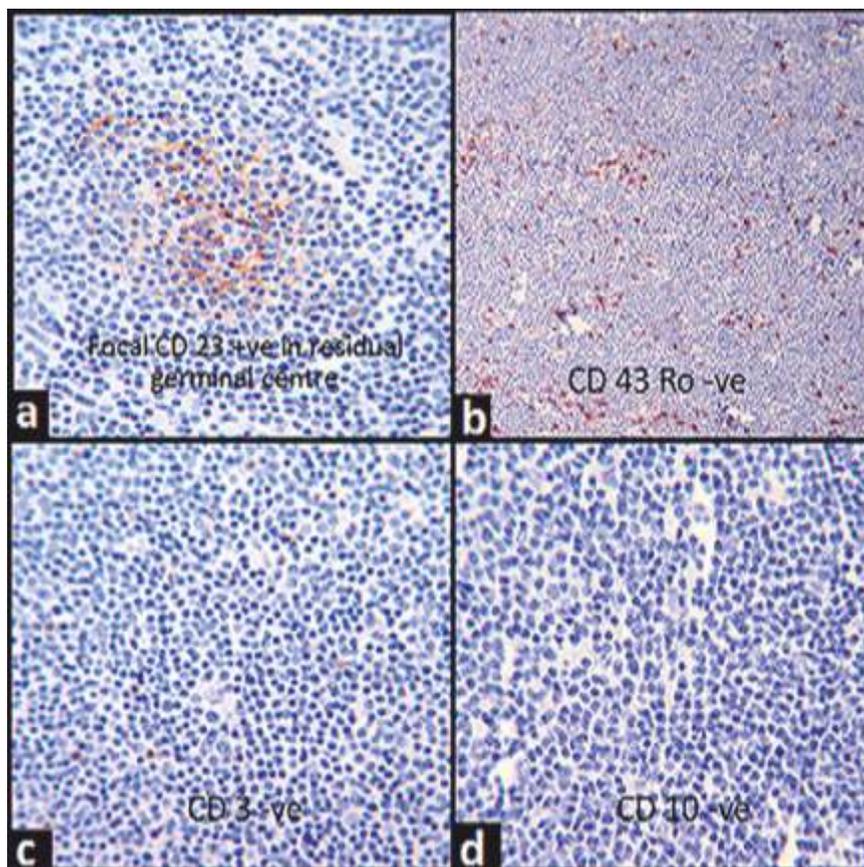


Fig. 5: Immunohistochemistry Showing Lymphoma Cells in Colon Negative for CD23 (a), CD45 RO, (b) CD3 and CD10 (d) (X400)

Discussion:

MLP accounts for 9% of primary GIT lymphomas [2]. Although first recognized in the 19th century by Briquet, MLP was first described by Cornes in 1961 as primary malignant lymphoid tumour of varying histologic types growing as sessile or pedunculated polyps over considerable segments of GIT [7]. Some authors consider it to be specific for MCL [3, 6] but many studies have shown that it is heterogenous entity [1, 2, 7, and 8]. Follicular lymphoma (FL), Mucosa Associated Lymphoid Tissue (MALT) lymphoma, B-cell chronic lymphocytic leukemia and adult-T cell lymphoma show similar gross features and are to be considered in differential diagnosis [2].

MCL is typically seen in adults with male predominance [2]. Bone marrow is involved in more than 50% of the cases at the time of diagnosis [5, 6]. Patients with MLP usually show a dominant mass in ileocecal region [7]. Rarely may it cause intussusceptions [6]. In our case the large ileal mass was the lead point causing ileo-colic intussusception. As the invagination was not tight there was no ischaemia. MCL is composed of cells resembling mantle zone of secondary follicles.

In our case the cells expressed CD20, CD5 with cyclin D1 overexpression and thus helped in differentiating from FL and MALT lymphoma. It is frequently characterized by t(11;14)(q13;q32) translocation [3]. Cytogenetic analysis or southern blot analysis to detect bcl-1 rearrangement was not done in our case. The exact etiology of MCL is not known. It is said to be derived from pregerminal center B cells [1, 8]. The characteristic mucosal pattern of MLP results from expression of integrin/homing receptor by lymphoid cells [7]. It is classified as B cell centrocytic lymphoma according to Keil's classification and diffuse small cleaved cell malignant lymphoma by working formulation. MCL has aggressive biological behavior with early systemic dissemination. It has the poorest prognosis of all NHL subtypes with the median failure free survival of 8-20 months [9]. Prognosis is significantly influenced by increased mitosis, blast morphology and leukemic involvement at the time of diagnosis [10]. The case is presented for its rarity and its importance in differentiating from other lesions presenting as MLP and intussusception.

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