

CASE REPORT

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A case with hydroureteronephrosis accompanied by Lupus Mesenteric Vasculitis

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Abstract

Lupus mesenteric vasculitis is an uncommon and severe complication of systemic lupus erythematosus (SLE). Frequently, these patients exhibit dilated bowel segments, focal or diffuse thickening of the intestinal walls, abnormal contrast enhancement in the intestinal wall, target or double halo, mesenteric edema, engorged mesenteric vessel and ascites in abdominal computerized tomography (CT) imaging. However, these signs lack specificity, limiting the diagnostic role of CT. Intestinal pseudo-obstruction in association with hydroureteronephrosis has been previously reported in SLE patients. Here in, we report a case of lupus mesenteric vasculitis occurring in conjunction with hydroureteronephrosis. Presence of hydroureteronephrosis may assist in improving the specificity of CT in lupus mesenteric vasculitis.

Keywords: Hydroureteronephrosis, mesenteric vasculitis, systemic lupus erythematosus

Introduction

Systemic lupus erythematosus (SLE) is an auto-immune condition with multi-organ and multi-system involvement that is characterized by the formation of auto-antibodies due to immune system dysfunction as well as the activation of the complement system [1]. Lupus mesenteric vasculitis (LVM) is the result of immune complex deposition in the intestinal walls and thrombosis in the intestinal vasculature. LVM is a severe complication of SLE characterized by pain, and intestinal ischemia associated with LVE may lead to life-threatening consequences [2].

Although a number of signs have been reported in abdominal CT imaging studies in SLE patients with abdominal pain, none is adequately specific for a diagnosis of LMV [3]. On the other hand, co-existence of intestinal pseudo-obstruction and hydroureteronephrosis has been previously reported in SLE patients [4]. In this case report, we describe a patient with SLE who presented with acute abdominal pain and who was subsequently found to have lupus mesenteric vasculitis in association with hydroureteronephrosis.

Case report

A 23-year old female patient presented to the emergency room with severe continuous abdominal pain of acute onset two days ago. She had no symptoms other than abdominal pain.

Reportedly, she had been diagnosed with systemic lupus erythematosus (SLE) one year ago with initial symptoms as arthralgia, malar rash, oral aphthous ulcers, and hydroxychloroquine and methyl prednisolone treatment had been given. However, the patient did not attend to follow-up controls regularly and was receiving only oral methyl prednisolone 32 mg/day for the past year. She also had a history of surgery for inguinal herniation 3 years ago. Physical examination showed stable vital signs and abdominal tenderness on palpation as well as rebound, more marked in the lower quadrants.

Laboratory examination showed no abnormality in complete blood count values, while CRP was 12 mg/dl (N=0-5), erythrocyte sedimentation rate was 48 mm/h (N=0-20), anti-ds DNA was +++, anti-nucleosomes ++, antinuclear antibody was 1/1000 granular-homogenous, C3 was 45 mg/dl (n=90-180), and C4 was 4.6 mg/dl (n=15-40), anti-cardiolipin antibody IgM and IgG were negative, lupus anticoagulant was negative. Liver and kidney function tests, urinalysis, amylase, and lipase were normal. Direct erect and supine abdominal x-rays were unremarkable, and abdominal ultrasound showed grade 2 hydroureteronephrosis on the right and wall thickening in the small intestinal wall segments suggestive of edema (Figure 1). A contrast-enhanced CT examination showed the following: contrast enhancement in the internal and external layers of the intestinal segments at more than one localization suggestive of LMV, intestinal wall segment thickening with double-halo sign, intraperitoneal free fluid, as well as right hydroureteronephrosis (Figure 2). The patient was receiving 32 mg of daily methylprednisolone and was considered to have LMV. After

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pulsed methylprednisolone at a daily dose of 1 g for three days, steroid therapy was continued at 1 mg/kg/day. Abdominal pain resolved completely within one week. A follow-up ultrasound examination showed improvement in intestinal wall thickening and hydroureteronephrosis. The patient was discharged on hydroxychloroquine and a tapering dose of steroids

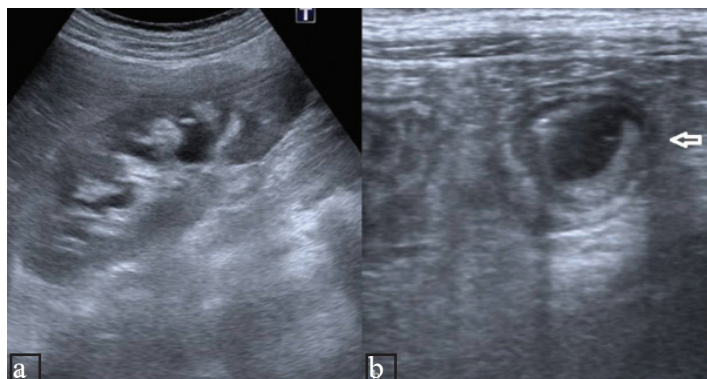


Figure 1. Abdominal ultrasound performed at the time of abdominal pain showing the following: a) grade 1 hydronephrosis in the collecting system of the right kidney, and b) diffuse symmetrical intestinal wall thickening (arrow) in both pelvic areas (more marked on the right) consistent with the edema in small intestinal walls. No free fluid was detected with ultrasound

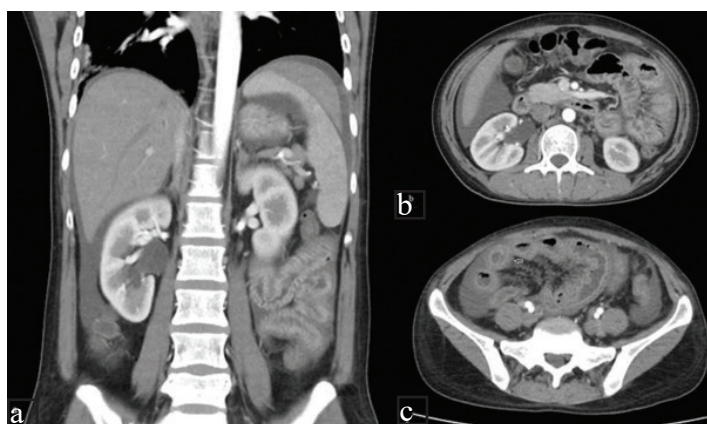


Figure 2. Coronal (a) and axial (b-c) cross-sections of contrast enhanced abdominal computerized tomography performed two days after ultrasound examination showing wall thickening in small intestinal segments suggestive of edema in addition to double halo sign (arrow) and grade 2 hydroureteronephrosis on the right. Also, free intraperitoneal fluid is present.

Discussion

Lupus mesenteric vasculitis represents a major cause of acute abdominal pain in SLE patients, manifesting itself by acute onset of diffuse and severe abdominal pain. Mesenteric arteritis, lupus arteritis, lupus enteritis, gastrointestinal vasculitis, intra-abdominal vasculitis, and acute gastrointestinal syndrome have been used to describe LMV [5,6]. Initial step in management requires excluding other causes of abdominal pain including acute gastroenteritis, peptic ulceration, acute pancreatitis, and peritonitis. LMV is characterized by immune complex deposition, complement activation with subsequent submucosal edema, and leukocytoclastic vasculitis involving mesenteric vessels and thrombus formation [4]. In the early stages of LMV, high-dose steroids are generally fairly effective and complications such as intestinal ischemia, necrosis, or perforation may be prevented by medical treatment. Abdominal computerized tomographic

(CT) imaging represents the most useful diagnostic modality for establishing a diagnosis of LMV. Typical signs of mesenteric vasculitis in abdominal CT include the target sign and the comb sign. Another important cause of acute abdomen in SLE patients is intestinal pseudo-obstruction (IPO) [5], which is a condition with impaired intestinal motility without mechanical or obstructive causes and which may be life-threatening if not treated promptly. IPO may also co-exist with hydroureteronephrosis and is diagnosed on the basis of clinical examination and imaging studies. Direct x-rays may display multiple air-fluid levels as well as enlarged small intestinal segments, while CT may also show increased intestinal wall thickness [7]. Pathophysiological mechanisms underlying the development of IPO in SLE have not been clearly elucidated, although proposed mechanisms include development of visceral smooth muscle injury and dysmotility subsequent to immune-complex mediated vasculitis. The smooth muscle dysfunction may be diffuse and may involve urinary and biliary systems as well [8-10]. Mesenteric ischemia due to LMV may be associated with a number of findings in abdominal CT including focal or diffuse intestinal wall thickening, enlarged intestinal segments, abnormal contrast enhancement in the intestines (target or double halo sign), mesenteric edema, engorgement of the vascular structures penetrating the intestinal wall, and ascites. Since mesenteric vasculitis may affect a multitude of vessels, intestinal wall thickening is of multi-focal nature. However these signs in abdominal CT examination are not specific for LMV and may also occur in other conditions such as pancreatitis, mechanical intestinal obstruction, peritonitis, or inflammatory bowel disease [11]. Our patient had severe and diffuse abdominal pain of sudden onset, with intestinal wall edema, target sign, and comb sign in abdominal CT examination. Also she had grade 2 hydroureteronephrosis on the right side. In this patient previously diagnosed with SLE, the intestinal findings were considered to be due to LMV and hydroureteronephrosis was thought to be secondary to the involvement of urinary smooth muscles. Despite hydroureteronephrosis, this patient did not have pseudo-obstruction. She developed LMV while receiving 32 mg of methylprednisolone on a daily basis due to SLE, and after diagnosed LMV, 1 g of pulsed methyl prednisolone was administered, and a significant improvement occurred within one week. This case suggests that hydroureteronephrosis may not only accompany IPO but also mesenteric vasculitis due to SLE. Presence of non-obstructive hydroureteronephrosis in conjunction with signs suggestive of mesenteric vasculitis in abdominal computerized tomography may assist in increasing the specificity of CT examination with respect to LMV.

Competing interests

The authors declare that they have no competing interest.

Financial Disclosure

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