Mantle Cell Lymphoma with Multiple Lymphomatous Polyposis Presenting with Intussusceptions

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ABSTRACT
Mantle cell lymphoma is an aggressive type of B-cell non-Hodgkin’s lymphoma that originates from small to medium sized lymphocytes located in the mantle zone of the lymph node. The gastrointestinal tract is the predominant site of extranodal involvement in the form of multiple lymphomatous polyposis. Multiple lymphomatous polyposis due to mantle cell lymphoma presenting with intussusception is uncommon and very few cases have been reported. We are reporting a Mantle cell lymphoma with multiple lymphomatous polyposis presenting with intussusception in a 62 years old male.

Keywords: Mantle-Cell Lymphomas, Intestinal Polyposis, Intususception

Introduction
Primary Non-Hodgkin’s lymphoma (NHL) of the gastrointestinal tract is the most common extranodal NHL and accounts for 4%-20% of all NHL (1). Mantle cell lymphoma (MCL) comprises 5%-10% of all NHL (2, 3). Gastrointestinal tract involvement occurred in 20% of MCL, which is commonly manifested as numerous, small, spherical, or hemispherical polyps, termed multiple lymphomatous polyposis (MLP) (1, 4). Intussusception, in which, the proximal segment of bowel telescopes into the lumen of an adjacent distal segment, can occur anywhere in the gastrointestinal tract. Intussusception is common in children and relatively rare in adults and caused by several factors. MLP presenting with intussusception is very rare and representing only 1% of patients with bowel obstruction. There are only few cases (less than 1%) of MCL causing intussusception has been reported in the literature (5;6).
We hereby report a case of MCL with MLP involving ileum, cecum, ascending colon and mesentery lymph nodes, clinically presented as intussusception and intestinal obstruction.

**Case History**

A 62 year old male presented with pain, tenderness in the right lower abdomen, associated with nausea and non-bilious vomiting. There was no history of fever or weight loss. Physical examination revealed normal vital signs, a soft distended abdomen with hyperactive bowel sounds, and a palpable tender mass in the right lower quadrant. No palpable peripheral lymphadenopathy.

Laboratory investigations showed low hematocrit (27%), normal peripheral leukocyte count and differential. Peripheral smear showed mild microcytic hypochromic anemia and bone marrow aspiration revealed mild erythroid hyperplasia. No evidence of marrow infiltration by leukemia/lymphoma.

A plain abdominal radiograph showed a nonspecific gas pattern in the bowel with fecal loading of the descending and sigmoid colon. No mediastinal lymphadenopathy on a chest radiograph. CT scan of abdomen revealed ileo-colic intussusception. No focal lesions/infiltrates in the liver and spleen.

Right hemicolectomy and extended ileal resection was performed. Postoperative recovery was uneventful.

Gross resected segment of small bowel, cecum, and ascending colon showed ileo-colic intussusception (Fig. 1a,1b) and on cut section revealed multiple sessile and pedunculated polyps of varying sizes ranging from 0.2cm to 2-4cm. in diameter (Fig. 2a). Multiple lymph nodes of varying sizes (0.2cm to 3.5cm) isolated from the mesentery (Fig. 2b). Histological examination of the ileum, colon, mesenteric mass, and lymph nodes showed diffuse infiltration of monotonous small lymphoid cells with irregular nuclei (Fig. 3). The neoplastic lymphoid cells were positive for Cyclin D1 (Fig. 4), CD20 and CD5; negative for CD23, CD10 and the histomorphology, immunophenotypic analysis was consistent with mantle cell lymphoma.

**Fig. 1-** Gross specimen of segment of small intestine, cecum, ascending colon (1a), and ileocolic intussusception (1b).
Mantle cell lymphoma (MCL) is an aggressive type of B-cell non-Hodgkin's lymphoma that originates from small to medium sized lymphocytes located in the mantle zone of the lymph node. It was classified as one of 11 types of B cell lymphoma in the Revised European-American Classification of Lymphoid Neoplasms on the basis of morphologic, immunophenotypic and genetic findings in 1994 (3).

Approximately 5%-10% of lymphomas are classified as MCL (2, 3). The gastrointestinal tract is the predominant site of extranodal involvement in the form of MLP and accounts for approximately about 9% of primary gastrointestinal lymphomas (3). Among 13.7% of primary gastrointestinal lymphomas, 41% are diffuse large B-cell lymphomas, 12% are lymphoblastic lymphomas, and 4% are mantle cell lymphomas.

**Fig. 2** - Cut section showing numerous intraluminal and serosal lymphomatous polyposis (2a) and resected mesenteric lymph nodes (2b).

**Fig. 3** - Histopathological examination of mantle cell lymphoma showing monotonous small lymphoid cells with irregular and slightly angular nuclei (Hematoxylin & Eosin staining ×40).

**Fig. 4** - Immunohistochemistry showing positive nuclear staining for Cyclin D1 (×100 magnification).
lymphoma, 35% are extra nodal marginal zone B-cell lymphoma and 2% are in the form of MLP (1). Kodama T et al. in their study of 35 patients with MLP, found 12 cases as MCL, 14 follicular lymphoma and 9 extranodal marginal zone B-cell lymphoma (7). Furthermore Fraga M et al. reported that MCL was developed without MLP (8). MLP is characterized by multiple polyloid lesions involving long segments of the gastrointestinal tract, ascending colon and the small bowel, particularly in the ileum and ileocecal region. Occasionally, however, numerous polyps are present throughout the entire gastrointestinal tract. A primary manifestation in the stomach is rare. Individual cases of MLP occurring in association with colonic adenocarcinoma have been described (9). Polyps may be sessile, pedunculated or both. They range in size from 0.1 to 2–5 cm and present with ulceration. Mesenteric lymph nodes are most commonly involved. Intussusception is uncommon in adults and is very rarely caused by MCL (5, 6). To our knowledge, there are only few cases (less than <1%) of mantle cell lymphoma causing intussusception has been reported in the literature (5, 6).

MLP can present with symptoms such as abdominal pain, diarrhea, bleeding, and less frequently, protein-losing enteropathy, intestinal malabsorption, or chylous ascites. Rarely, MLP presents as an acute abdomen due to perforation or intestinal obstruction (10, 11).

Risk factors of intestinal lymphomas include diffuse nodular lymphoid hyperplasia with or without primary immunodeficiencies (12), patients with inflammatory bowel disease using immunosuppressive medications (13), male patients with Crohn’s disease (14), congenital and acquired immunodeficiency states, and immunosuppression after solid organ transplantation.

Upper gastrointestinal endoscopy, enteroscopy and colonoscopy are important tools in diagnosing MLP to assess the locations of the polyps and obtain tissue biopsies. Immunohistochemical markers are essential in distinguishing mantle cell from other types of lymphoma. The mantle cell neoplastic cells were positive for Cyclin D1, CD20, CD5 and CD79a, but negative for BCL6, CD23 and CD10. The absence of CD23 is useful to distinguish MCL from small lymphocytic lymphoma and the presence of CD5 useful in the differential diagnosis with follicular and marginal zone lymphomas. Cytogenetic analysis of MCL shows rearrangement of the bcl-1 locus on chromosome 11 due to t [11:14] (q13:q32) translocation, accompanied by cyclin D1 antigen over expression (15).

MCLs usually respond poorly to conventional therapeutic regimens and are associated with short median survival. Median survival with standard treatment for MCL patients remains between 3 and 4 years (16).

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References