

## Case Report

# Lupus enteritis as a flare up of systemic lupus erythematosus- a case report and review

Nikitha Janga, Jagadeesan M.\*, Kavitha M. M., Kannan R.

Department of General Medicine, Saveetha Medical College Hospital, Chennai, Tamil Nadu, India

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**\*Correspondence:**

Dr. Jagadeesan M.,

E-mail: [drjagadeesan@gmail.com](mailto:drjagadeesan@gmail.com)

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### ABSTRACT

Systemic lupus erythematosus (SLE) generally affects young to middle-aged women, commonly presenting as a triad of fever, rash, and joint pain. Abdominal pain is a common symptom in patients with SLE. The leading causes of abdominal pain in SLE are lupus enteritis, pancreatitis, pseudo-obstruction, acalculous cholecystitis, mesenteric thrombosis, hepatic thrombosis, medications like (NSAIDs, MMF, steroids, HCQ), colon perforation. The incidence of abdominal pain in patients with SLE ranges from 8-40%, and the commonest cause is lupus enteritis. The following case describes a young woman presenting with lupus enteritis as a manifestation of SLE, the importance of early disease recognition, utilities of abdominal computed tomography (CT) in diagnosis, and current treatment protocols for lupus enteritis.

**Keywords:** SLE, Lupus enteritis, Abdominal pain

### INTRODUCTION

Lupus enteritis is defined as the inflammation of the small bowel and its vasculature, with supportive image and/or biopsy findings. Review of literature revealed the incidence of lupus enteritis or mesenteric vasculitis to be 0.2% to 53% and was correlated with a mortality rate as high as 53% if complicated with hemorrhage, ulceration, infarction, or perforation, or if treatment or diagnosis is delayed.<sup>1</sup> The clinical picture of lupus enteritis is often nonspecific, with mild to severe abdominal pain, diarrhea, and vomiting being the cardinal manifestations. Lupus enteritis most commonly affects the jejunum and ileum. The rectum is rarely involved, because of collateral circulation mechanism for lupus enteritis is complement activation. The activation of complement system can promote diffuse microvascular injury and increased vascular permeability. Lupus enteritis is seldom confirmed on histology, making CT the gold standard for diagnosis.<sup>2</sup> The initial treatment of lupus enteritis is generally high dose intravenous methylprednisolone,

immunosuppression with azathioprine or cyclophosphamide. The following case describes a young woman presenting with lupus enteritis as a manifestation of SLE.

### CASE REPORT

A 28-year-old woman known case of SLE on remission for past one and half years presented with 3 days history of abdominal pain and vomiting for 2 days and a history of diarrhea for 2 days. The patient described the pain as diffuse, constant, and dull in intensity, without any alleviating factors or associated symptoms. The patient was treated in a nearby hospital as drug-induced gastritis with intravenous pantoprazole 40 mg twice a day for 3 days. Because of persistent pain, the patient reviewed with us. Considering her baseline disease of lupus, we suspected lupus enteritis and proceeded with CT abdomen with contrast studies. Contrast enhanced CT (CECT) abdomen showed smooth circumferential wall thickening of entire small bowel loops with mild distention, maximal

luminal diameter measuring 2.5 cm with a bowel wall thickening of 6 mm (Figure 1), increase in mesenteric vasculature (Figure 2) along with free fluid in the Morrison pouch and the pelvis suggestive of lupus vasculitis involving small bowel territories and ascites



**Figure 1: CT of abdomen of bowel loops with edema also referred to as target sign.**



**Figure 2: CT of engorgement of mesenteric vessels known as comb sign.**

Line immune assay was strongly positive for SSA, SSB, and ds-DNA. Her complements were low (C3 58 mg/L (normal range-88-206 mg/dl), C4 8 mg/L (normal range-12-72 mg/dl)). The patient was then treated with intravenous fluids, intravenous antibiotics, and pulse methylprednisolone 500 mg IV/day for 5 days. The patient's abdominal pain showed dramatic improvement within 36 hours of initiating steroids and complete resolution of abdominal pain was observed. The patient was discharged with a tapering dose of steroids,

mycophenolate mofetil (500 mg TDS), and hydroxychloroquine (200 mg HS).

She was diagnosed with SLE when she presented with mono neuritis multiplex. There were no mucocutaneous or constitutional manifestations during that phase. Complete blood count was within normal limits with an erythrocyte sedimentation rate of 95 mm/hr and C-reactive protein was negative. Her ANA-Hep2 done by IIF method showed a 3+ speckled pattern, with 4+intensity. Corresponding line immune assay was strongly positive for, ribosomal P protein (PO), SS-A /RO60, and SS-A/RO52. Her complements were low C3 20 mg/L (80 to 160) C4 was 7 mg/L (20 to 40). She was treated with pulse methylprednisolone, followed by 0.75 mg/kg BW oral prednisolone which was slowly tapered, and mycophenolate mofetil (2.5 gm) as immunosuppressants along with hydroxyl chloroquine (200 mg HS). She was on complete remission for one and a half years. The patient was off steroids for the past 6 months, MMF at the maintenance of 750 mg for the past 1 year, and her complements were normal.

**DISCUSSION**

Lupus enteritis presents with signs and symptoms, such as abdominal pain, ascites, nausea, vomiting, diarrhea, and fever. In the study of major gastrointestinal manifestations in SLE, prevalence of LE was reported as 0.2-6.4% in the Western literature and estimated to be 3.4-5.8% in SLE patients in Asia.<sup>3-6</sup> Ko et al reported 11 of 15 cases (73.3%) had CT features of LE as the clinical suspicion for this complication was high in their patients.<sup>7</sup> It may be concluded that LE is the most common cause of acute abdominal pain severe enough to have admissions and imaging studies in SLE patients both in Asia and in the West.<sup>8</sup>

The pathogenesis is unclear but has been attributed to immune-complex deposition and complement activation, with subsequent submucosal edema. Biochemical investigations shall aid in the diagnosis of lupus enteritis. Our patient had strongly positive SSA, SSB, dsDNA, and decreased complement levels.

CT scan of the abdomen with contrast is considered the gold standard. The classic findings suggestive of lupus enteritis in CT are bowel wall thickening greater than 3 mm, known as target sign, engorgement of the mesenteric vessels known as combs sign, and increased attenuation of mesenteric fat. Target sign is also seen in other conditions such as intestinal angioedema, mesenteric vein thrombosis, inflammatory bowel disease, and intestinal infections. Arterial narrowing and distended loops of the bowel can be found out by arteriography. One should always consider mesenteric vasculitis in the differential diagnosis as overlooking the diagnosis can have grave consequences.<sup>9</sup>

Lupus enteritis can lead to intestinal necrosis and perforation if untreated. Signs of perforation may be subtle and masked in patients taking steroids; therefore, abdominal pain in a lupus patient must be addressed and evaluated. Gastrointestinal visualization by endoscopy is usually not helpful nor it is necessary in making the diagnosis of lupus enteritis since only superficial tissue is analyzed.

Steroids are usually considered in the first-line management of lupus enteritis. For patients with mild abdominal pain oral prednisolone 0.5-1 mg/kg per day is given and in patients with severe abdominal pain pulse therapy of methylprednisolone 250 mg to 1 g IV daily followed by oral prednisolone 0.5-1 mg/kg per day is given. Patients who do not respond to pulse dose steroids, IV cyclophosphamide (1 mg/kg) daily, or mycophenolate can be considered. Additional, rituximab has been effective in severe relapsing lupus enteritis in some cases reports.<sup>10</sup>

When a rapid response to immunosuppressive therapy is not achieved, surgical intervention (laparotomy) for possible bowel perforation or large area of ischemia should be considered. We were able to control her symptoms with pulse methylprednisolone 500 mg IV/day for 5 days. Her abdominal pain showed dramatic improvement within 36 hours of initiating steroids and complete resolution of abdominal pain was observed. Even if patients respond initially to steroids, there is a high predilection for recurrence (23%).

A predictor of the risk of recurrence for lupus enteritis is bowel wall thickness greater than 9 mm and the recurrence rate of lupus enteritis correlates with a lower cumulative dosage of prednisolone and a shorter duration of treatment.<sup>11</sup>

Prognosis is generally excellent for patients with lupus enteritis given its good response to steroids.

## CONCLUSION

Lupus enteritis is a rare manifestation of SLE. whenever an SLE patient presents with abdominal pain a high suspicion of lupus enteritis should be considered as a differential diagnosis. Diagnosis of lupus enteritis requires a combination of high clinical suspicion from symptoms, laboratory testing, and imaging. Treatment usually depends on severity. In this moderately severe lupus enteritis, high-dose steroids were an effective initial treatment.

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