

Case Report

Lymphomatous polyposis of colon presenting as intussusception in an adult male

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Abstract

Mantle cell lymphoma (MCL) comprises 2.5-7% of all non-Hodgkin's lymphomas, and the gastrointestinal tract is involved in about 20% of cases. Multiple lymphomatous polyposis is an uncommon disease that is regarded as the intestinal form of MCL. Although it has variable clinical presentations but intussusception as a presenting symptom is very rare.

Key words: : Mantle cell lymphoma, Multiple lymphomatous polyposis

Introduction

The gut mucosa contains more lymphocytes than the other immune system organs. However, paradoxically, only 10% of lymphomas occur in the gut ⁽¹⁾. Mantle cell lymphoma (MCL) is a rare subtype of B cell non-hodgkin's lymphoma (NHL) and accounts for 2% of all NHL. The gastrointestinal (GI) tract is involved in 10-20% of patients characterized by the presence of multiple lymphomatous polyps ⁽²⁾.

Multiple lymphomatous polyposis (MLP) of colon is a rare disease, and it is very important to precisely establish the histological type of lymphoma as the prognosis and treatment is quite different ⁽³⁾. We present a case of multiple lymphomatous polyposis due to mantle cell lymphoma involving ileum, ileocecal region, appendix, ascending colon and mesenteric lymph nodes presenting with intussusception in a 45 year male.

Case History

A 45 year old previously healthy male presented to the emergency with constant pain in the right lower abdomen associated with nausea and vomiting since three days. Physical examination revealed normal vital signs, a soft distended abdomen with hyperactive bowel sounds and a palpable tender mass in the

right lower quadrant. Laboratory studies revealed – normal haematological and Biochemistry, biochemistry profile and ESR – 50 mm at one hour. The patient underwent right hemicolectomy and specimen was sent for histopathology. The postoperative period was uneventful and the patient was discharged on the seventh postoperative day.

The specimen consisted of ascending colon measuring 20 cm and ileum measuring 28 cm in length. On cutting open, intussusception was identified in ileocecal region. Throughout the length of ascending colon, cecum, ileum and appendix, mucosa revealed polyps ranging in size from 0.1 to 2 cm. Multiple mesenteric lymph nodes were found enlarged varying in diameter from 0.5 to 4 cm which were solid grey white on cut section (Fig 1). Histopathological examination revealed diffuse lymphoproliferative process. The lymphoid cells were monomorphic, medium sized with slightly irregular nuclei and were arranged in a diffuse pattern. Immunohistochemical studies showed positive staining for CD20, CD5 and cyclin D1 while CD10 was negative, thus confirming the diagnosis of mantle cell lymphoma (Fig 2). The patient was treated with CHOP regime and was lost to follow up.

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Figure 1: a) Gross specimen of right hemicolectomy revealing ileocecal intussusception along with enlarged mesenteric lymph nodes b) on cutting open, multiple polyps identified in the ileum, appendix and caecum and colon.

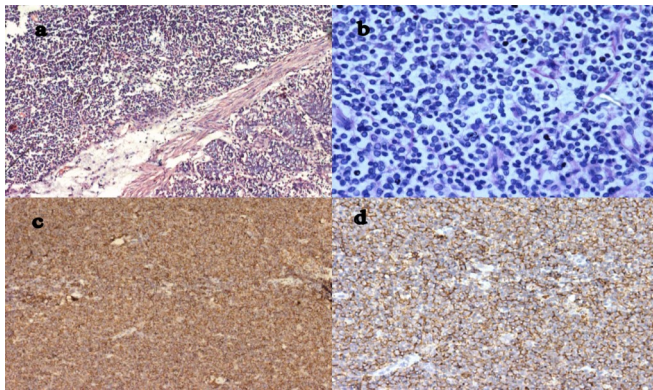


Figure 2: Microscopic examination revealing diffuse infiltration of the gut wall by monomorphic, medium sized lymphocytes [a) H & E; x100 and b) H & E; x200], which on immunohistochemistry showed positivity for cyclin D1 (c) and CD 5 (d).

Discussion

Primary GI lymphomas are rare conditions. They are most common in the stomach, followed by small intestine and colon. Approximately 15- 30% of primary extranodal lymphomas occur in the GI tract, although primary GI tract lymphomas account for only 1 to 10% of all GI malignancies. The great majority of GI tract lymphomas present with a generally solitary lesion, but not as multifocal involvement ⁽¹⁾. MCL is a distinct clinicopathologic entity of non- Hodgkin B cell characterized by a monotonous proliferation of small to medium-sized lymphocytes with co-expression of CD5, CD20, and specific marker of cyclin D1 and frequent (t11;14) (q13;q32) translocation. Macroscopic appearance of MCL in gastrointestinal tract is variable: from tumoral mass, ulcer, mucosal thickness to multiple polypoid lesions ⁽³⁾. MLP is an extremely rare disease. Males are more frequently affected and the disease usually appears during the fifth and sixth decades of life ⁽¹⁾. It presents with symptoms such as abdominal pain, diarrhoea, bleeding, less frequently as protein-losing enteropathy, intestinal malabsorption, or chylous ascites and rarely, as an acute abdomen due to

perforation or intestinal obstruction ⁽⁴⁾. In MLP, the colon is involved in the majority of cases, followed by the small intestine. The ileocecum is frequently the original focus of MCL involvement and remains the primary site of disease. Despite having tropism for the ileocecum, appendiceal MCL has rarely been documented as a mass lesion thickening of the entire mucosal aspect ⁽³⁾. Clinically, MLP may be confused with epithelial polyp and other types of lymphoma especially marginal zone B cell lymphoma of MALT, diffuse large B cell lymphoma, follicular lymphoma and peripheral T cell lymphomas or T/NK cell ⁽²⁾. Definitive diagnosis of MLP requires histological examination of the specimen with histomorphologic and immunophenotypic analysis ⁽⁴⁾.

Surgery is the mainstay of therapy for intussusception in adult patients ⁽⁴⁾. The prognosis of Gastrointestinal MCL is poor, with a mean survival time of less than three years. Response to chemotherapy is seen in up to half of the patients. COP (cyclophosphamide, doxorubicin, prednisolone), anthracycline containing regimens and CHOP (cyclophosphamide, doxorubicin, vincristine and prednisolone) are used as conventional chemotherapies for MCL. Another treatment is rituximab (a chimeric monoclonal antibody) which achieves response rates of about 30% when used alone and >90% when combined with an anthracycline containing regimen ⁽¹⁾. Innovative strategies utilizing bortezomib, temsirolimus or radioimmunoconjugates remain under active investigation ⁽⁴⁾.

References

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