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From Symptoms to Solution: Case Series on Lupus Enteritis from a Tertiary Care Center from South India

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ABSTRACT
This study aimed to analyze the clinical, laboratory, imaging and treatment outcomes of eight patients diagnosed with lupus enteritis (LE) to better understand its presentation and prognosis. It was a retrospective case series examining the demographic, symptomatic, laboratory and radiological characteristics of LE in patients with systemic lupus erythematosus (SLE). The results showed that all patients presented with abdominal pain, with common additional symptoms including diarrhea and abdominal distension. Laboratory findings highlighted active lupus, including low complement levels, anemia, leukopenia and thrombocytopenia. Imaging showed ascites, bowel wall edema and the target sign, which are characteristic of LE. Treatment involved high-dose corticosteroids and immunosuppressive therapy, with 75% of patients recovering and 25% dying, indicating a significant mortality risk. The study concluded that lupus enteritis is a severe, potentially life-threatening complication of SLE that requires prompt recognition and aggressive immunosuppressive therapy to improve outcomes.

INTRODUCTION

Abdominal pain is a common and often challenging clinical presentation among patients with systemic lupus erythematosus (SLE), associated with a broad range of underlying causes that may be SLE-related or unrelated to the disease itself. These causes include classic abdominal pathologies, as well as complications specific to the disease's pathophysiology and treatment. Among SLE-related causes, lupus enteritis (LE) stands out as a potentially severe, yet rare complication characterized by inflammation or vasculitis within the small bowel. The pathogenesis of LE is thought to involve immune complex deposition and complement activation, resulting in submucosal edema and other forms of gastrointestinal injury[1,2]. According to the British Isles Lupus Assessment Group (BILAG) 2004 criteria, LE is defined as vasculitis or inflammation specifically affecting the small bowel, with confirmation typically supported by imaging modalities such as computed tomography (CT) or histopathological biopsy findings[3]. CT imaging is particularly critical for diagnosing LE, as it can identify characteristic signs, including bowel wall thickening and vascular compromise, which are essential for differentiating LE from other causes of abdominal pain in SLE[1,2]. Beyond LE, SLE patients may experience abdominal pain related to disease-specific complications such as pancreatitis and intestinal pseudo-obstruction, as well as infections secondary to immunosuppressive therapies[2]. The exact prevalence of lupus enteritis remains unclear, with varying reports describing it as a common gastrointestinal manifestation in SLE patients, while other sources label it as rare[1]. Additionally, inconsistent terminology-such as mesenteric arteritis, intestinal vasculitis, lupus peritonitis and abdominal serositis-complicates the understanding and recognition of LE[3]. This study presents eight new cases of lupus enteritis managed at a tertiary care center in South India. It aims to elucidate the epidemiology, clinical characteristics, serological profiles and outcomes associated with LE further to enhance understanding and improve management strategies for this poorly defined yet significant cause of abdominal pain in SLE patients.

MATERIALS AND METHODS

This case series includes eight patients diagnosed with lupus enteritis (LE) managed at the Department of Clinical Immunology and Rheumatology, Sri Ramachandra Institute of Higher Education and Research (SRIHER), Chennai. Data were collected retrospectively over the past six years (2018-2024) to evaluate patient demographics, clinical features, laboratory parameters, imaging findings, treatments, and outcomes in SLE patients with abdominal pain indicative of LE.

Inclusion and Exclusion Criteria: The study included patients who met the American College of Rheumatology (ACR) 2019 criteria for SLE and presented with abdominal pain and CT findings suggestive of LE. Abdominal CT was essential to confirm features consistent with LE, such as bowel wall thickening and characteristic inflammatory signs. Patients with infectious causes of enteritis or other non-SLE-related gastrointestinal issues were excluded to ensure a focus on lupus-related pathology.

Data Collection: Collected variables were extracted from patient records, focusing on the following:

- **Demographic Data:** Age, sex and duration of SLE diagnosis before the onset of LE.
- **Clinical Features:** Documented symptoms included abdominal pain, diarrhea, abdominal distension, fever and acute cutaneous lesions.
- **Laboratory Data:** Laboratory markers evaluated included complement levels (C3, C4), anti-dsDNA antibody levels, direct Coombs test (DCT) positivity, hemoglobin levels (for anemia), leukocyte counts (for leukopenia), platelet counts (for thrombocytopenia) and the presence of proteinuria.
- **Imaging Findings:** Abdominal CT findings were reviewed in detail for each case, focusing on features characteristic of LE, including:
- Bowel wall edema.
- Ascites.
- Target sign.
- Comb sign (mesenteric vessel engorgement).
- Surrounding fat stranding.
- **Treatment:** All patients received corticosteroids as first-line therapy. Additional immunosuppressive agents, such as cyclophosphamide, were used in most cases, with one patient receiving rituximab.
- **Outcomes:** Outcomes documented included patient survival, complication rates (such as intestinal gangrene) and recurrence.

This collection of clinical and imaging variables provides a detailed overview of LE presentation and treatment response, aiming to contribute further insights into managing this rare gastrointestinal manifestation in SLE patients.

Demographics: This case series includes eight patients diagnosed with lupus enteritis (LE). Out of these, seven were female (87.5%) and one was male (12.5%), reflecting a higher prevalence in females. The average age at presentation was 30 years, with an age range of 17-55 years.

Clinical Features: The predominant symptom among all patients was abdominal pain, present in 100% of cases. Other symptoms included diarrhea and abdominal distension, each seen in 50% of patients. Fever and acute cutaneous lesions were less common, present in 25% of cases each.(Fig. 1).

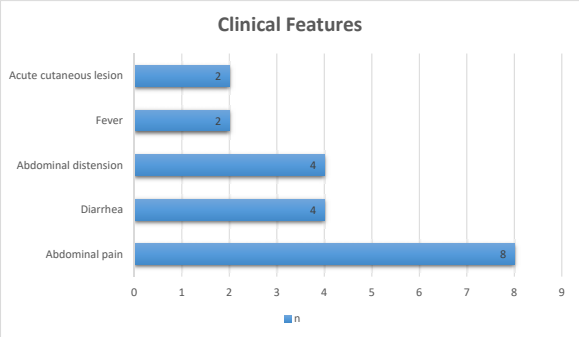

Fig. 1: Clinical Features

RESULTS AND DISCUSSIONS

Laboratory Findings: Laboratory data revealed several abnormalities characteristic of active SLE. Decreased complement levels were observed in the majority, with low C3 levels in 75% of patients and low C4 levels in 25%. Direct Coombs test (DCT) positivity and anemia were also prominent, found in 75% of patients. Leukopenia, thrombocytopenia and proteinuria were each observed in half of the cases. Elevated anti-dsDNA antibodies were noted in 25% of patients. (Table 1).

Table 1: Laboratory Findings

Laboratory Finding	n	%
↓C3	6	75
↓C4	2	25
↑Anti-dsDNA	2	25
DCT positivity	6	75
Anemia	6	75
Leukopenia	4	50
Thrombocytopenia	4	50
Proteinuria	4	50

Imaging Findings: Abdominal CT findings provided critical diagnostic support for LE. The most frequent imaging findings were:(Fig. 2).

- **Ascites** in all patients (100%).
- **Target sign** in 75%.
- **Bowel wall edema** in 62.5%.
- **Comb sign** (engorgement of mesenteric vessels) in 50%.
- **Surrounding fat stranding** in 37.5%.

One patient (14%) also had concurrent cystitis and hydroureteronephrosis.

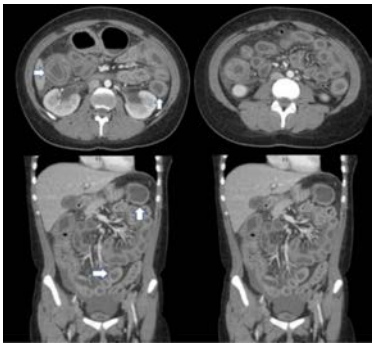

Fig. 2: CT-Abdomen Arrows are Showing Target Signs

Treatment and Outcomes: All patients received corticosteroids as the first-line treatment, with additional immunosuppressive therapy in most cases. Cyclophosphamide was administered in 62.5% of patients, while one patient received rituximab. Among these cases:

- **Six Patients:** (75%) recovered and are in regular follow-up.
- **Two Patients:** (25%) died, indicating a significant mortality risk associated with severe LE.
- **One Patient**: (14%) developed intestinal gangrene, necessitating surgical intervention.

This analysis highlights the severe, potentially life-threatening course of LE and the importance of prompt recognition and aggressive immunosuppressive treatment to improve outcomes.

Systemic lupus erythematosus (SLE) is a multifaceted autoimmune disorder characterized by the presence of autoantibodies and autoreactive T lymphocytes, which drive widespread organ damage. Its clinical presentation is highly variable, reflecting its impact across multiple organ systems, including the skin, kidneys, lungs and the gastrointestinal (GI) tract. GI involvement is notably common in SLE patients, where the disease itself contributes to a range of manifestations such as recurrent oral ulcers, lupus hepatitis, autoimmune pancreatitis, protein-losing enteropathy and lupus enteritis (LE)[5,6]. Additionally, gastrointestinal symptoms and liver dysfunction in these patients may also stem from the adverse effects of medications, particularly non-steroidal anti -inflammatory drugs, which are commonly used to manage the inflammation and pain associated with SLE. Given the diverse GI complications seen in SLE, it remains critical for clinicians to distinguish between disease-related manifestations and medication side effects to ensure optimal management and prevent long-term organ damage.

Demographics: In our cohort, most patients (87.5%) were female, consistent with the known higher prevalence of SLE in women. The average age at presentation was 30 years, with a range from 17-55 years. These findings are consistent with those reported by other studies, including Janssens[1] who observed a similarly high prevalence of LE in women and noted that the disease often affects young adults, with their cohort having a median age of 30 years. Chen[5] also found a similar demographic trend, with a higher incidence of LE in females and a mean age of 30 years at diagnosis.

Clinical Features: Abdominal pain was the most common symptom in our patients, occurring in all cases (100%), which aligns with findings from Janssens[1] where abdominal pain was present in 97% of cases. Diarrhea and abdominal distension were seen in 50% of our patients, which is slightly higher than the 32% and 42% observed in the study by Janssens[1] respectively. The presence of fever and cutaneous lesions in 25% of our cohort is in line with Janssens[1] where fever occurred in 20% of cases. The significant variation in the prevalence of symptoms in these studies may be attributed to differences in study populations, diagnostic criteria and follow-up durations.

Laboratory Findings: Laboratory findings in our study revealed several abnormalities indicative of active lupus, most notably low C3 and C4 levels, which were seen in 75% and 25% of our patients, respectively. This is consistent with the findings of Janssens[1] where 88% of patients had low complement levels and Chen[5] where reduced complement C3 levels were independently associated with LE. Other significant findings in our cohort included positivity for the direct Coombs test (75%) and the presence of anemia, leukopenia and thrombocytopenia (each 50%). These laboratory features reflect active SLE disease, as also observed in Kwok[7] who found a similar frequency of hematologic abnormalities in their cohort of SLE patients with LE. Proteinuria, observed in 50% of cases in our study, is a well-recognized feature of systemic lupus erythematosus (SLE), often reflecting lupus nephritis. This finding is consistent with the literature. Janssens[1] reported proteinuria in 47% of lupus enteritis (LE) cases, aligning closely with our observations. Proteinuria in the context of LE may indicate concurrent renal involvement, further emphasizing the systemic nature of SLE and the overlap between renal and gastrointestinal manifestations. Chen[5] also highlighted the significance of proteinuria in SLE patients, reporting it as a common laboratory abnormality, often correlating with disease activity. These findings underline the importance of assessing renal function in SLE patients presenting with gastrointestinal symptoms, as it may provide additional insights into disease severity and guide the choice of immunosuppressive therapies.

Imaging Findings: Abdominal imaging, particularly CT scans, provided critical support in diagnosing LE. In our cohort, ascites was the most common finding (100%), followed by the target sign (75%) and bowel wall edema (62.5%). Janssens[1] similarly found bowel wall edema (95%) and ascites (78%) to be the most frequent CT findings. The target sign, indicative of bowel wall ischemia, was observed in 75% of our patients, which is in line with the 71% reported by Janssens[1] The presence of the Comb sign (50%) and surrounding fat stranding (37.5%) in our cohort was also consistent with findings in the literature, where mesenteric abnormalities and fat stranding are often observed in LE. These imaging findings are highly suggestive of active mesenteric involvement and ischemia, critical for distinguishing LE from other abdominal pathologies.

Treatment and Outcomes: All our patients received corticosteroids as first-line therapy, with additional immunosuppressive agents in the form of cyclophosphamide (62.5%) and rituximab (12.5%). Janssens[1] reported that corticosteroids were administered to all patients and immunosuppressive therapy, including cyclophosphamide, was commonly used, with a recurrence rate of 25%. Our treatment protocol was similar, but we observed a 25% mortality rate, with two patients dying due to severe disease progression. Chen[5] noted that high-dose corticosteroids were required in most LE patients, with a 25% recurrence rate, while Kwok[7] reported good responses to high-dose steroids in most patients, with no deaths directly attributed to LE. In our study, one patient developed intestinal gangrene and required surgical intervention, highlighting the potential for severe complications in LE, as seen in Chen[5] where some patients developed intestinal necrosis. This emphasizes the need for early intervention and aggressive immunosuppressive therapy to mitigate such life-threatening complications.

CONCLUSION

Lupus enteritis is a rare and poorly understood cause of abdominal pain in patients with SLE. This condition may progress to intestinal necrosis and perforation if untreated. Co-existence with active nephritis is also observed with LE. There is an excellent response to steroids and immunosuppression. Timely diagnosis is

crucial for the adequate management of this rare manifestation of SLE. A high index of clinical suspicion is mandatory for diagnosing LE.

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