

Lupus with “thumb-prints,” “targets,” and “combs”

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A 28-year-old woman was undergoing evaluation for proteinuria (urine protein/creatinine ratio=9.6 [normal<0.3]). Two days following kidney biopsy, she presented to the emergency department with abdominal pain, vomiting, and obstipation. In the previous 3 months, she had experienced frequent episodes of abdominal pain, which used to subside spontaneously. She also had a history of intermittent fever and absence of bowel sounds. Examination revealed distended abdomen with rebound tenderness and absence of bowel sounds. Reduced breath sounds were noted over the infrascapular area on both sides. Laboratory test results were as follows: hemoglobin, 9 g/dL; total leukocyte count, 9779 cells/mm³; erythrocyte sedimentation rate (ESR), 76 mm in the first hour; positive anti-nuclear antibody (1:640, homogeneous); C3, 61 mg/dL; and C4, 8 mg/dL. An erect radiograph of the abdomen excluded any bowel perforation. Computed tomography of the chest and abdomen revealed pleural effusion on both sides with diffuse thickening and edema of the bowel wall giving rise to “thumb-printing” (Figure 1) and “target” sign (Figure 2). The mesenteric vasculature (“comb” sign) and ascites were accentuated (Figure 2). Kidney biopsy revealed Class IV lupus nephritis. The patient was diagnosed with systemic lupus erythematosus (SLE) with nephritis and mesenteric vasculitis and was treated with high-dose pulsed methylprednisolone followed by monthly pulses of intravenous cyclophosphamide and tapering doses of oral prednisolone. Her bowel symptoms improved remarkably after 4 days of treatment. At the 6-month follow-up, she was relieved of abdominal symptoms and her ESR and proteinuria had normalized. She was started on maintenance therapy with azathioprine (2 mg/kg).

Mesenteric vasculitis is a rare manifestation of lupus, which may present with abdominal pain, intestinal obstruction, abdominal distension, or gastrointestinal bleeding. In patients with SLE, mesenteric vasculitis is the most common cause of severe abdominal pain and requires admission (1). Signs of peritonitis can be observed in some cases, which mandates ruling out intestinal perforation. It is commonly associated with evidence of active SLE in other organ systems (2). It can be fatal if not diagnosed and treated promptly with glucocorticoids, to which good response is frequently noted (1, 3). Cyclophosphamide has been successfully administered in steroid refractory cases (3).

Small vessels of the bowel wall, which are involved in lupus mesenteric vasculitis, cannot be visualized using mesenteric angiography. However, angiography can be performed on suspicion of involvement of larger arteries, which occurs in polyarteritis nodosa, atherosclerosis, or thrombosis. In addition, the most common site of involvement is the small bowel, which is not easily amenable to biopsy. Because of these limitations, computed tomography of the abdomen is the most useful modality for diagnosis. It reveals characteristic features such as edema of the bowel wall, giving rise to the “target” sign on cross-sections (4).

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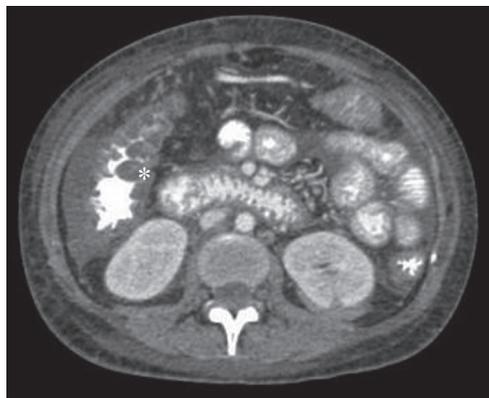


Figure 1. Contrast-enhanced CT of the abdomen showing thumb-like projections of the intestinal mucosa into the lumen: “thumb-print” sign (asterisk).

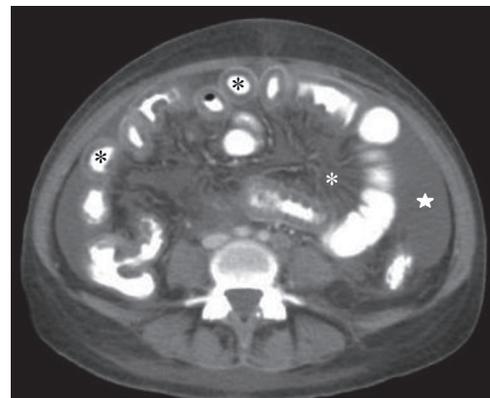


Figure 2. Contrast-enhanced CT of the abdomen showing edema of the bowel wall: “target” sign (black asterisks); prominence of mesenteric vasculature: “comb” sign (white asterisk) and ascites (star).

Thumb-like projections of the intestinal mucosa into the lumen appear as "thumb-printing" on longitudinal sections, which indicate bowel wall ischemia (4). Prominence of mesenteric vessels resembles the teeth of a comb ("comb sign") (4). Prompt treatment is necessary to prevent complications such as pneumatosis intestinalis and bowel perforation.

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