CASE REPORT

Mantle Cell Lymphoma with a Single Protruding Lesion as the Cause of Intussusception

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Abstract:
Mantle cell lymphoma (MCL) is a malignant lymphoma of the gastrointestinal tract that mostly presents as multiple lymphomatous polyposis (MLP); however, MLP with intussusception is rarely reported in MCL. Furthermore, a single protruding lesion with intussusception has never been reported in primary small intestinal MCL. A 70-year-old man presented with pain in the right lower abdomen. Computed tomography and colonoscopy revealed ileocecal intussusception. Ileocecal resection was performed. Histology and immunohistochemistry of the resected specimen showed MCL with a single protruding lesion. The patient was successfully treated with surgery alone and remains in complete remission at the three-year follow-up.

Key words: mantle cell lymphoma, protruding lesion, intussusception

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Introduction

Mantle cell lymphoma (MCL) is a non-Hodgkin’s lymphoma (NHL) composed of small lymphoid cells. Approximately 6% of lymphomas are classified as MCL. MCL generally occurs in adults with a median age of 60 years and has a male predominance (1). The primary presentation of extra-nodal disease occurs in one quarter of patients and frequently involves the Waldeyer’s ring and gastrointestinal tract. Primary gastrointestinal MCL accounts for 1% to 4% of primary gastrointestinal lymphomas (2). Multiple lymphomatous polyposis (MLP) is one of the most common primary gastrointestinal presentations of MCL and accounts for 9% of primary gastrointestinal lymphomas (3). It frequently occurs in the colon and small bowel, particularly in the ileum and ileocecal region. However, MCL may also present endoscopically as the protruding type, fold-thickening type, ulceration type, and superficial type throughout the gastrointestinal tract (4).

Malignant lymphoma is an uncommon cause of intussusception, causing less than 1% of all cases of intussusception (5). A single protruding lesion in the small intestine combined with intussusception has never been reported in patients with MCL. We herein report a case of primary small intestinal MCL with a single protruding lesion causing ileocecal intussusception.

Case Report

A 70-year-old previously healthy man presented with a 3-day history of constant pain in the right lower abdomen, associated with nausea, without a fever or weight loss. He had no history of gastrointestinal or hematological diseases or remarkable family history. A physical examination revealed normal vital signs, a soft distended abdomen with normoactive bowel sounds, and a palpable tender mass in the right lower quadrant. His laboratory data were as follows: white blood cell count, 6,400/μL; hemoglobin level, 13.0 g/dL; C-reactive protein level, 0.98 mg/dL; and soluble interleukin-2 receptor level, 790 U/mL. Lactate dehydrogenase levels were normal. Contrast-enhanced computed tomography (CE-CT) of the abdomen showed intussusception and a mass in the terminal ileum (Fig. 1A and B). In addition, CE-CT of the neck, chest, and pelvis revealed no lymph node enlargement or organ involvement besides in the ileocecal region.

Total colonoscopy demonstrated ileocolic intussusception and a large submucosal tumor-like protruding mass sinking into the cecum out of the terminal ileum. The mass had a smooth surface with multiple, irregularly shaped ulcerations.
and a diameter of about 50 mm (Fig. 2A). The mass returned to the terminal ileum on colonoscopic manipulation, resolving the intussusception without operation (Fig. 2B). Esophagogastroduodenoscopy revealed no involvement of the upper digestive tract.

Multiple biopsies of the mass showed diffuse infiltration of monotonous small to small-to-medium sized lymphoid cells (Fig. 3A and B). In addition, an immunohistochemical analysis of the biopsy tissues demonstrated positive staining for CD20 and CD5 (Fig. 3C and D); therefore, MCL was suspected. Surgery was planned to prevent intussusception from recurring.

The patient underwent ileocecal resection and ileocolic anastomosis. A gross examination of the resected specimen showed a single solid, fleshy protruding lesion, which had a cobblestone-like appearance, in the terminal ileum (Fig. 4A and B). The lesion measured 8.8×5.5 mm. There were no other polyps in the resected specimen.

The histology of the lesion in the terminal ileum revealed a monotonous population of intermediate-sized lymphoid cells with irregular nuclear and extensive submucosal infiltration into the mucosa and muscularis propria (Fig. 5A and B). A flow cytometry analysis showed a monoclonal B-cell population with the following marker profile: surface immunoglobulin γ+, CD19+, CD20+, CD5+, CD3-, CD10-, CD11c-, and CD23-. An immunoperoxidase analysis performed on a glass slide showed positive nuclear staining of the tumor cells for cyclin D1 (Fig. 5C). These findings were considered consistent with MCL.

Thereafter, the patient underwent staging investigations. Positron emission tomography (PET) revealed no tracer uptake. A bone marrow biopsy and chromosomal study were also performed; however, no abnormal results were observed. In addition, double-balloon enteroscopy was performed to examine the small intestine, and no other lesions were noted. Based on these findings, we diagnosed the patient with primary small intestinal MCL with a single protruding lesion. The Mantle Cell Lymphoma International Prognostics Index (MIPI) score was 3.0, and the disease was classified as stage I according to the Ann Arbor staging system.

Owing to the patient’s refusal, old age, and low MIPI score, adjuvant chemotherapy was deferred, but we carefully followed up with the patient. Postoperatively, the patient never developed symptoms. Even at three years postoperatively, the patient never developed symptoms. Even at three years postoperatively, the patient never developed symptoms. Even at three years postoperatively, the patient never developed symptoms. Even at three years postoperatively, the patient never developed symptoms. Even at three years postoperatively, the patient never developed symptoms. Even at three years postoperatively, the patient never developed symptoms. Even at three years postoperatively, the patient never developed symptoms. Even at three years postoperatively, the patient never developed symptoms. Even at three years postoperatively, the patient never developed symptoms. Even at three years postoperatively, the patient never developed symptoms. 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Discussion

MCL is a subtype of NHL, comprising about 7% of adult cases of NHL (6). Patients typically present at a median age of 60 years with lymphadenopathy and advance-stage disease (III or IV) (7). MCL is a well-defined entity characterized by the overexpression of cyclin D1 (6). Despite aggressive treatment, MCL has a poor prognosis, with a median overall survival of two to five years, because of its accelerated proliferation, non-specific clinical presentation, and early relapse (7).

MCL most commonly involves the gastrointestinal tract in the form of MLP. MLP accounts for 9% of primary gastrointestinal lymphomas (3), and approximately 10% of patients with MCL present with MLP. Multiple polyps typically involve long segments of the gastrointestinal tract. The ileocecal region is most frequently involved, whereas the esophagus and anus are rarely affected (8, 9). Most patients with MLP will have MCL, but this presentation may also occur in follicular lymphoma and marginal zone lym-
The lymphoid infiltrate fills the submucosa in the terminal ileal lesion [Hematoxylin and Eosin (H&E) staining, ×4]. B: The cells are small to medium in size with irregular nuclei, coarse chromatin, indistinct nucleoli, and pale, scant cytoplasm. Transformed lymphocytes are absent (H&E staining, ×200). C: Cyclin D1 is positive in most nuclei (×400).

A single protruding lesion is the atypical pattern of small intestinal involvement, and with this condition, a large mass causing intussusception, particularly involving the ileocecal region, is a rare but real complication. This case of primary small intestinal MCL with an unusual presentation adds to the spectra of clinical manifestations of small intestinal MCL. Primary small intestinal MCL should be included in the differential diagnosis of a single protruding lesion in the small intestine. Awareness of such occurrences is necessary and might help refine diagnostic and therapeutic methods for primary small intestinal MCL. Physicians should consider this possibility in any patient with a palpable tender mass in the right lower quadrant.

Therapy for MCL is based on the individual patient’s risk. A simple wait-and-watch approach at the beginning is considered appropriate therapy for elderly patients or patients with a low MIPI. Chemotherapy can be started once patients develop symptoms. Initial aggressive chemotherapy with or without bone marrow transplant is recommended in young symptomatic patients. The median duration of remission is 1.5 to 3 years (17). In the current patient, the wait-and-watch approach was followed postoperatively, which was a safe option that allowed for the preservation of a good quality of life and avoidance of side effects from the early initiation of cytotoxic and biological therapies.

The authors state that they have no Conflict of Interest (COI).

References

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