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Unmasking Lupus Enteritis: A Rare Cause of Acute Abdominal Pain in Systemic Lupus Erythematosus (SLE)

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Background

Systemic Lupus Erythematosus (SLE) is a multisystem autoimmune disorder with diverse clinical manifestations, including constitutional, cutaneous, and articular involvement. Lupus enteritis, a rare but serious complication, occurs in 0.2% to 5.8% of SLE patients and is characterized by ischemic changes in the bowel due to mesenteric vasculitis. If not promptly diagnosed and treated, it can lead to bowel infarction, perforation, and increased mortality. Given its rarity and potentially severe outcomes, early recognition is crucial.

Case report

- We report a case of a 34-year-old female diagnosed with SLE in 2004, presenting with positive antinuclear antibodies (ANA), anti-dsDNA, low complement levels, inflammatory polyarthritis, autoimmune hemolytic anemia (AIHA) with relapses, and lymphopenia. She had been on a tapering dose of prednisolone and maintained on cyclosporin (75 mg twice daily) and hydroxychloroquine (200 mg daily). Notably, she had a history of transaminitis while on azathioprine and mycophenolate mofetil.
- In August 2023, she presented with acute abdominal pain, distension, nausea, and vomiting. Physical examination revealed dehydration, epigastric tenderness, and abdominal distension. Laboratory findings were unremarkable except for mild leukocytosis. A plain radiograph demonstrated dilated small bowel loops, and an abdominal computed tomography (CT) scan showed diffuse small bowel thickening with target signs, indicating extensive lupus enteritis involving the small bowel.
- The patient was diagnosed with lupus enteritis complicated by ischemic bowel and serositis. Electrolyte imbalances were corrected, and she received high-dose pulse steroid therapy (500 mg/day for three days), followed by 0.5 mg/kg/day prednisolone and Rituximab. Clinical improvement was observed by the second day, with resolution of abdominal symptoms.
- She was transitioned to maintenance therapy with hydroxychloroquine (200 mg daily) and a tapering regimen of prednisolone. Two weeks later, she received the second dose of Rituximab and remained asymptomatic. Gradual steroid tapering was implemented over the following weeks. One month later, she remained symptom-free at follow-up.

Investigations

Test	Values	Reference range
Haemoglobin	11.7	12.0- 15.0 g/L
WBC	9	4.0-10.0 x 10^9/L
Platelet	405	150-400 X 10^9/L
Urea	6.8	3.2-8.2 mmol/ L
Sodium	134	136-145 mmol/ L
Potassium	3.3	3.4-4.5 mmol/ L
Creatinine	67	49- 90 umol/L
Albumin	43	32- 48 g/L
ALP	170	46- 116 U/L
ALT	61	10-49 U/L
C3/ C4	0.5/ 0/04	
C- reactive protein	13.81	
Serum procalcitonin	0.37	

Table 1: Blood investigation results

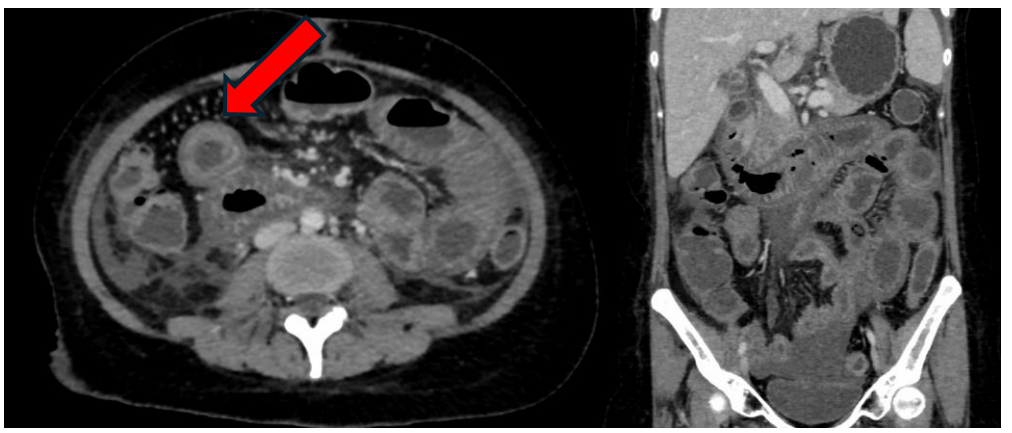

Figure 1: Abdominal CT scan – dilatation of intestinal segments, with circumferential, multisegmented mural thickening of over 3mm, affecting mainly the duodenum until the distal ileum, with associated submucosal oedema ("target sign" – arrow)

Discussion

- Lupus enteritis is a rare but recognized manifestation of SLE, occurring in patients with active disease [1]. It is characterized by inflammation of the small bowel, presenting with acute abdominal pain, nausea and vomiting. In our patient, acute onset of diffuse abdominal pain, nausea, and vomiting in the setting of serologic flare (hypocomplementemia) prompted consideration of lupus-related gastrointestinal involvement. These classic symptoms of gastrointestinal involvement in the context of active SLE, confirmed with imaging and clinical response to immunosuppressive therapy.
- Immune complex deposition in the small-vessel walls of the mesentery leads to complement activation, endothelial injury, and submucosal edema. This inflammatory cascade manifests radiologically as bowel-wall thickening , the "target sign", findings observed in our patient [3].
- Abdominal pain in SLE has a broad differential, including medication-induced pancreatitis, mesenteric ischemia, infectious enterocolitis, and serositis. Key distinguishing features of lupus enteritis include rapid radiologic changes, concomitant serologic activity (low C3/C4, rising anti-dsDNA), and a prompt response to immunosuppression.
- High-dose corticosteroids are the mainstay of therapy, with most cases showing rapid resolution of symptoms and radiologic improvement
- Timely recognition of lupus enteritis is crucial as delayed treatment can lead to serious complications such as bowel ischemia, perforation, or stricture formation. Our case highlights the importance of considering lupus enteritis in SLE patients presenting with abdominal pain, even in the absence of other systemic flare markers. Prompt diagnosis and early immunosuppressive treatment can prevent complications and improve outcomes.

Conclusion

This case underscores the importance of considering lupus enteritis in SLE patients presenting with acute abdominal symptoms, given its rarity and life-threatening complications. Prompt diagnosis and timely intervention with immunosuppressive therapy, including corticosteroids and Rituximab, can significantly improve outcomes. This report highlights the clinical characteristics, diagnostic approach, and successful management of lupus enteritis, emphasizing the role of early detection in enhancing patient survival and quality of life.

References

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