Mesenteric Vasculitis as a Rare Initial Presentation of Systemic Lupus Erythematosus: A Case Report

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Abstract

Systemic lupus erythematosus (SLE) is a multisystem autoimmune disease that can also affect the gastrointestinal tract (GIT). The GIT symptoms are common in SLE patients, occurring in up to 40%–50% of the patients and may involve almost any organ along the GIT. Abdominal pain is the most common symptom, and an important cause of this is lupus mesenteric vasculitis (LMV). LMV is a very rare presentation of SLE, and in some cases, can precede the typical manifestations of SLE. Here, we report such a case where a 30-year-old Saudi woman presented clinically with a 2-week abdominal pain and diarrhea; the patient had not previously been diagnosed with SLE. Laboratory investigations and abdomen computed tomography imaging confirmed the diagnosis of LMV. Early recognition of this condition and its proper management improve the outcome of this serious rare initial presentation of SLE.

Keywords: Abdominal pain, mesenteric vacuities, systemic lupus erythematosus

INTRODUCTION

Systemic lupus erythematosus (SLE) is a multisystem autoimmune disease that can affect, among many systems, the gastrointestinal tract (GIT).¹ GIT-related symptoms are common in SLE patients, occurring in up to 40%–50% of the patients, and may involve almost any organ along the GIT. Abdominal pain is the most common symptom, with it being reported in about 75% of the patients.²,³

An important cause of abdominal pain in SLE patients is lupus mesenteric vasculitis (LMV), also known as lupus enteritis, which was first described by Hoffman and Katz in 1980.⁴ LMV is a very rare presentation of SLE, occurring in 0.2%–9.7% of cases with the active disease and <1% in those with inactive disease, yet it is one of the most serious complications of SLE with high mortality. In some cases, LMV can precede the typical manifestations of SLE.²,⁵ In terms of organs involved in patients with LMV, a recent study found that jejunum was involved in 83% of the cases, ileum in 84%, colon in 19%, duodenum in 17% and rectum in 4%.³ Other complications of SLE gastrointestinal manifestations include protein-losing enteropathy, intestinal pseudo-obstruction, hepatic involvement, bowel gangrene and pancreatitis. In addition, intussusception, which is a rare diagnosis in adults, has also been reported as the initial presenting symptom.⁵,⁶,⁷

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Here, we report a case of a young patient presenting with LMV but without the classical manifestations of SLE. To the best of the authors’ knowledge, this is the first such reported case from the Gulf region.

CASE REPORT

A previously well 30-year-old female presented to our hospital with diffuse abdominal pain and diarrhea for 3 weeks. The pain was vague in nature and of a severity index 4–5 on a 10-point pain scale. The patient described the diarrhea as watery without blood or mucus, of large amount that occurred 4–5 times/day, sometimes awakened her from sleep and aggravated by eating. The patient occasionally suffered episodes of vomiting, which did not contain blood. Her appetite had been poor and she lost 4 kg in weight during the 3-week period. However, there was no fever, sweating, skin rash, oral or genital ulcer, hair loss, joint pains or yellow sclerae. In addition, there was no significant family history of note or contact with a tuberculosis patient, and she had never smoked. On examination, the patient looked well and was not in distress. Examination of her systems was unremarkable, apart from mild epigastric tenderness. In particular, there was no skin rash, mouth ulcers or joint swelling.

A provisional diagnosis of inflammatory bowel disease was considered. Other differential diagnoses such as infective colitis, tuberculous enteritis, systemic vasculitis and intestinal lymphoma were also considered.

Laboratory investigations revealed microcytic hypochromic anemia with normal erythrocyte sedimentation rate and C-reactive protein levels. The blood biochemistry, including liver and kidney functions, was within normal ranges, apart from a slightly low serum albumen level [Table 1].

Upper endoscopy revealed antral gastritis, and colonoscopy showed evidence of mild proctosigmoiditis but normal terminal ileum. Biopsies from the sigmoid colon showed mild focal nonspecific chronic inflammatory cell with mild edema, but no evidence of inflammatory bowel disease or tuberculosis. On the other hand, biopsies from the duodenum revealed preserved villous architecture without evidence of inflammation.

Further workup with abdominal computed tomography (CT) showed edematous and circumferential thickening of the small bowel wall, with abnormal bowel enhancement [Figure 1a and b]. Abdominal CT angiography showed normal mesenteric vessels with no microaneurysms. Based on the clinical presentation and these CT findings, vasculitis, in particular systemic lupus enteritis, was highly suspected. Blood tests for SLE were requested, and the patient was treated with oral prednisolone at a dose of 60 mg/day, which was to be gradually tapered. Her condition improved dramatically, as the pain subsided, and she was discharged home after 2 weeks. However, when seen in the clinic 4 weeks later, the patient had developed facial swelling and acne as side effects of steroids. Thus, steroid therapy was gradually tapered to 5 mg daily, and 50 mg of azathioprine was prescribed. Unfortunately, 1 week later, she developed severe side effects of azathioprine that included abdominal pain, nausea, vomiting and marked fatigue with elevation of liver enzymes. These adverse effects completely resolved a week after azathioprine was discontinued. At this point, she started to have arthralgia, hair loss and mouth ulcers.

Blood tests for SLE confirmed a positive antinuclear antibodies and double-stranded DNA, together with low complements and negative antineutrophil cytoplasmic antibody-associated vasculitis. Accordingly, based on more than 4 of 11 criteria by scoring 10 points according to the new diagnostic criteria [8], a diagnosis of SLE was confirmed. The dose of prednisolone was increased to 40 mg/day and hydroxychloroquine at a dose of 250 mg was started. Her symptoms improved completely, and prednisolone was then tapered gradually to a maintenance dose of 10 mg/day.

When seen at the clinic 2 weeks later, the patient’s condition had significantly improved, with abdominal CT revealing significant improvements in the bowel edema [Figure 2].

DISCUSSION

LMV is a rare manifestation of SLE with high rates of mortality if not promptly diagnosed [3,9,10]. LMV presents with acute, diffuse abdominal pain and may be associated with nausea and vomiting, diarrhea or hematochezia. A recent review regarding emergency complications of SLE stated that LMV is the most important cause of

Figure 1: (a) CT showing circumferential bowel wall thickening and edema; (b) sagittal CT showing circumferential bowel wall thickening, edema and increased bowel enhancement (double halo or target sign)
acute abdominal pain in SLE patients.\(^7\) Similarly, in a study by Janssens \textit{et al.},\(^3\) where data from seven similar patients were reported, the clinical presentations included abdominal pain (97%), vomiting (42%), diarrhea (32%) and fever (20%). In agreement with this, our patient had both abdominal pain and diarrhea as the main symptoms. Other laboratory tests usually reflect SLE activity by showing low complement levels (88%), anemia (52%), leuko- or lymphocytopenia (40%) and thrombocytopenia (21%).\(^7\)

In the absence of history of SLE, the diagnosis of LMV can be difficult. In most cases, SLE is diagnosed before the first episode of enteritis,\(^7\) while in about 10% of the cases, SLE is diagnosed simultaneously with the first episode of enteritis;\(^12,13\) only few patients present with enteritis before the diagnosis of SLE.\(^13,14\) In our patient, the clinical features of SLE appeared a month after the initial presentation of enteritis, when the steroid dose was tapered off and when the serological tests confirmed the diagnosis of SLE.

The clinical presentation of mesenteric vasculitis in our patient was supported by the abdominal CT images, the findings of which revealed bowel wall edema and thickening, as described previously in 95% of the patients.\(^7\) Other CT findings in similar cases include ascites (78%), the characteristic target sign (71%), mesenteric abnormalities (71%) and bowel dilatation (24%).\(^3,15,16\) However, histological confirmation is difficult and feasible only in some cases (6%).

Patients diagnosed with SLE lupus mesenteric enteritis should promptly receive corticosteroids as a first-line therapy. Additional immunosuppressants and antitumor necrosis factor agents may be administered either from the initial episode, or in case of relapse, which usually occur in about 25% of patients.\(^9,10\) Such relapses can be treated with pulse steroids and cyclophosphamide as well as biologic therapy.\(^16-19\)

To the best of the authors’ knowledge, this is the first such reported case from the Gulf region. However, several worldwide published case reports have highlighted the different presentations of LMV such as the one reported by Ju \textit{et al.}\(^20\) In general, the diagnosis of LMV is confirmed by abdominal CT and emergency treatment with pulse steroids is needed. In terms of management, treatment with pulse cyclophosphamide and immunoglobins has shown to be a useful modality.\(^9,10,17-19\)

**CONCLUSION**

The present report and similar cases in the literature call attention to the presence of LMV as a rare initial manifestation of SLE. Early recognition of this condition and its proper management improve the outcome of an otherwise lethal condition.

**Declaration of patient consent**

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has provided her consent for her images and other clinical information to be reported in the Journal. The patient understand that her name and initials would not be published and due efforts will be made to conceal her identity, but anonymity cannot be guaranteed.
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Conflicts of interest
There are no conflicts of interest.

REFERENCES

1. Bodh V, Kalwar R, Sharma R, Sharma B, Mahajan S, Raina R, et al. Lupus enteritis: An uncommon manifestation of systemic lupus erythematosus as an initial presentation. J Dig Endosc 2017;8:134-6.
2. Luman W, Chua KB, Cheong WK, Ng HS. Gastrointestinal manifestations of systemic lupus erythematosus. Singapore Med J 2001;42:380-4.
3. Janssens P, Arnaud L, Galiier I, Mathian A, Hie M, Sene D, et al. Lupus enteritis: From clinical findings to therapeutic management. Orphanet J Rare Dis 2013;8:67.
4. Hoffman BI, Katz WA. The gastrointestinal manifestations of systemic lupus erythematosus: A review of the literature. Semin Arthritis Rheum 1980;9:237-47.
5. Halleux DS, Wallace DJ. Gastrointestinal manifestations of systemic lupus erythematosus. Curr Opin Rheumatol 2000;12:379-85.
6. Bort J, and Gerner E. Lupus gastrointestinal tract vasculopathy: Lupus “Enteritis” Involving the entire gastrointestinal tract from esophagus to rectum. Case Rep Gastroenterol 2017;11:48-53.
7. Li Z, Xu D, Wang Z, Wang Y, Zhang S, Li M, et al. Gastrointestinal system involvement in systemic lupus erythematosus. Lupus 2017;26:1127-38.
8. Tedeschi SK, Johnson SR, Boumpas D, Daikh D, Dorner T, Jayne D, et al. Developing and refining new candidate criteria for SLE classification: An international collaboration. Arthritis Care Res (Hoboken) 2018;70:571-81.
9. Yuan S, Ye Y, Chen D, Qiu Z, Zhan Z, Lian F, et al. Lupus mesenteric vasculitis: Clinical features and associated factors for the recurrence and prognosis of disease. Semin Arthritis Rheum 2014;43:759-66.
10. Hill PA, Dwyer KM, Power DA. Chronic intestinal pseudo-obstruction in systemic lupus erythematosus due to intestinal smooth muscle myopathy. Lupus 2000;9:458-63.
11. Marco JL, Chhakchhuak CL. Complications of systemic lupus erythematosus in the emergency department. Emergency Medicine. 2018;50:6-16.
12. Ezehnilavan S, Priyamvada PS, Haridasan S, Rajesh NG, Parameswaran S. Acute gastric dilatation in a patient with lupus nephritis: An uncommon presentation of lupus mesenteric vasculitis. Saudi J Kidney Dis Transpl 2018;29:429-34.
13. Tian XR, Zhang X. Gastrointestinal involvement in systemic lupus erythematosus: Insight into pathogenesis, diagnosis and treatment. World J Gastroenterol 2010;16:2971-7.
14. Bringer J, Richard JL, Ribstein J, Barinéon G, de Seguin C, Béraud JJ, et al. Recurrent ileitis in disseminated lupus erythematosus. Therapeutic and nutritional problems. Rev Med Interne 1981;2:61-6.
15. Endo H, Kondo Y, Kawagoe K, Ohya TR, Yanagawa T, Asayama M, et al. Lupus enteritis detected by capsule endoscopy. Intern Med 2007;46:1621-2.
16. Ko SF, Lec TY, Cheng TT, Ng SH, Lai HM, Cheng YF, et al. CT findings at lupus mesenteric vasculitis. Acta Radiol 1997;38:115-20.
17. Anand A, Malur K, Kawale J, Nadkar MY. Mesenteric vasculitis in a case of systemic lupus erythematosus. J Assoc Physicians India 2016;64:70-3.
18. Glijn N, Korstwagen LA, Lam-Tse WK. Systemic lupus erythematosus (SLE): An unusual cause of ileocolic intussusception. BMJ Case Rep 2017;2017:bcr2017220185.
19. Fanouriakis A, Kostopoulou M, Alunno A, Aringer M, Bajema I, Boletis JN, et al. Overview of the management and prognosis of systemic lupus erythematosus in adults. Ann Rheum Dis 2019;78:736.
20. 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;5:273-81.