Multiple Lymphomatous Polyposis of the Intestine with Ileocecal Intussusception Due to Mantle Cell Lymphoma: A Case Report of a 34-Year-Old Man

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Patient: Male, 34
Final Diagnosis: Mantle cell lymphoma
Symptoms: Ileocecal intussusception
Medication: —
Clinical Procedure: —
Specialty: Gastroentrology and Hepatology

Objective: Rare disease
Background: Multiple lymphomatous polyposis of the gastrointestinal tract can be associated with the B-cell lymphoma variant, mantle cell lymphoma, with most cases having been described in patients who are more than 50 years-of-age. A rare case of multiple lymphomatous polyposis due to mantle cell lymphoma is reported in a 34-year-old man.

Case Report: A 34-year-old man presented with paroxysmal abdominal pain followed by spontaneous remission, which had been previously diagnosed as gastritis. An episode of ileocecal intussusception occurred, which was confirmed on imaging studies. The diagnosis of multiple lymphomatous polyposis due to mantle cell lymphoma was confirmed following ileocecal resection and histopathology. The patient refused to receive chemotherapy following surgery. Currently, at two-year follow-up, no further abnormality has been found. A review of the literature has shown the importance of endoscopic evaluation in the diagnosis of lymphomatous polyposis.

Conclusions: Multiple lymphomatous polyposis due to mantle cell lymphoma has rarely been described in young patients under the age of 50 years. Gastrointestinal endoscopic examination is important for the early diagnosis of multiple lymphomatous polyposis.

MeSH Keywords: Intussusception • Lymphoma • Lymphoma, B-Cell

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Multiple lymphomatous polyposis of the gastrointestinal tract can be associated with the B-cell lymphoma variant, mantle cell lymphoma [1]. Most cases of multiple lymphomatous polyposis due to mantle cell lymphoma have been described in patients who are more than 50 years-of-age [1]. This report is of a case of multiple lymphomatous polyposis occurring in a man aged 34 years, who presented with paroxysmal abdominal pain followed by spontaneous remission for nine months, prior to ileocecal intussusception, and highlights the importance of gastrointestinal endoscopic examination.

Case Report

A 34-year-old man presented with a nine-month history of episodes of severe abdominal pain lasting for ten minutes, followed by periods of remission. Gastric endoscopy at the local hospital showed gastritis and the pain was relieved by medication. Severe symptoms occurred at seven months, and at five months prior to hospital admission, which could be relieved by medication for gastritis each time. One week prior to hospital admission, the patient developed severe, constant abdominal pain, without nausea or vomiting, but without remission, and was admitted to hospital for investigation.

![Figure 1. Intussusception resulting from multiple lymphomatous polyposis due to mantle cell lymphoma.](image)

(A) Plain abdominal radiograph shows a nonspecific air pattern. (B) Computed tomography (CT) scan shows ileocecal intussusception, thickening of the bowel wall, and enlargement of the mesenteric lymph nodes. (C, D) Colonoscopy shows polypoid lesions of the large bowel, consistent with multiple lymphomatous polyposis.
Figure 2. Pathological features of multiple lymphomatous polyposis due to mantle cell lymphoma. (A) Gross examination of the excised bowel shows thickening of the mucosal folds and multiple polypoid lesions involving the ileum and cecum, with lymph nodes. The colonic mucosa is normal. (B) Low power photomicrograph of a tissue section from the large bowel polyp. Hematoxylin and eosin (H&E) staining. (C) Photomicrograph of a tissue section from one of the polyps shows a uniform population of infiltrating lymphocytes. H&E ×400. (D) Photomicrograph of the immunohistochemistry shows positive staining for cyclin D1. (E) Photomicrograph of the immunohistochemistry shows positive staining for CD20. (F) Photomicrograph of the immunohistochemistry shows positive staining for CD5. (G) Photomicrograph of the immunohistochemistry shows negative staining for CD10. (H) Photomicrograph shows that no lymphoepithelial lesions are seen. H&E ×200.
The patient underwent exploratory laparotomy, and an ileocecal intussusception was identified, followed by ileocecal resection with extended ileal resection. The gross examination of the resection specimen showed thickening of the mucosal folds and multiple polyoid lesions that ranged from 0.2 cm to 0.6 cm in diameter, involving the ileum and cecum, with the enlarged lymph nodes, while the colonic mucosa was normal (Figure 2A).

Histological examination of the resection specimen showed that the polyps contained a uniform lymphoid cell infiltrate (Figure 2B, 2C). Immunohistochemistry showed that the lymphoid cells were uniformly positive for the B-lymphocyte marker CD20 (Figure 2D), and also for cyclin D1 (Figure 2E), and CD5 (Figure 2F), but were negative for CD10 (Figure 2G). No lymphoepithelial lesions were present (Figure 2H). The diagnosis of mantle cell lymphoma was confirmed.

Following surgery, the patient refused to transfer to the Department of Hematology for chemotherapy and requested clinical follow-up only. Currently, the patient has been followed-up for two years and remains in a stable condition. Follow-up with positron emission tomography and computed tomography (PET/CT) and single balloon enteroscopy examination failed to find other lesions, and no extra-intestinal lymphoma has been detected. However, a longer patient follow-up will be required.

**Discussion**

The gastrointestinal tract is the most common site for extranodal malignant lymphoma [2]. Multiple lymphomatous polyposis is recognized in patients with mantle cell lymphoma, but is not a specific feature of this form of lymphoma, and is also found in other types of both T-cell lymphoma and B-cell lymphoma [3, 4]. A study that reviewed 455 cases of lymphomas involving the gastrointestinal tract showed a prevalence of 0.4% for mantle cell lymphoma [5].

Multiple lymphomatous polyposis can present with diverse clinical symptoms including abdominal pain, diarrhea, bleeding, protein-losing enteropathy, intestinal malabsorption, or chylous ascites. The clinical symptoms for the patient in this report included atypical abdominal pain followed by remission. Because most cases of multiple lymphomatous polyposis occur in patients more than 50 years of age, in this 34-year-old patient with gastrointestinal symptoms, initially only gastric endoscopy was performed, and gastritis was confirmed. However, no colonoscopic examination was performed until the patient presented with intussusception, which emphasizes the importance of a thorough gastric and colonic endoscopy examination for patients with chronic abdominal pain.

Multiple lymphomatous polyposis most commonly occurs in the ascending colon, followed by the small intestine, stomach, and duodenum, and particularly involves the ileum and the ileocecal region [6]. Although intestinal involvement with mantle cell lymphoma presented with multiple lymphomatous polyposis in this case, gastric involvement by mantle cell lymphoma can be diverse and include both gastritis and ulcerative lesions [7, 8].

Mantle cell lymphoma is derived from naïve B-cell population of the lymphoid mantle zone. Using immunohistochemistry, mantle zone lymphoma cells are negative for CD23 and positive for CD5 [9]. Cytogenetically, a rearrangement of the Bcl-1 locus on chromosome 11 due to t(11:14) (q13:q32) translocation is found in mantle cell lymphoma, accompanied by cyclin D1 overexpression [9]. As in this case, the histological features of mantle cell lymphoma show uniform, small-sized lymphoid cells, with few mitoses, without the formation of lymphoid follicles or germinal centers, and without lymphoepithelial lesions. In this case, immunohistochemistry confirmed uniform cell positivity for B-cell markers, as well as CD5 and the diagnostic marker for mantle cell lymphoma, cyclin D1; the lymphoma cells expressed the cell surface B-cell marker, CD20, and were also positive for Bcl-2 and CD79a, but were negative for CD10, Bcl-6, MUM-1, CD21, and CD23. The immunohistochemistry findings confirmed the diagnosis of mantle cell lymphoma. In the case of this patient, there was no family history of lymphoma.

On colonoscopy and radiology, the differential diagnosis of multiple lymphomatous polyposis includes adenomatous or hamartomatous polyposis, which cannot be distinguished macroscopically but require tissue diagnosis by histology. When multiple lymphomatous polyposis is confined to involve the terminal ileum and ileocecum, as in this case, benign lymphoid hyperplasia should also be considered in the differential diagnosis. Other small B-cell lymphoproliferative diseases, including nonspecific lymphofollicular hyperplasia, nodal marginal zone B-cell lymphoma, follicular lymphoma, and small lymphocytic lymphoma should be considered in the differential diagnosis.

The prognosis of gastrointestinal mantle cell lymphoma is poor with a mean survival time of less than three years. Chemotherapy for mantle cell lymphoma includes COP (cyclophosphamide, doxorubicin, and prednisolone), anthracycline-containing regimens, and CHOP (cyclophosphamide, doxorubicin, vincristine, and prednisolone) [10]. A variety of new therapies are now being developed for the treatment of mantle cell lymphoma, including bortezomib, which represents the first drug specifically approved for this type of lymphoma. Also, bendamustine, mTOR inhibitors, and lenalidomide have also shown specific activity against mantle cell lymphoma [11]. Because the patient, in this case, was relatively young,
stem cell transplantation could also have been considered. Unfortunately, the patient refused further chemotherapy for a variety of reasons and a follow-up only was undertaken. At the time of writing, the patient is in a stable condition, which may be attributed to the limited gastrointestinal lesions and the timely treatment.

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Conclusions

As this case has shown, multiple lymphomatous polyposis is a rare condition that may also occur in patients under 50 years of age. A thorough gastrointestinal endoscopic examination is important in the early diagnosis of multiple lymphomatous polyposis.