中文題目：紅斑性狼瘡患者的急性腹痛鑑別診斷: 瀰漫性大型 B 細胞淋巴癌，以模仿腸繫膜血管炎方式呈現

英文題目：Diffuse large B lymphoma mimics mesenteric vasculitis in a lupus woman with acute abdominal pain.

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Background

Systemic lupus erythematosus (SLE) is an autoimmune disease that affects multiple organs, including skin, joints, kidney, nerve system and gastrointestinal (GI) tract. Acute abdomen (AA) in lupus patients is a medical urgency and demands intensive care for the high mortality in previous studies[1]. Diagnosis of AA is challenging in lupus patients because clinical manifestation might be masked by long-term use of steroid or immunomodulation therapy. Early introduction of computerized tomography (CT) is efficient and rapid for the differential diagnosis, comprising lupus related (such as lupus mesenteric vasculitis, LMV) and non-lupus related etiology (such as appendicitis, cholecystitis). Diagnosis of LMV is largely dependent on abdominal CT finding, including bowel distention, bowel wall thickening, Comb sign, target signs and mesenteric fat attenuation[2]. Complete bowel rest, immediate therapy of intravenous glucocorticoid after early recognition of LMV by abdominal CT may avoid potentially life-threatening perforation and other fatal complications[3]. However, diagnosis of LMV based on image definition required to be thoroughly surveyed, correlating with lupus disease activity and clinical symptoms carefully before treatment decision. In this paper, we will discuss a lupus woman with AA, presented with typical LMV finding on CT but different final diagnosis.

Case report

A 49-year-old woman suffered from progressive onset of diffuse abdominal pain for 3 days, with acute exacerbation over periumbilical region. Other associated symptoms included nausea, dyspepsia and constipation, without demonstrating fever, vomiting, or blood in stool. She had been taking long-term hydroxychloroquine and low dose steroid for lupus, which was diagnosed ten years ago with stable disease activity. Due to inadequate control for abdominal pain which rapidly progressed in hours, she was referred to the emergency depart of a medical center. Physical examination at admission revealed that she was alert, but pale and cold-sweating, with a body temperature of 36.4℃, blood pressure of 122/78 mmHg, respiratory rate of 19

breaths/min and pulse rate of 104 beats/min. Abdominal examination revealed abdominal distention, diminished bowel sound and rebounding tenderness over periumbilical region, without presence of skin ecchymosis. Hemogram demonstrated leukopenia, microcytic anemia, and thrombocytopenia. A contrast-enhanced abdomen CT revealed intramural bowel gas, segmental edematous change and thickening of small bowel, as well as comb sign formation (Figure 1A and 1B).

Under the impression of small bowel perforation, she received emergent laparotomy, which revealed perforation at ileum. About 70-80cm ileum was resected, with side-to-side anastomosis. Tissue biopsy of the resected bowel presented with large lymphoid cells disrupting the underlying structural integrity of the follicle center and positive stain for CD20, compatible with diffuse large B cell lymphoma. There was no evidence of mesenteric vessel involvement.

Discussion

Gastrointestinal involvement is common in lupus patients, and LMV is the most common and serious complication. We presented a case of lupus woman, presented with similar images finding as LMV by abdominal CT. However, image finding is not specific to LMV, and diagnosis acquires further clinical consideration, such as SLEDAI (SLE disease activity). A case-control study indicated that non-lupus related AA should be considered in patient with SLEDAI less than 5[1]. However, another study performed by Lee CK et al. provided contradictory evidence, indicating that SLEDAI did not correlate well with the occurrence of LMV, but leukopenia was associated with the LMV occurrence [4]. Another character for differential diagnosis is that, patients with LMV response well to intravenous glucocorticoid, and almost resulted in complete remission. For patients with inadequate response or recurrent abdominal pain after glucocorticoid therapeutic trial, diagnosis other than LMV should be taken into consideration. In conclusion, image diagnosis by CT for LMV is efficient, but not specific in patients with lupus, especially those with leukopenia or low disease activity at initial presentation.

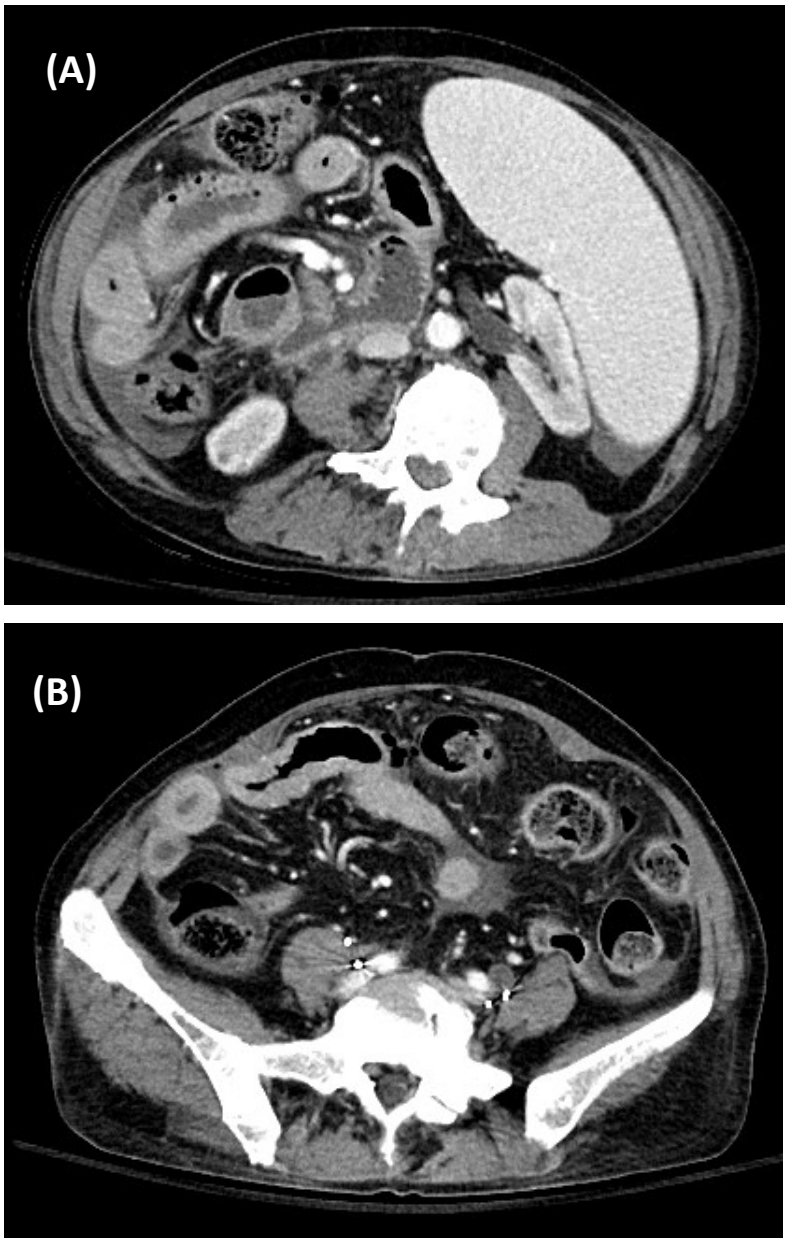

Figure 1. Abdominal CT of a 49-year-old woman with acute abdominal pain. (A) bowel wall thickening with intramural bowel gas, suspicious of bowel perfomation (B) bowel distention and Comb sign formation.

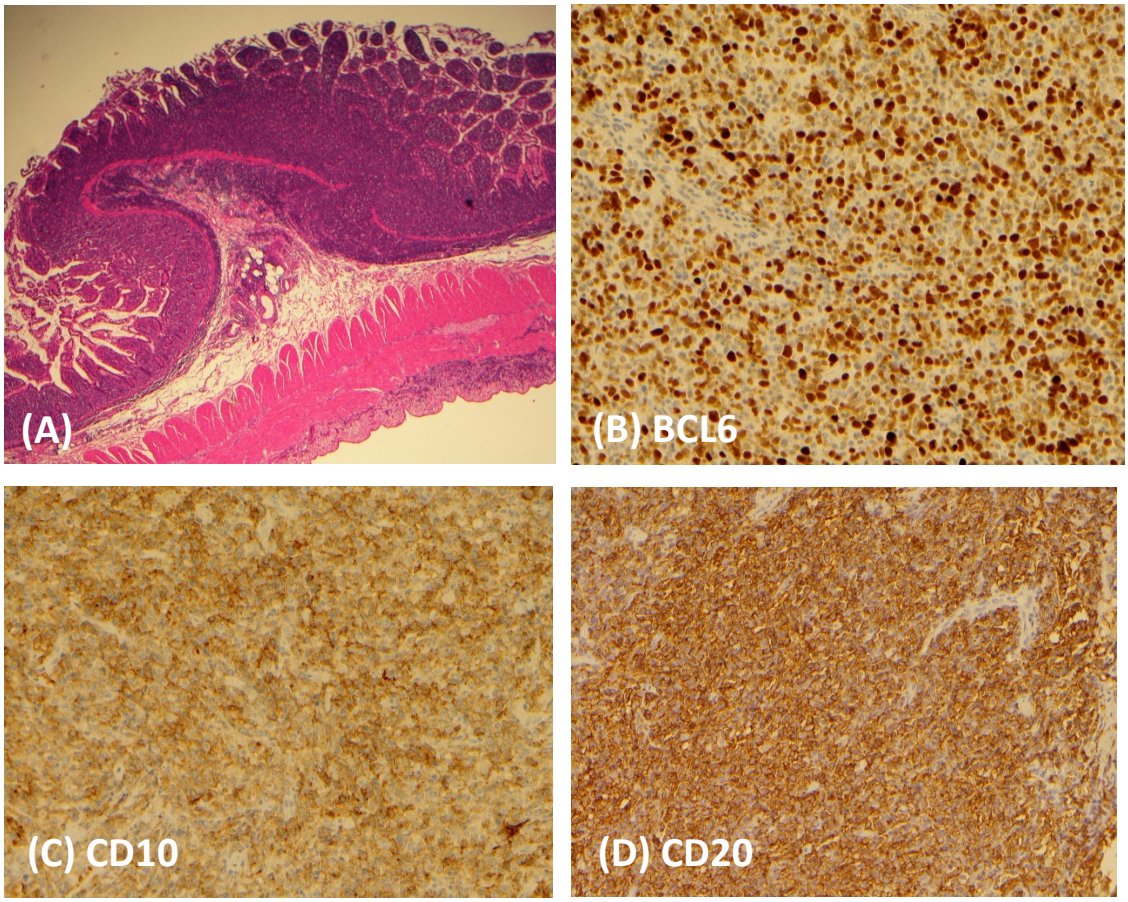

Figure 2. Pathologic finding of the dissected ileum (A) global view (B)BCL 6 stain (C) CD 10 stain (D) CD 20 stain.

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