Case Report

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). Intussception, 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 intussception 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).

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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.

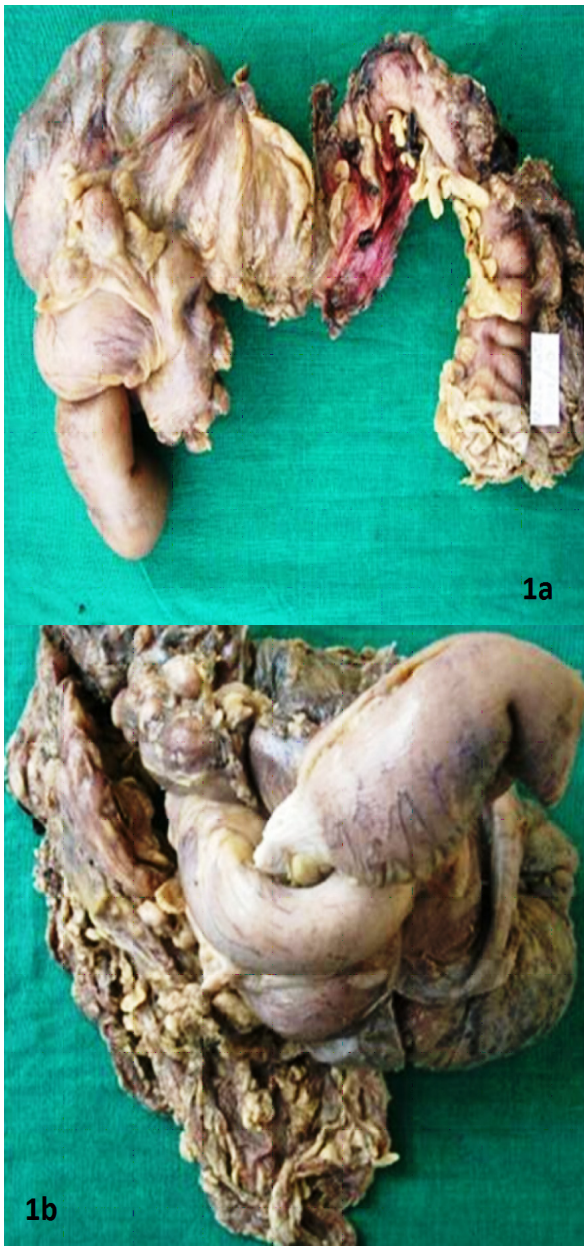

Fig. 1- Gross specimen of segment of small intestine, cecum, ascending colon (1a), and ileocolic intussusception (1b).

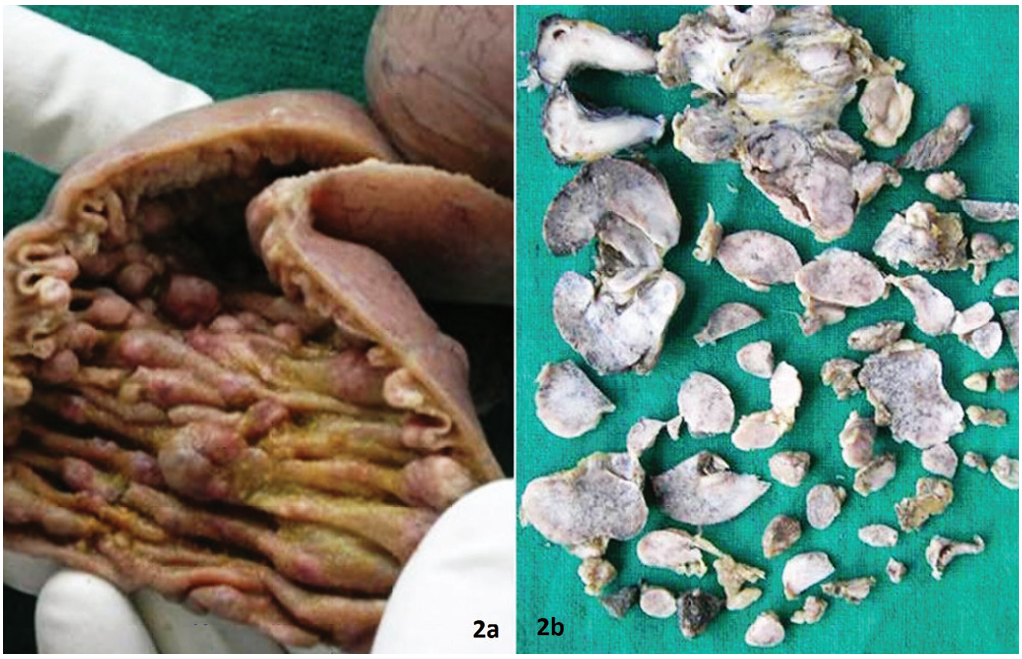

Fig. 2- Cut section showing numerous intraluminal and serosal lymphomatous polyposis (2a) and resected mesenteric lymphnodes (2b).

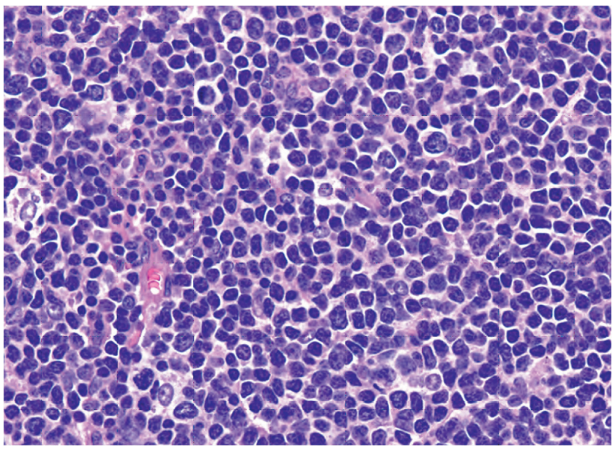

Fig. 3- Histopathological examination of mantle cell lymphoma showing monotonous small lymphoid cells with irregular and slightly angular nuclei (Hematoxylin & Eosin staining ×40)

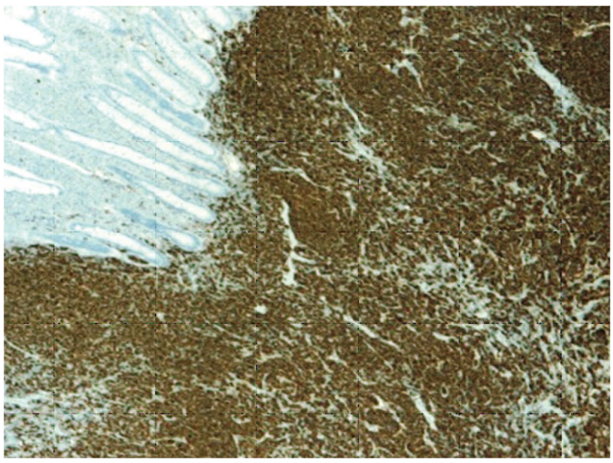

Fig. 4- Immunohistochemistry showing positive nuclear staining for Cyclin D1 (×100 magnification)

Discussion

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). Aamong 13.7% of primary gastrointestinal lymphomas, 41% are diffuse large B-cell

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 polypoid 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, diarrhoea, 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. Immunohistochemocal 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 CD 79a, but negative for BCL6, CD23 and CD10. The absence of CD 23 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 chromosome11 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).

Acknowledgements

The authors declare that there is no conflict of interests.

References

1. Tamura S, Ohkawauchi K, Yokoyama Y, Higashidani Y, Daibata M, Hiroi M, *et al.* Non-multiple lymphomatous polyposis form of mantle cell lymphoma in the gastrointestinal tract. J Gastroenterol 2004;39(10):995-1000.

2. Campo E, Raffeld M, Jaffe ES. Mantle-cell lymphoma. Semin Hematol 1999;36(2):115-27.

3. Harris NL, Jaffe ES, Stein H, Banks PM, Chan JK, Cleary ML, *et al.* A revised European-American classification of lymphoid neoplasms: a proposal from the International Lymphoma Study Group. Blood 1994;84(5):1361-92.

4. Saito T, Toyoda H, Yamaguchi M, Nakamura T, Nakamura S, Mukai K, *et al.* Ileocolonic

lymphomas: a series of 16 cases. Endoscopy 2005;37(5):466-9.
5. Grin A, Chetty R, Bailey D. Mantle cell lymphoma as a rare cause of intussusception: a report of 2 cases. Ann Diagn Pathol 2009;13(6):398-401.
6. Kella VK, Constantine R, Parikh NS, Reed M, Cosgrove JM, Abo SM, *et al.* Mantle cell lymphoma of the gastrointestinal tract presenting with multiple intussusceptions--case report and review of literature. World J Surg Oncol 2009; 7:60.:60.
7. Kodama T, Ohshima K, Nomura K, Taniwaki M, Nakamura N, Nakamura S, *et al.* Lymphomatous polyposis of the gastrointestinal tract, including mantle cell lymphoma, follicular lymphoma and mucosa-associated lymphoid tissue lymphoma. Histopathology 2005;47(5):467-78.
8. Fraga M, Lloret E, Sanchez-Verde L, Orradre JL, Campo E, Bosch F, *et al.* Mucosal mantle cell (centrocytic) lymphomas. Histopathology 1995;26(5):413-22.
9. Kanehira K, Braylan RC, Lauwers GY. Early phase of intestinal mantle cell lymphoma: a report of two cases associated with advanced colonic adenocarcinoma. Mod Pathol 2001;14(8):811-7.
10. Ruskone-Fourmestraux A, Delmer A, Lavergne A, Molina T, Brousse N, Audouin J, *et al.* Multiple lymphomatous polyposis of the gastrointestinal tract: prospective clinicopathologic study of 31 cases. Groupe D'etude des Lymphomes Digestifs. Gastroenterology 1997;112(1):7-16.
11. Kauh J, Baidas SM, Ozdemirli M, Cheson BD. Mantle cell lymphoma: clinicopathologic features and treatments. Oncology (Williston Park) 2003;17(6):879-91.
12. Matuchansky C, Touchard G, Lemaire M, Babin P, Demeocq F, Fonck Y, *et al.* Malignant lymphoma of the small bowel associated with diffuse nodular lymphoid hyperplasia. N Engl J Med 1985;313(3):166-71.
13. Farrell RJ, Ang Y, Kileen P, O'Briain DS, Kelleher D, Keeling PW, *et al.* Increased incidence of non-Hodgkin's lymphoma in inflammatory bowel disease patients on immunosuppressive therapy but overall risk is low. Gut 2000;47(4):514-9.
14. Bernstein CN, Blanchard JF, Kliewer E, Wajda A. Cancer risk in patients with inflammatory bowel disease: a population-based study. Cancer 2001;91(4):854-62.
15. Li JY, Gaillard F, Moreau A, Harousseau JL, Laboisse C, Milpied N, *et al.* Detection of translocation t(11;14)(q13;q32) in mantle cell lymphoma by fluorescence in situ hybridization. Am J Pathol 1999;154(5):1449-52.
16. Lenz G, Dreyling M, Hiddemann W. Mantle cell lymphoma: established therapeutic options and future directions. Ann Hematol 2004;83(2):71-7.