# Intestinal Mucosal-Associated Lymphoid Tissue Lymphoma Mimicked Cryptogenic Multifocal Ulcerous Stenosing Enteritis

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To the Editor: A 50-year-old man was admitted to Peking Union Medical College Hospital with recurrent abdominal pain, vomiting, and melena. In the past 2 years, he had visited several hospitals for abdominal discomfort and was diagnosed as ileus or subileus. Normal mucosa was shown in both gastroscopy and colonoscopy. Three months before admission, the patient started suffering from recurrent hematemesis and melena. Physical examination only showed abdominal tenderness, without lymphadenopathy. His hemoglobin was 94 g/L. The lymphocyte count, platelet count, liver function, tumor markers, and erythrocyte sedimentation rate (ESR) were normal. Antinuclear antibodies, anticytoplasmic antibodies, and anti-saccharomyces cerevisiae antibody were undetectable. Small-bowel barium enema showed multiple ulcerations and stenosis in the jejunum and proximal hold-up [Figure 1a]. Computed tomography (CT) confirmed the presence of multiple jejunal strictures, prestenotic dilation, and bowel wall thickening [Figure 1b]. The 18F-fluorodeoxyglucose positron emission tomography-CT (PET-CT) suggested intestinal inflammation. The appearance of skip lesions made us consider a rare disease called cryptogenic multifocal ulcerous stenosing enteritis (CMUSE). Thus, the patient was treated with 40 mg daily prednisone, which was slowly tapered. Significant clinical improvement occurred and the patient was discharged.

Two months later, the patient was presented to emergency department for abrupt abdominal pain. Abdominal X-ray revealed subphrenic free air; thus, the patient was taken to surgery. The surgeon found more than 20 areas of strictures, starting from the Treitz ligament all the way to proximal ileum [Figure 1c and 1d]. The perforation area was located at 170 cm distal to Treitz ligament with thickened adjacent mesentery and local pus accumulation. The partial resection of jejunum was performed. Histopathological finding was atypical lymphocytes infiltrating from mucosa to the whole muscular layer and serosa [Figure 1e]. Immunohistochemistry demonstrated strongly positive stained with CD20 [Figure 1f], confirming the diagnosis of lymphoma of mucosal-associated lymphoid tissue (MALT). The patient was treated with rituximab, cyclophosphamide, epirubicin, vindesine, and prednisone for 6 courses and remained in partial remission till now.

MALT lymphoma comprises about 5% of all non-Hodgkin's lymphoma. Although gastrointestinal tract is frequently involved, small intestine is a relatively uncommon site of origin, and only 9% involves jejunum.[1] Multiple superficial ulcers, causing significant stenosis, normal ESR, and good response to glucocorticoids, were typical characteristics of CMUSE.[2] However, since the ulcerations were localized to the mucosa and submucosa instead of transmural inflammations, the diagnosis of CMUSE was questioned when the patient encountered intestinal perforation. Ultimately, our patient was diagnosed as MALT lymphoma, which partly explains the symptoms temporarily improved by glucocorticoids. In spite of this, the macroscopical presentation in this case was an unusual pattern. According to the clinicopathological analysis of 143 primary intestinal lymphomas,[1] tumorous type was the most common pattern (86.7%). Saito *et al*.[3] reviewed colonoscopy features of MALT lymphoma, confirming that the main appearance was solitary or multiple, sessile, and semipedunculated protrusions

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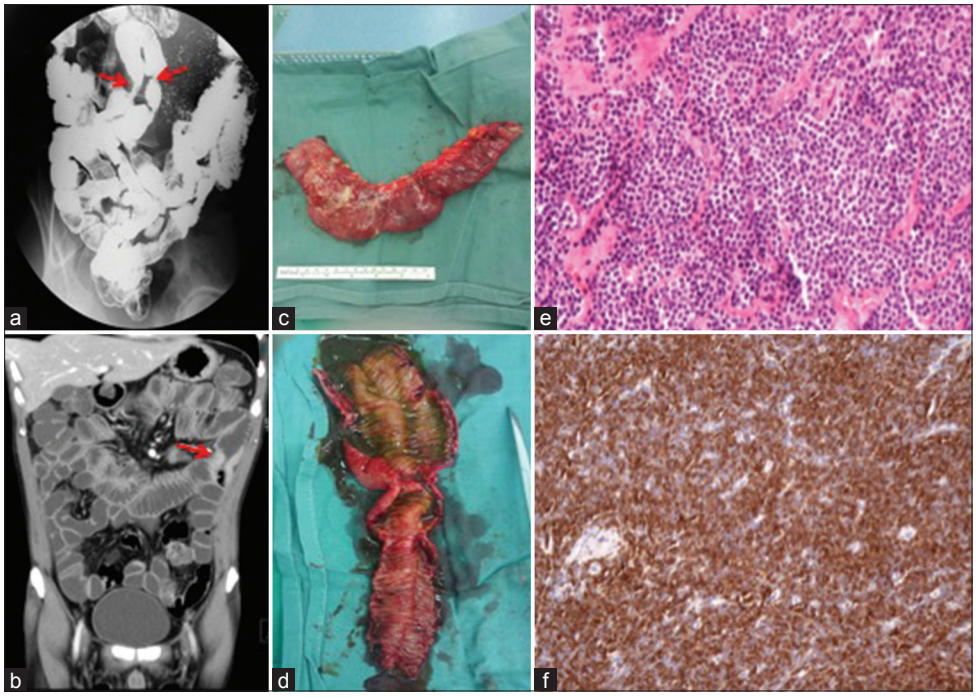


**Figure 1:** A 50-year-old man with lymphoma of mucosal-associated lymphoid tissue. (a) Multiple intestinal stenosis in small-bowel barium enema; (b) multiple jejunal strictures and bowel wall thickening in computed tomography; (c) perforation area with local pus and debris accumulation; (d) multiple intestinal strictures with significant bowel wall thickening; (e) histopathological finding showed infiltrated atypical lymphocytes (H & E staining, original magnification, ×150); (f) immunohistochemical examination demonstrated strongly positive stained with CD20 (original magnification ×150).

covered with seemingly normal mucosa, instead of skip-pattern stenosis and superficial ulcerations in our case.

Findings on PET in our patient indicated inflammation rather than malignancy. MALT lymphoma, as a clinically indolent subtype of lymphoma, might have variable FDG-avidity on PET-CT.[4] In a prospective study,[5] PET failed to visualize all the biopsy-confirmed MALT lymphoma; therefore, double-balloon enteroscopy, capsule endoscopy, radiographic imaging, and pathological study would be critical in early diagnosis of small-intestinal lymphoma.

## Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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## Conflicts of interest

There are no conflicts of interest.

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