

Photoclinic

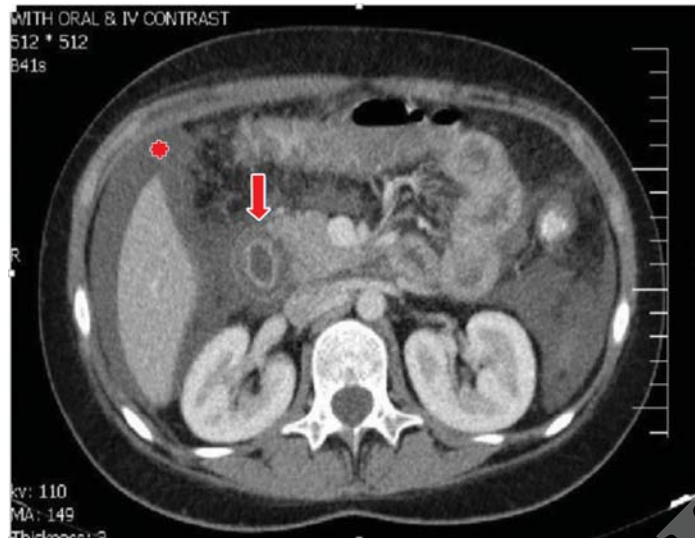


Figure 1. Abdominal CT scan showing ascites (asterisk) and target sign (arrow)



Figure 2. Abdominal CT scan showing comb sign (arrow)

Cite this article as: Sagharnia S, Darabi F, Jamshidi AR. Photoclinic. *Arch Iran Med*. 2016; **19(11)**: 821 – 822.

A 17-year-old woman with 2 weeks history of generalized crampy abdominal pain, progressive nausea/vomiting, and loose stools was admitted to the emergency department. She was a known case of systemic lupus erythematosus (SLE) with a history of associated mucocutaneous symptoms (Malar rash), nephritis (proteinuria 600 mg/24 h which wasn't biopsied), arthritis and positive serology (positive ANA and Anti ds-DNA, with titer 2 times greater than normal values) from five years ago. Due to nephritis, she was taking Mycophenolate Mofetil (MMF), which was tapered from 2/5 gr/day to 1 gr/day during 5 years. She was also under the treatment with hydroxychloroquine 200 mg daily and Prednisolone 5 mg/day at the presentation. She hadn't had any SLE flare up in the previous 5 years and her kidney function was under the control with about 50 mg proteinuria/24 h. Her serologic

tests were also become normal during this period. At the time of admission, physical examination revealed no fever or tachypnea and just mild tachycardia without any heart murmur. Arterial pulses were normal. Malar rash was evident and the abdomen had mild generalized tenderness in the deeper touch. Blood test showed mild leukocytosis, normal liver enzyme and creatinine level, increased levels of ANA and Anti ds-DNA antibodies (about 2 times greater than normal values) and low complement. Urine analysis was normal and 24 h proteinuria was 300 mg. Stool exam was normal. Chest X-ray, and echocardiography were normal too. Early Gastro-duodenoscopy was done in first 36 hours of her admission that was also normal. Contrast enhanced abdominal CT-scan revealed ascites, diffuse circumferential wall thickening and submucosal edema of small bowel with abnormal enhancement (target sign) (Figure 1) and comb like pattern of mesenteric vessels (comb sign) (Figure 2).

Sara Sagharnia MD¹, Farideh Darabi MD¹, Ahmadreza Jamshidi MD¹
 Author's affiliations: ¹Rheumatology Research Center, Shariati Hospital, Tehran University of Medical Sciences, Tehran, Iran.
 •Corresponding author and reprints: Ahmadreza Jamshidi MD, Rheumatology Research Center, Shariati Hospital, Tehran University of Medical Sciences, Tehran, Iran. Telefax: +98-21-84902405, E-mail: jamshida@tums.ac.ir.
 Accepted for publication: 24 August 2015

What is your diagnosis?
 See the next page for diagnosis

Photoclinic Diagnosis:

Small Vessel Mesenteric Vasculitis

The small vessel mesenteric vasculitis (MV) has a global prevalence ranging from 0.2% to 9.7% among patients with lupus and 29% to 65% in those with lupus and acute abdominal pain.¹⁻⁴ The pathology of Lupus MV comprises immune-complex deposition and complement activation, with subsequent submucosal edema, leukocytoclastic vasculitis and thrombus formation.^{1,5} Abdominal CT scan is thought to be the most useful modality for the early diagnosis of mesenteric ischemia⁶ and can demonstrate prominence of mesenteric vessel with a palisade pattern supplying dilated loops (comb sign), ascites, and diffuse or focal circumferential bowel wall thickening, as well as submucosal edema with abnormal enhancement (target sign). Gastroendoscopy and colonoscopy sometimes reveal findings of ischemia and ulceration.^{1,5} Because lupus mesenteric vasculitis typically involves small vessel of bowel submucosa, mesenteric angiography is not usually diagnostic.⁵ However, angiography can be helpful in ruling out larger vessel that causes mesenteric ischemia such as polyarthritis nodosa. Mesenteric vasculitis has a high mortality rate with a reported estimate 50% mortality depending of timing and institution of corticosteroid treatment and surgery.² Medical treatment involves immediate, high-dose, intravenous steroid therapy¹ and if a rapid response to medical treatment is not achieved, surgical intervention is mandatory.⁷ In this patient, in addition to supportive managements, intravenous methylprednisolone (1 gr daily for 3 days) followed by oral

prednisolone 50 mg/day (1 mg/kg) was prescribed and MMF dose was also increased to 2 gr/day. The general condition of the patient markedly improved in the third day and she was discharged from the hospital after one week. She was well 2 weeks later on the follow-up and tapering of prednisolone was initiated.

References

1. Ju JH, Min JK, Jung CK, Oh SN, Kwok SK, Kang KY, et al. Lupus mesenteric vasculitis can cause acute abdominal pain in patients with SLE. *Nat Rev Rheumatol.* 2009; 5: 273 – 281.
2. Tian XP, Zhang X. Gastrointestinal involvement in systemic lupus erythematosus: insight into pathogenesis, diagnosis and treatment. *WJG.* 2010; 16(24): 2971.
3. Kwok SK, Seo SH, Ju JH, Park KS, Yoon CH, Kim WU, et al. Lupus enteritis: Clinical characteristics, risk factor for relapse and association with anti-endothelial cell antibody. *Lupus.* 2007; 16(10): 803 – 809.
4. Lee CK, Ahn MS, Lee EY, sheen JH, Co YS, Ha HK, et al. Acute abdominal pain in systemic lupus erythematosus: focus on lupus enteritis (gastrointestinal vasculitis). *Ann Rheum Dis.* 2002; 61(6): 547 – 550.
5. Sultan SM, Ioannou Y, Isenberg DA. A review of gastrointestinal manifestations of systemic lupus erythematosus. *Rheumatology (Oxford).* 1999; 38 (10): 917 – 932.
6. Ko S, Lee T, Cheng TT, Ng SH, Lai HM, Cheng YF, et al. CT findings at lupus mesenteric vasculitis. *Acta Radiologica.* 1997; 38(1): 115 – 120.
7. Medina F, Ayala A, Jara LJ, Becerra M, Miranda JM, Fraga A. Acute abdomen in systemic lupus erythematosus: the importance of early laparotomy. *Am J Med.* 1997; 103(2): 100 – 105.