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A case of intestinal MALT lymphoma characterized by diffuse capillary hyperplasia nodules with hemorrhage

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Dear Editor,

A 52-year-old man presented to our department, complaining of recurrent episodes of hematochezia for half a month. Colonoscopy showed diffuse nodular mucosal eminences of approximately 2-3 mm in size in the colorectum and terminal ileum, presenting a "cobblestone" appearance, with visible capillary hyperplasia that are prone to rupture and bleeding upon light touch (Figure 1A). Narrow-band imaging (NBI) showed that the hyperplastic capillaries presented "root-like" branching (Figure 1B). We performed deep biopsies in areas with dense capillary networks or prone-to-bleeding regions at the nodules of the rectum, sigmoid colon, descending colon, transverse colon, ascending colon, ileocecal region, and terminal ileum. Histopathological examination of all biopsy specimens revealed moderate-to-severe chronic mucosal inflammation with activity (+) and prominent lymphoid hyperplasia (Figure 1C-D). Immunohistochemical staining demonstrated that the tumor cells were positive for CD20, CD79a, Bcl-2, Ki67 (20%), CD43. Combined with the morphological features of HE-stained sections, the findings suggested extranodal marginal zone lymphoma of mucosa-associated lymphoid tissue (indolent) (Figure 1E-F).



Discussion

Extranodal marginal zone lymphoma of mucosa-associated lymphoid tissue (MALT lymphoma) is a low-grade B-cell lymphoma and a subtype of marginal zone lymphoma (MZL). It originates from memory B cells in the mucosa-associated lymphoid tissue (MALT) (1). In all non-Hodgkin lymphoma (NHL) cases, MALT lymphoma accounts for 5-8% (2); among MALT lymphomas, gastric MALT lymphoma makes up 50-70%, while intestinal MALT lymphoma only constitutes 5-15% (2-3). Intestinal MALT lymphoma that concurrently involves the colorectum and small intestine is even rarer. Gastrointestinal MALT lymphoma, an indolent B-cell lymphoma, carries a risk of transformation to aggressive lymphoma. Approximately 5-15% of gastrointestinal MALT lymphomas may transform to diffuse large B-cell lymphoma (DLBCL), which is significantly higher than the transformation rate of 1-3% in gastric MALT lymphoma. Following transformation, the 5-year survival rate declines from over 80% during the indolent phase to 40-50% in the aggressive phase (4).

Endoscopic findings of gastrointestinal MALT lymphoma are varied and nonspecific. Macroscopically, they can be classified into three types: a. Mucosal elevated type: single or multiple nodular elevated mucosa, mostly smooth surface, with visible irregular vascular networks. Multiple elevated lesions may show a "cobblestone-like" change. b. Ulcerative type: shallow ulcers with irregular margins, and surrounding mucosa may be congested or edematous. c. Infiltrative type: diffuse mucosal roughness, erosion, or congestion, sometimes with thickened mucosal folds resembling "brain gyri". In this case, the intestinal nodules, smooth-surfaced and without erosion or ulceration, were often misdiagnosed as benign lesions. About 75% of primary intestinal MALT lymphomas are initially misdiagnosed as benign, mainly due to endoscopists' insufficient understanding of the endoscopic features of intestinal MALT lymphoma. For this case, the main endoscopic features were: a. Multisegmental diffuse homogeneous nodular mucosal elevations. b. Capillary hyperplasia with a "root-like" pattern under NBI. c. A tendency for contact-induced bleeding.



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Figure 1. (A) Colonoscopy showed diffuse nodular mucosal eminences of approximately 2-3 mm in size in the colorectum and terminal ileum, presenting a "cobblestone" appearance, with visible capillary hyperplasia that are prone to rupture and bleeding upon light touch. (B) NBI identified the hyperplastic capillaries presented "root-like" branching. (C-D) H&E staining of biopsy specimens from the rectum to the terminal ileum. (E-F) Immunohistochemical staining of biopsy specimens from the rectum to the terminal ileum.