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CASE REPORT

Lupus mesenteric vasculitis with gastrointestinal bleeding as the primary manifestation: a case report

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Abstract

Systemic lupus erythematosus (SLE) is a complex autoimmune disease where the body loses tolerance to its own antigens, particularly nuclear antigens. Abnormal responses from T and B cells lead to the production of autoantibodies and the formation of immune complexes in tissues, triggering complement activation, inflammation, and irreversible organ damage. SLE can affect any part of the body, resulting in diverse clinical symptoms. One rare manifestation of SLE is lupus mesenteric vasculitis (LMV), which presents with vague symptoms, abnormal laboratory findings, and specific imaging features. LMV, although uncommon, can progress to severe complications such as bowel perforation, haemorrhage, and even mortality. Here, we report a case of LMV with the involvement of multiple organ systems (including mucocutaneous, musculoskeletal, serosal cavities, and haematological systems), presenting initially with life-threatening intractable gastrointestinal bleeding, and complicated by severe pulmonary infection. By sharing this case, we aim to enhance clinicians' confidence in managing critical SLE cases and raise awareness about disease surveillance.

Key words: Hematological involvement; Lupus mesenteric vasculitis; Lupus nephritis; Multiple organ damage; Systemic lupus erythematosus

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Introduction

Systemic lupus erythematosus (SLE), an autoimmune disease, affects joints and various organs, including the skin, kidneys, lungs, nervous system, and haematological system (Tselios and Urowitz, 2017). The availability of self-antigens, activation of the innate immune system, and dysfunction of the adaptive immune system are thought to promote the progression of SLE (Kaul et al, 2016). The complications of SLE are diverse and severe, and may include lupus pneumonitis, lupus encephalopathy, intestinal pseudo-obstruction, gastrointestinal bleeding, and vasculitis (Alves et al, 2016). It is important for clinicians to recognize SLE and its complications early in its course and treat the disease promptly and intensively. Here, we present a rare case of SLE with multisystem damage in a 48-year-old Chinese woman.

Case report

A 48-year-old female patient presented with facial redness persisting for 3 months and bloody stools for 1 week. She was admitted to our hospital on 6 November 2023. Upon physical examination, the patient exhibited signs of malnutrition and facial pigmentation on her face. Scattered petechiae were observed on her lower extremities, while petechiae measuring approximately 5 × 1 cm were noted on her abdomen, alongside depigmented areas measuring about 5 × 6 cm on her sacrum.

Laboratory analyses revealed elevated erythrocyte sedimentation rate and high-sensitivity C-reactive protein levels, along with decreased haemoglobin and platelet counts. Notably, antinuclear antibody was detected at a titre of 1:1000, while anti-double-stranded DNA and anti-SM antibodies were significantly elevated. Additionally, complement levels were decreased (Table 1). Based on the patient's clinical presentation and laboratory findings, she fulfilled the, 2019 ACR/EULAR classification criteria for SLE and received a diagnosis

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Table 1. Laboratory data

Parameters	Result	Reference interval
Complete blood counts		
White blood cells ($\times 10^9$/L)	7.61	3.5–9.5
Red blood cells ($\times 10^{12}$/L)	1.6	3.85–5.1
Haemoglobin (g/L)	49	115–150
Platelets ($\times 10^9$/L)	73	100–300
Urinalysis		
24-hour urinary protein quantification (mg/24h)	692	20–141
Occult blood	–	–
Biochemistry		
Erythrocyte sedimentation rate (mm/h)	131.5	0–26
Procalcitonin (ng/mL)	7.11	0–0.1
High-sensitivity C-reactive protein (mg/L)	13.90	0–5
Lactate dehydrogenase (U/L)	331	120–250
Aspartate transaminase (U/L)	106	13–35
Uric acid (umol/L)	502.9	150–370
Albumin (g/L)	13.5	40–55
Creatinine (umol/L)	181	41–73
Thrombin time (s)	26.10	14–21
D-dimer (Ug/mL)	14.31	0–1
Plasma fibrinogen concentration (g/L)	1.16	2–4
Ferritin (ng/mL)	1330.7	10–291
Immunology		
Antinuclear antibody	1:1000+	–
Anti-RNP antibody (RU/mL)	204.6	0–19.99
Anti Ro52 antibody (RU/mL)	91.95	0–19.99
Anti-nucleosome antibody (RU/mL)	>400.00	0–19.99
Anti-dsDNA antibody (RU/mL)	>800	0–9.99
Anti-histone antibody (RU/mL)	>400	0–19.99
Anti-SSA antibody (RU/mL)	>400.00	0–19.99
Anti-SM antibody (RU/mL)	204.66	0–19.99
Immunoglobulin G (g/L)	25.80	7.6–15.2
Immunoglobulin A (mg/L)	11200	800–4530
Complement 3 (mg/L)	374	790–1520
Complement 4 (mg/L)	61.2	160–380
Direct Coombs test	+	–

(Aringer et al, 2019). The SLE Disease Activity Index (SLEDAI-2000) score was 26, indicating severe disease activity.

The patient experienced repeated passage of bright red stool, accompanied by a large amount of blood clots, occurring 5–6 times daily for over 10 days. Notably, the patient did not report abdominal pain, nausea, or vomiting of blood. Abdominal computed tomography (CT) imaging (Figure 1A) revealed thickening and swelling of the wall of the gastric antrum wall, along with prominent oedema of the peritoneum and mesentery, accompanied by indistinct fat spaces. Due to the suspicion of lupus complicated with mesenteric vasculitis, the patient underwent methylprednisolone 240 mg pulse therapy for 3 days (adjusted for the patient's weight of 30 kg), followed by maintenance treatment with methylprednisolone 80 mg per day, simultaneously immunoglobulin 15 g treatment for 3 days. The patient experienced recurrent fever during hospitalisation, and chest CT indicated pulmonary infection (Figure 2A), with sputum smear revealing the presence of abundant yeast-like fungi and a significantly elevated fungal d-dextran level of 742.400 pg/mL. Treatment comprised a regimen of meropenem, fluconazole, and compound sulfamethoxazole tablets. Given the possibility of Pneumocystis carinii pneumonia, anti-infective therapy, blood transfusion, and symptomatic support measures were administered. Gradually, the patient's gastrointestinal bleeding ceased, and lung infection was brought under control. Subsequently, Telitacicept was initiated to manage lupus activity, tailored to the patient's condition.

Following treatment, the patient exhibited absorption of double pneumonia lesions, as well as resolution of left pleural effusion and pericardial effusion. Furthermore, there was a notable reduction in peritoneal and mesenteric thickening and oedema, along with subsidence of intestinal canal swelling and slight blurring of the fat space (Figure 1B and Figure 2B). The patient's platelet count increased, anaemia ameliorated, albumin and complement levels rose, while anti-dsDNA antibody, inflammatory markers, and D-dimer

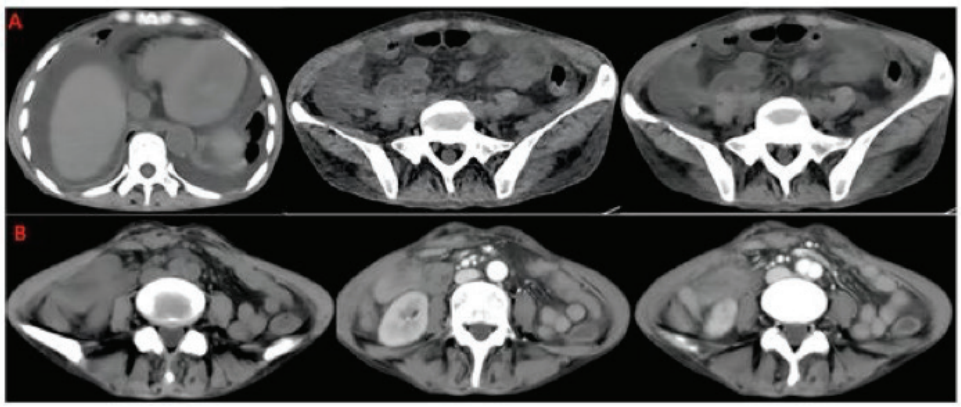

Figure 1. Comparison of abdominal CT before and after treatment (A: 8 November 2023, B: 20 November 2023).

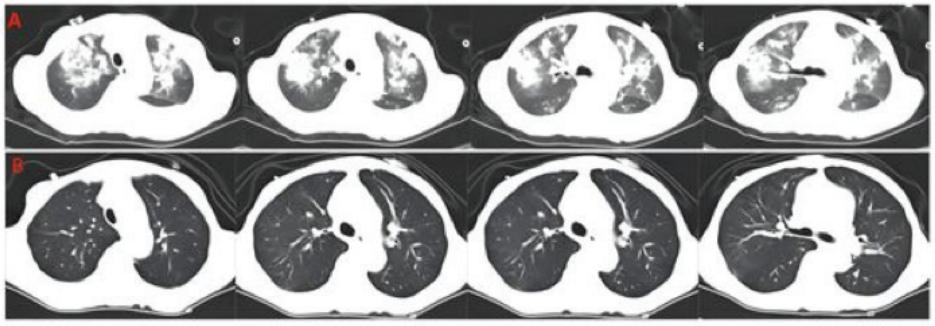

Figure 2. Comparison of chest CT before and after treatment (A: 11 November 2023, B: 18 November 2023).

levels decreased (Figure 3). Consequently, the patient's overall condition significantly improved, leading to discharge.

Discussion

Systemic lupus erythematosus is a chronic autoimmune condition that can impact nearly all organ systems, leading to diverse clinical presentations and numerous complications (Tselios and Urowitz, 2017). Multisystem involvement is considered a significant prognostic indicator (van Vollenhoven et al, 2014). The patient we are reporting exhibits nearly all the organ damage associated with SLE, including facial redness, joint pain, pericarditis, and pleurisy, as well as the involvement of the haematological system, kidneys, and gastrointestinal tract.

Lupus mesenteric vasculitis (LMV) represents a rare yet severe complication observed in SLE patients, with a prevalence ranging from 0.2% to 9.7% among this population (Yuan et al, 2014). The clinical manifestations of LMV are non-specific, with abdominal pain being the most commonly reported symptom (Alsalameh and Hauggaard, 2013). Roughly 90% of LMV patients experience abdominal pain of varying intensity, often accompanied

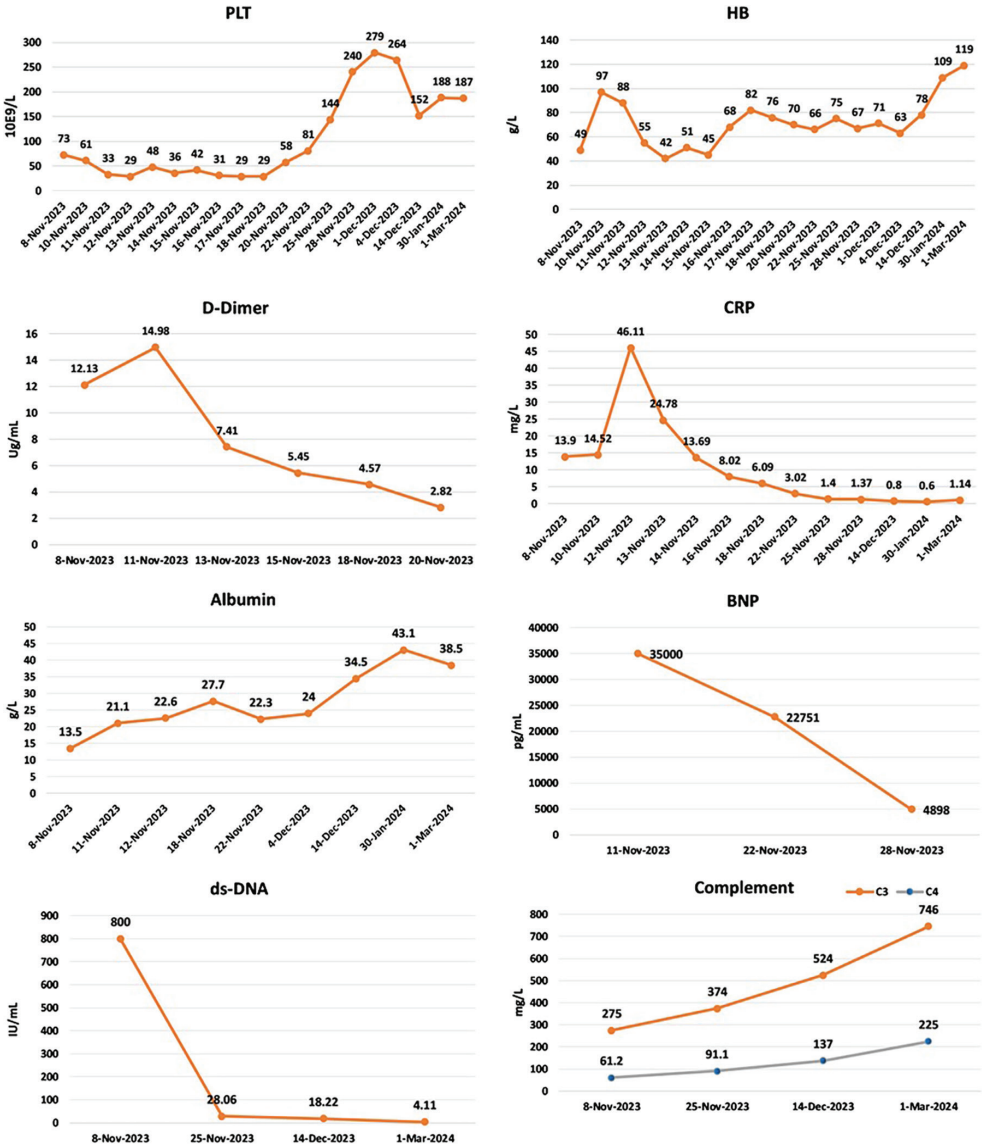

Figure 3. Changes to relevant indicators during hospitalization. PLT, platelet; HB, haemoglobin; CRP, C-reactive protein; ds-DNA, double-stranded DNA; BNP, pro-brain natriuretic peptide.

by symptoms such as nausea, abdominal distension, diarrhoea, and vomiting (Chen et al, 2021). Clinicians should consider LMV when observing abdominal symptoms occur in SLE patients, underscoring the importance of prompt abdominal CT imaging, particularly with contrast enhancement, which enhances sensitivity in detecting intestinal abnormalities. Three typical findings on abdominal CT findings in LMV patients include bowel wall thickening (>3.0 mm), resulting in the "target sign" due to separation of the mucosa and muscle layers; mesenteric vasodilation creating the "comb sign"; and increased attenuation of mesenteric fat (Chu et al, 2014). The concurrent presence of the "target sign" and "comb sign" is highly specific to LMV, and aids in diagnosis.

The patient we report initially presented with facial redness, followed by bloody stools that progressed to massive bleeding. Subsequent abdominal CT revealed thickening and swelling of the gastric antrum wall, significant thickening and oedema of the peritoneum and mesentery were significantly thickened and oedematous (>3 mm), and the fat space was blurred fat spaces. A diagnosis of LMV was established, with mesenteric vasculitis considered the cause of gastrointestinal bleeding. While the predominant clinical manifestations of LMV include abdominal pain (70%–100%), vomiting (60%–80%), and diarrhoea (30%–60%), gastrointestinal bleeding may occur in approximately 3% of cases, potentially leading to intestinal perforation (Sran et al, 2014).

At present, standardised guidelines for treating LMV are unavailable, with glucocorticoids typically regarded as the primary treatment (Nozari and Divsalar, 2014). These medications can effectively prevent severe complications such as intestinal ischaemia, necrosis, and perforation (Ju et al, 2009). Most LMV patients experience symptom relief following high-dose corticosteroid therapy, with or without additional immunosuppression (Chen et al, 2021). In the case of the patient discussed herein, gastrointestinal bleeding of this patient gradually ceased and symptoms improved following high-dose corticosteroid therapy.

Currently, no universally accepted diagnostic and treatment protocol exists for LMV, and early and accurate diagnosis remains challenging, primarily relying on exclusionary criteria. Thus, differential diagnosis plays a crucial role. Furthermore, no single biological test serves as a specific diagnostic marker. While pathological diagnosis is considered the "gold standard," its invasive nature limits its routine use. Therefore, CT serves as a non-invasive diagnostic tool commonly employed in LMV diagnosis of LMV.

In this case, the patient presented with underlying lupus and multisystem involvement. Although she experienced gastrointestinal bleeding, she did not have typical symptoms of LMV such as abdominal pain and vomiting were absent, leading to an initial oversight of LMV. However, after multidisciplinary discussions and consideration of this possibility, timely administration of this disease, high-dose corticosteroid therapy resulted in gradual cessation of gastrointestinal bleeding.

This case serves as a reminder to clinicians that when encountering gastrointestinal bleeding in patients with a history of lupus, even in the absence of typical LMV symptoms of LMV such as abdominal pain and vomiting, it is crucial to consider the possibility of this condition and rule out other differential diagnoses. Multidisciplinary collaboration is essential in accurately assessing the patient's condition, identifying key issues, and ultimately improving the survival rates of critically ill patients.

Learning points

- The patient exhibits facial redness, joint pain, pericarditis, pleurisy, and haematological involvement.
- A severe lung infection and LMV with gastrointestinal bleeding are complicating factors.
- The patient has LMV is diagnosed, with gastrointestinal bleeding as the primary manifestation.
- High-dose glucocorticoid pulse therapy effectively halts gastrointestinal bleeding due to LMV.

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Availability of data and materials

All data included in this study are available upon request by contact with the corresponding author.

Author contributions

JRC and YHP designed the research. JRC drafted the manuscript. YHP revised the manuscript and provided funding support. QW and QRW analyzed the data. All authors contributed to important editorial changes in the manuscript. All authors read and approved the final manuscript. All authors have participated sufficiently in the work and agreed to be accountable for all aspects of the work.

Ethics approval and consent to participate

This study was approved by the Institutional Medical Ethics Committee of the Affiliated Hospital of North Sichuan Medical College (2023ER180-1). The patient provided an informed consent form.

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Conflict of interest

The authors declare no conflict of interest.

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