

Lupus Enteritis: A Rare Manifestation of SLE A Case Report

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Abstract: Systemic lupus erythematosus is a multifactorial chronic autoimmune disease which may affect almost any organ. Gastrointestinal Systems are common in systemic lupus erythematosus. they are usually attributable to infections medicine side effects, and other underlying conditions. In rare cases, they are caused by the autoimmune process itself. Lupus enteritis can be the manifestation of SLE flare. Although involvement are more common, patient may presents to us with peritoneal involvement which warrants urgent investigations and treatment and it is associated with grave prognosis. High index of suspicion should be maintained when evaluating SLE patients presenting with gastrointestinal symptoms to prevent diagnosis and treatment delays that could lead to serious complications such as bowel necrosis, perforation. This case report is reminder to physicians regarding Lupus enteritis. CT scan of abdomen is diagnostic modality of choice. Manifestations usually improve promptly to corticosteroid therapy. High index of suspicion should be maintained when evaluating SLE patients presenting with gastrointestinal symptoms to prevent diagnosis and treatment delays that could lead to serious complications such as bowel necrosis, perforation

Keywords: Systemic Lupus Erythematosus, Lupus nephritis, gastrointestinal symptoms, SLE flare, Perforation, immune complexes

1. Introduction

Systemic Lupus Erythematosus (SLE) is multisystem autoimmune disease in which organs and cells undergo damage initially mediated by tissue binding auto antibodies and immune complexes. Its clinical course is usually fluctuating, characterized by intermittent flares involving any organ or system of the body. Gastrointestinal symptoms are present in upto 40 to 50% patients of SLE^{[1][2]}. Nevertheless, they can be secondary to infections, medication use, protein losing enteropathy, inflammatory bowel disease associated celiac disease, pseudoobstruction, pancreatitis, SLE enteritis. Term lupus enteritis refers to inflammation of bowel wall due to SLE activity. Pathogenesis may include immune complex deposition in the bowel wall or small vessel vasculitis^[1]. Pain in patients of SLE might reflect the underlying vasculitis or thrombosis which can lead to ischemia and perforation if not promptly treated. Nausea, sometimes with vomiting, diarrhea can be manifestation of SLE flare^[4]. Current knowledge of lupus enteritis is limited based on case reports and series. This case report may be eye opener to physician dealing with gastrointestinal manifestations in patients of SLE as perforation and peritonitis can be missed.

2. Case Presentation

A 33 year old female patient known case of lupus (had multiple flare episodes in past) with hyperthyroidism brought by relative to the hospital with chief complaints of Loose stools multiple episodes since 5-6 days, Multiple episodes of vomiting since 5-6 days, Pain in abdomen since 2 days, Loss of appetite since 5-6 days. The patient was apparently all

right 6 days prior to admission when she developed loose stools which were sudden in onset 7-8 episodes per day watery in nature foul smelling associated with non bilious vomiting 4-5 episode per day and pain in abdomen generalized non radiating dull in nature associated with loss of appetite.

There was no complains of fever, joint pain and swelling, blood or mucus stained stools, breathlessness, convulsions, no abnormal psychiatric behavior. Patient gave uneventful obstetric history. The patient was a diagnosed case of cutaneous lupus on hydroxychloroquine 200mg BD and Omnacortil 5mg OD taking irregularly. She was also a known case of hyperthyroidism on Neomercazole 5mg OD. On examination patient was conscious oriented obeying verbal commands. She had mild pallor hyperpigmented patches on face, abdomen, both arms which were non itchy maculopapular with non raised edges. There was no cyanosis, no icterus, no edema, no lymphadenopathy. Her abdomen was soft diffusely tender, but there was guarding and no rigidity.

Surgical opinion was taken. Her Total Leucocyte Count 14400, Pletlet 1.88 lakh, Neutrophils 91%, ESR was 40, CRP was 2 mg/dl. Her differentiated diagnosis that we entertained were infective gastroenteritis, lupus enteritis, lupus pancreatitis and protein losing enteropathy, pseudoobstruction. Patient was thought to have acute gastroenteritis of infective etiology and was started on antibiotics, probiotics, and intravenous fluids. After 2 days on this line of treatment patients loose stools and vomiting and pain in abdomen persisted and rash increased in size and severity. Higher antibiotics started. But despite patient did

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not improved and her ESR and CRP was also high (ESR 30, CRP 1.8mg/dl). Ultrasonography was done that was suggestive of loculated collection in pouch of Douglas approx. 40 cc, inflamed edematous thickened bowel loops suggestive of? ileitis? primary peritonitis? Sealed off perforation. Patient developed fever and diagnosis of SLE enteritis was thought So started on intravenous steroids. Patient responded to steroids she had no stools. Contrast enhanced Computed Tomography scan was done suggestive of loculated collection abutting and displacing the terminal ileum and extravasation of contrast through it suggestive of sealed off perforation. Finally Patient was transferred to surgery and on exploratory laparotomy a single ileal perforation of size 1cm *1cm *3cm proximal to the Ileocecal junction was found and resection and anastomosis was done. But, unfortunately, patient died during surgical follow up

3. Discussion

Systemic lupus erythematosus (SLE) is a worldwide chronic autoimmune disease which may affect every organ and tissue. Genetic predisposition, environmental triggers, and the hormonal milieu, interplay in disease development and activity. Clinical manifestations and the pattern of organ involvement are widely heterogeneous, reflecting the complex mosaic of disrupted molecular pathways converging into the SLE clinical phenotype^[6]. The potential severity of SLE-related GI manifestations is concerning, considering that more than 50% of SLE patients develop GI symptoms at some point during the course of illness.^[7] The incidence and prevalence of GI involvement during the course of SLE disease vary widely. This could be due to less attention being paid to GI manifestations than other organ symptoms, such as lupus nephritis. According to an autopsy study, 60–70% of SLE patients had evidence of peritonitis, whereas only 10% showed clinical manifestations throughout their lives^[8]. Lupus enteritis is a rare and poorly understood cause of abdominal pain in patients with systemic lupus erythematosus^[9]. Its clinical symptoms mostly includes abdominal pain (97%), vomiting (42%), diarrhea (32%) and fever (20%)^[9]. Abdominal CT scan is considered the first-line diagnostic modality^[11]. There are three classic findings: bowel wall edema and enhancement (“target sign”), engorgement/increased number of mesenteric vessels (“comb sign”), and increased attenuation of mesenteric fat^[10,11]. Ascites is also commonly present. These findings are nonspecific and can also be seen with other conditions, such as intestinal obstruction, pancreatitis, or inflammatory bowel disease^[10]. Coordination with the radiology department is important as the radiologist might not be aware of the presence of SLE in clinical history, which can lead to delayed diagnosis. Nonspecific findings like mucosal edema, erosions, and ulcers have been reported to be associated with lupus enteritis^[12]. First-line treatment includes systemic corticosteroids. Appropriate bowel rest, hydration, and electrolyte repletion are essential supplementary therapies. Most patients experience symptomatic improvement with corticosteroid therapy^[10,12]. Appropriate bowel rest, hydration, and electrolyte repletion are essential supplementary therapies.

The most serious complications are life-threatening and

include bowel wall ischemia and perforation. Suspicion for perforation should be high and warrants close clinical and radiological monitoring since systemic corticosteroids can mask signs of peritonitis^[2]

Bowel infarction and perforation in patients with SLE are often caused by acute ischemic enteritis of the small intestine, which is also described as lupus mesenteric vasculitis^[13]. Lupus mesenteric vasculitis is considered to be caused in patients with SLE by circulating autoantibodies that form an immune complex deposition in blood vessels, which can lead to the development of vasculitis and thrombosis of the vessels supplying the intestine. An inadequate blood supply to the intestine results in ulceration, infarction and perforation^[13,14]. The prognosis of SLE patients with intestinal perforation is poor. Therefore, early diagnosis and appropriate interventions are crucial to the management of the disease. The majority of SLE cases have been found to be long-term users of corticosteroids and immunosuppressants, which are known to weaken the immune system^[16,17]. In our case patient SLE patient presented us as acute gastroenteritis diagnosed as SLE enteritis but eventually died due to intestinal perforation. Mortality of perforation is upto 2.7%^[9]. Takashi et al. found half of the patients (6 of 11 patients) with SLE had intestinal perforation as a complication and died^[18]

4. Conclusion

Lupus enteritis occurs in only about 0.2 to 5.8% of patients with SLE and is vasculitis or inflammation of small bowel wall. It is rarer for it to be the only clinical feature of SLE flare. Dilemma exists between a rare diagnosis of sealed off perforation peritonitis in SLE and even rarer diagnosis of SLE enteritis. But physical signs of bowel perforation and ischemia are often masked by medication with corticosteroids and immunosuppressants. High index of suspicion should be maintained when evaluating SLE patients presenting with gastrointestinal symptoms to prevent diagnosis and treatment delays that could lead to serious complications such as bowel necrosis, perforation even death.





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