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# A rare case of lupus enteritis

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**Abstract**
Systemic Lupus Erythematosus (SLE) is an autoimmune disorder with protean manifestations involving multiple organs. SLE can present as a benign disorder with skin and joint involvement to a life-threatening disorder with renal or central nervous system disorder. We report a rare case of Lupus Enteritis in a young female patient.

**Keywords:** lupus enteritis, Systemic Lupus Erythematosus (SLE)

## Introduction

Systemic Lupus Erythematosus (SLE) is an enigmatic disorder aptly called "disease of thousand faces." It has varied clinical and laboratory manifestations with a plethora of autoantibodies. The first case of SLE was reported in India in 1955 [1]. Prevalence was said to be 3.2/100000 of the population in rural area-based study done in 1993. Gastrointestinal symptoms are common in SLE patients, and more than half of them are caused by adverse reactions to medications and viral or bacterial infections. Though not as common as lupus nephritis, SLE-related gastrointestinal involvement is clinically important because most cases can be life-threatening if not treated promptly. Lupus mesenteric vasculitis is the most common cause, followed by protein-losing enteropathy, intestinal pseudo-obstruction, acute pancreatitis and other rare complications such as celiac disease, inflammatory bowel diseases, *etc*.

## Case Report

22 years old female, a k/c/o SLE for last 3 years admitted with history of intractable vomiting for last 3 days. At the onset of illness patient had watery diarrhea for one day followed by abdominal distension.

On general physical examination patient was conscious and oriented to time place and person. She had tachycardia with pulse rate of 124/min, supine blood pressure of 96/60 mm of Hg, afebrile and maintaining 98 % of oxygen saturation at room air. Her abdomen was distended and tender on deep palpation with tympanic note on percussion all over the abdomen with sluggish bowel sounds. Keeping possibility of acute intestinal obstruction her abdominal X rays were obtained which revealed dilated gut loops involving both small and large intestine. Ultrasound abdomen showed dilated gut loops with gross ascites with bilateral hydroureteronephrosis and distended gall bladder. CECT abdomen was done which revealed bowel wall edema, abnormal bowel-wall enhancement (double halo or target sign), dilatation of bowel lumen and ascites along with engorgement of mesenteric vessels, increased number of visible vessels (comb sign). Ds DNA was positive with C3: 40 (84-168) mg/dl & C4: 7.8 (15-50) mg/dl which indicated SLE Flare.

Keeping diagnosis of SLE flare with Lupus Enteritis with Intestinal Pseudo obstruction with bilateral hydroureteronephrosis patient was given methyl prednisolone pulse along with usual care. Patient started improving on Day 4$^{th}$ after steroid pulse and was discharged on day 10$^{th}$ of admission.

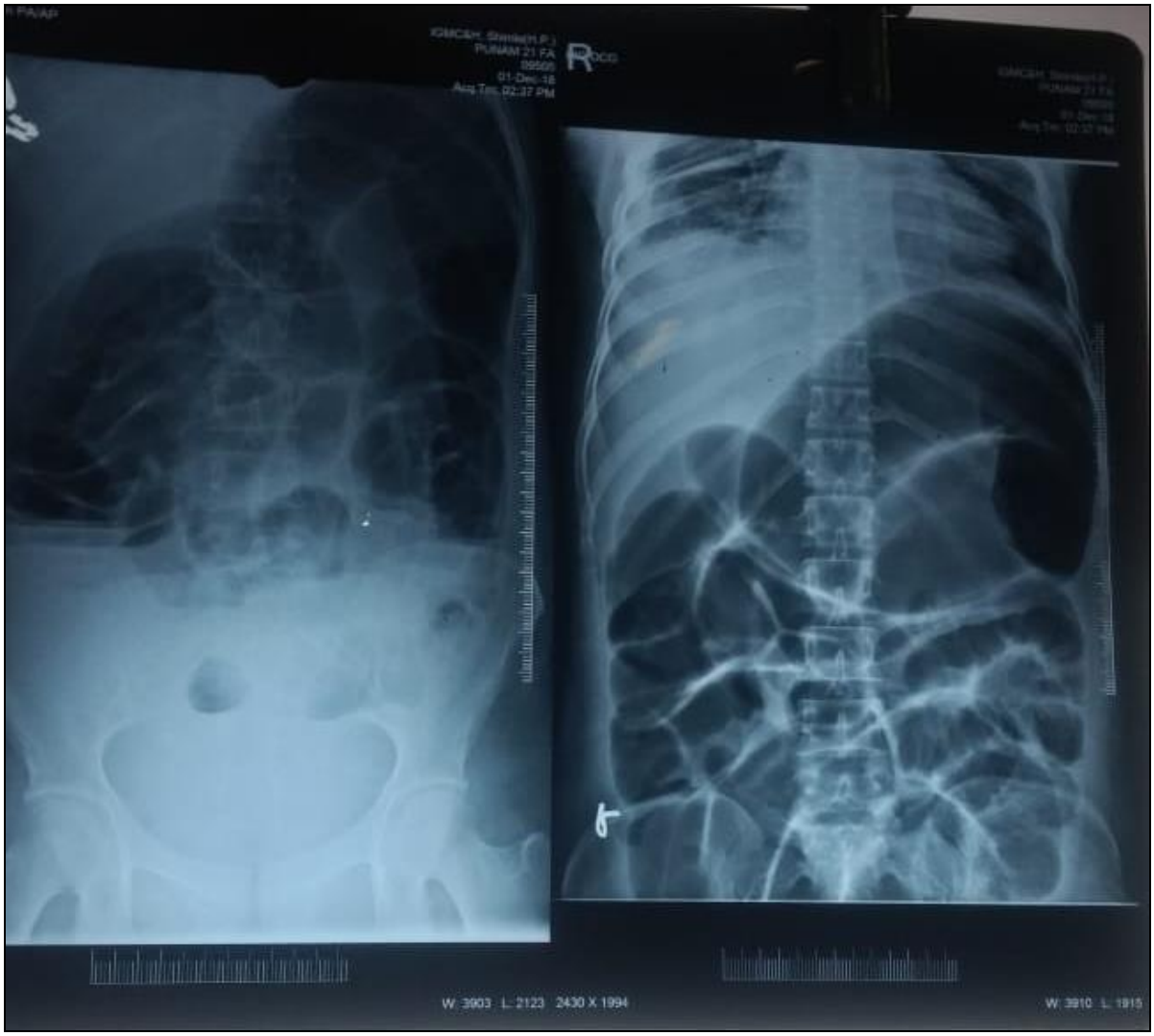

**Fig 1:** Abdominal Xray showing dilated small and large bowel loops.

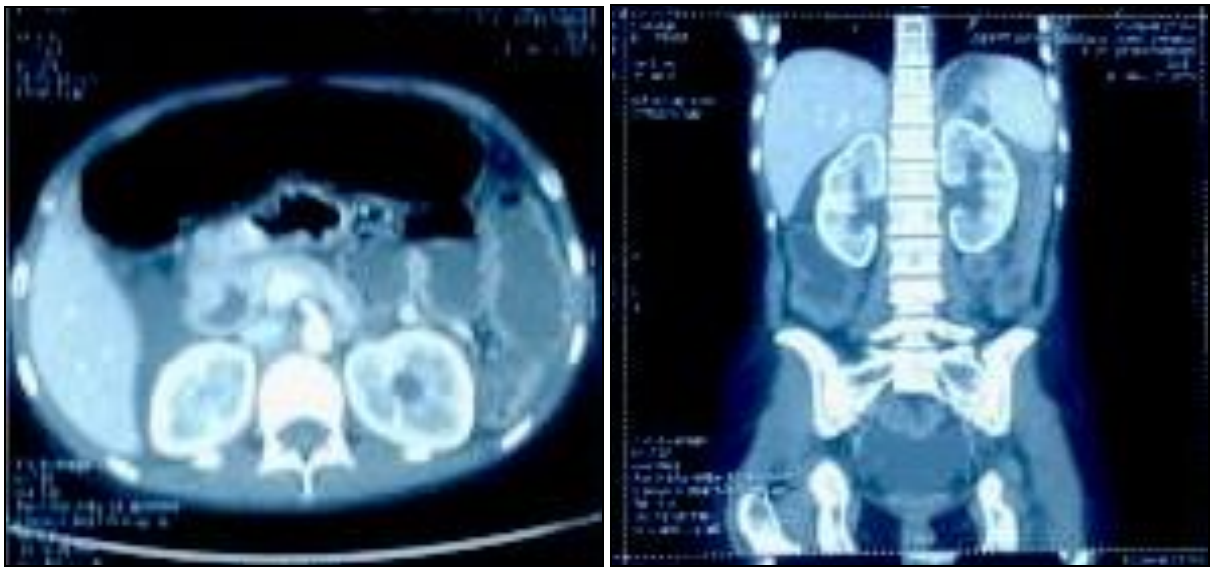

**Fig 2:** Sagittal and Coronal Plane of CECT abdomen showing bilateral hydronephrosis.

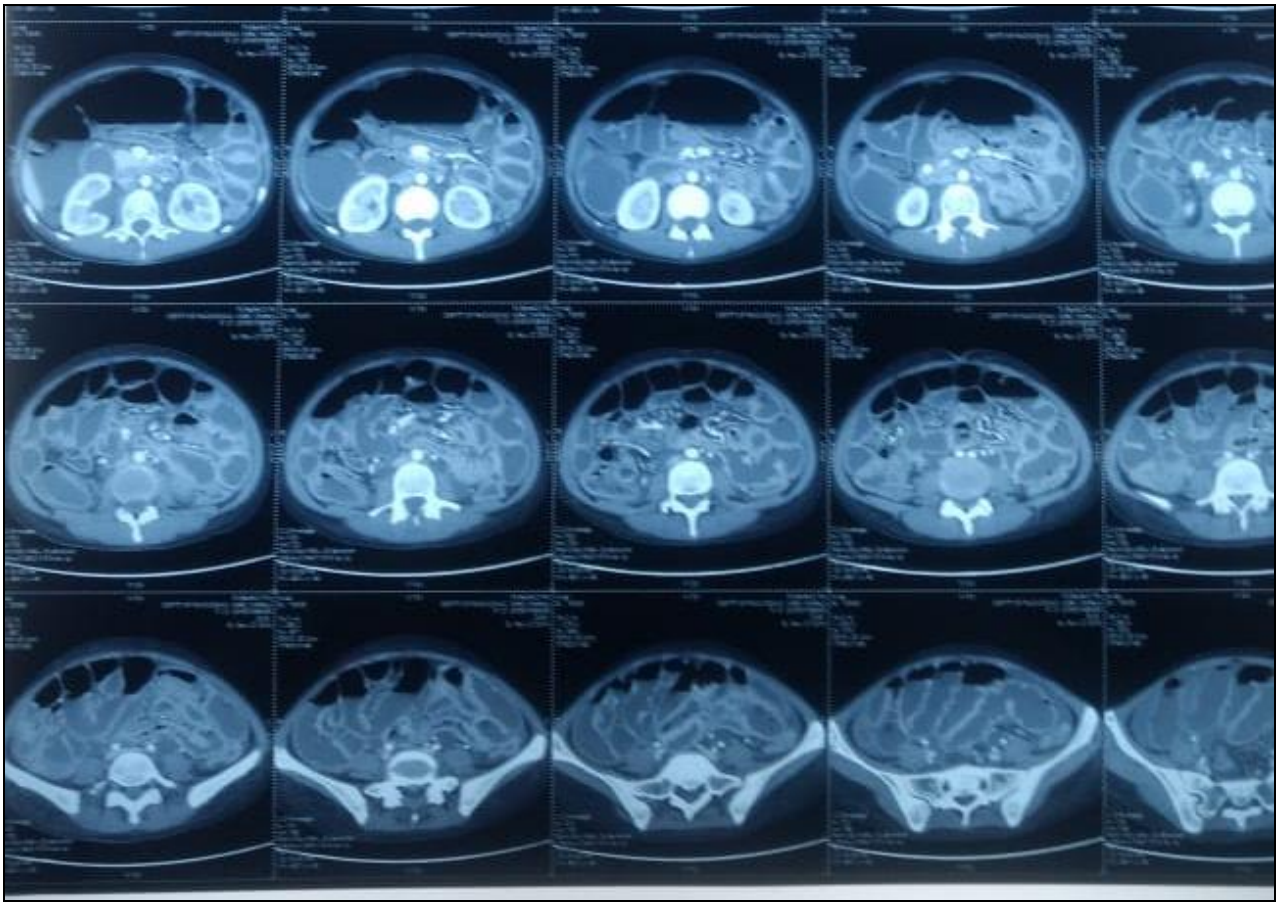

**Fig 3:** Multiple Sagittal sections of CECT abdomen at different levels showing dilated small and large bowel loops, target sign and increased mesenteric vascularity.

## Discussion

A number of terms have been used to describe lupus enteritis, including mesenteric arteritis, lupus enteritis, lupus arteritis, lupus vasculitis, gastrointestinal vasculitis, intra-abdominal vasculitis and acute gastrointestinal syndrome. Lupus enteritis is one of the main causes of acute abdominal pain in SLE patients. About 8%-40% of SLE patients have acute abdominal pain during the stage of active disease [1], but Lupus Enteritis is generally an uncommon condition in SLE patients. In Asia, the reported overall prevalence of Lupus enteritis in patients with SLE is 2.2%-9.7% [2-4]. Ju *et al* [5] reported that the global prevalence of LMV ranges from 0.2% to 9.7% among all SLE patients and from 29% to 65% in patients who had acute abdominal pain. The predisposing factors for LMV are unknown. The proposed trigger factors include bacterial infections that lead to changes of intestinal flora, cytomegalovirus infection, eosinophilia, non-steroidal anti-inflammatory drugs, chemicals, metallic particulates, animal viruses, helminth infection, caffeine, phosphodiesterase-4-inhibitors, adenosine diphosphate, certain foods and herbal medicines [4]. Inflammatory vasculitis secondary to immune complex deposition and thrombosis of the intestinal vessels secondary to circulating anti-phospholipid antibodies are the proposed pathogenic mechanisms of LMV [6-9]. Macroscopically, the appearance of LMV varies from segmental oedema to ulceration, gangrene and perforation [5]. Both small arteritis and venulitis are found in LMV. Microscopically, fibrinoid necrosis of subserosal vessels and leukocytoclasis on the vascular wall, as well as edematous submucosa with mild diffuse inflammatory infiltration of mononuclear cells, can be observed. In the muscular layer, intravascular fibrin thrombus and hemorrhage around the small veins can be found. Both the inflammatory and thrombotic mesenteric vasculopathy of LMV can cause mesenteric ischemia. LMV can cause very severe abdominal symptoms and signs and sometimes is diagnosed as acute surgical abdomen. Typically, the abdominal pain caused by LMV is diffuse in pattern, in some cases accompanied by rebound tenderness and abdominal muscle guarding. The symptoms of LMV vary from mild, nonspecific abdominal pain, bloating or loose stool, to necrosis and intestinal perforation which manifest as severe extensive gastrointestinal bleeding or acute surgical abdomen. It is noteworthy that in LMV patients with bowel perforation, the typical signs can be absent. Other manifestations of LMV include anorexia, nausea, vomiting, dysphagia, hematemesis, postprandial fullness, diarrhea and melena.

## Diagnosis

Accurate diagnosis of LMV is critical to allow prompt treatment to avoid unnecessary surgical intervention. As the clinical symptoms and laboratory parameters are non-specific, and bowel specimens are not always available, the diagnosis of LMV relies on abdominal computed tomography (CT) scan which allows both the bowel wall and the abdominal vasculature to be visualized. Advances in CT technology have been extremely helpful in detecting ischemia and for evaluating the causes of abdominal pain. Common CT findings in patients with LMV include dilated bowel, focal or diffuse bowel wall thickening, abnormal bowel wall enhancement which is also called “target sign”,

mesenteric edema, stenosis or engorgement of mesenteric vessels which is also called "the comb sign" and ascites (Figure (Figure1).1). Segmental or multifocal involvement of the small and large bowel loops with intervening normal bowel segments indicates ischemic change, which is almost always indicative of vasculitis. Ultrasonography is also useful for both the diagnosis and follow-up of LMV. Small intestinal wall oedema and thickening can be visualized under ultrasonography. Irregular thickening and projection of folds in multiple segments of the duodenum and the terminal ileum accompanied by "thumb print" signs in double-contrast radiography suggest ischemic changes. Gastroscopy and colonoscopy can show ischemia and ulcerative changes. However, endoscopy-guided biopsy might not yield a definitive diagnosis of LMV because the affected vessels are usually located in an inaccessible area. Laparoscopy can be used in the diagnosis of LMV [8, 9].

**Management**

Early diagnosis and appropriate intervention can avoid potentially fatal complications of LMV. Because the primary lesions of LMV are inflammatory ischemic vasculitis, immediate and aggressive anti-inflammatory immunosuppressive treatment should be initiated as soon as the diagnosis of LMV is made. The treatments include high dose intravenous infusion of methylprednisolone or an equivalent agent and complete bowel rest. For patients with recurrent LMV and those who do not have adequate response to intravenous prednisolone alone, intravenous cyclophosphamide should be initiated. The initial dosage of cyclophosphamide is 1 mg/kg daily and the dosage is gradually tapered when LMV is stabilized [6]. They suggested that for patients with high risk of recurrence, immunosuppressive agents should be initiated as early as possible. When a rapid response to immunosuppressive therapy is not achieved, surgical intervention for possible bowel perforation or large area of ischemia should be considered. Early laparotomy within 24 to 48 h is critical for improving the prognosis of LMV patients.

**Prognosis**

The prognosis of LMV varies in reports from different areas of the world. This might be due to genetic differences. Reports from Europe and North America indicate that the prognosis of LMV is poor and some fatalities can occur. Some reports claimed that the mortality of LMV could be as high as 50% [6]. The prognosis of LMV depends on the extent of vascular involvement, the prompt implementation of immunosuppressive therapy, and the time of surgical intervention. On the basis of published reports, the prognosis of LMV can be improved if abdominal CT is used to aid the diagnosis and if prompt immunosuppressive therapy is implemented.

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