CASE REPORT

Rectal ulcers induced by systemic lupus erythematosus

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SUMMARY

A 28-year-old woman presented with diarrhoea, haematochezia, tenesmus and rectal pain for 2 months. She was diagnosed with systemic lupus erythematosus (SLE) 8 years ago and remained on prednisone, azathioprine and hydroxychloroquine. Blood work revealed a positive ANA (antinuclear antibody test), anti-dsDNA 749 IU/mL (0–300 IU/mL), C3 0.22 g/L (0.65–1.65 g/L) and C4 0.05 g/L (0.16–0.60 g/L). Stool studies were unremarkable. MRI of the pelvis showed a rectum with eccentric wall thickening. Flexible sigmoidoscopy showed severe proctitis with multiple deep ulcers and diffuse submucosal haemorrhage. Rectal biopsy revealed crypt architectural distortion and reactive fibrosis in the lamina propria. The patient was given mesalamine suppository for 2 weeks with minimal improvement. Repeat flexible sigmoidoscopy showed a coalesced 3×4 cm full-thickness rectal ulcer. Therefore, the patient was given intravenous methylprednisolone for 3 days, followed by intravenous cyclophosphamide for 2 weeks. Her symptoms resolved and repeat flexible sigmoidoscopy showed fibrotic healing of the rectal ulcers.

BACKGROUND

Systemic lupus erythematosus (SLE) may affect any part of the gastrointestinal tract,¹ but colonic involvement is rare and occurs in only 0.2%.² The left colon is more commonly affected than the right due to the presence of 'watershed' areas which arise from anastomoses of the superior mesenteric artery, inferior mesenteric artery and middle rectal artery, while the rectum is relatively protected owing to its rich and dual vascular supply from branches of the inferior mesenteric artery and internal iliac artery.³ Rectal ulcers are therefore exceedingly rare and solitary lesions are theoretically more likely to occur in SLE.

A 28-year-old Chinese woman presented with a 2-month history of worsening diarrhoea, haematochezia, tenesmus, rectal pain, anorexia and weight loss. She denied any recent travel or sick contacts and had not been sexually active. She was diagnosed with SLE 8 years ago and since then she had remained on prednisone (4 mg daily) and hydroxychloroquine (200/400 mg on alternating days). About 6 months ago, azathioprine (75 mg daily) was added to better control her SLE. Physical examination showed mild left lower quadrant tenderness, livedo reticularis on upper back and extremities, and no active joints. Blood work revealed a haemoglobin of 69 g/L (115–160 g/L), albumin of 22 g/L (40 g/L), normal liver enzymes,

positive ANA, anti-dsDNA of 749 IU/mL (0–300 IU/mL), C3 of 0.22 g/L (0.65–1.65 g/L), C4 of 0.05 g/L (0.16–0.60 g/L), negative antiphospholipid antibodies, negative cytomegalovirus (CMV) serology, and negative *Entamoeba histolytica* serology. Stool C&S, O&P, *Clostridium difficile* toxin, Shiga toxin, acid-fast bacilli (AFB) smear and *E. histolytica* ELISA were all negative. MRI of the pelvis showed a diffusely abnormal rectum with eccentric wall thickening and areas of ulceration but no defined fistulous tract or abscess. Given the history and abnormal MRI, a flexible sigmoidoscopy was performed and revealed moderate-to-severe proctitis with multiple deep ulcers seen immediately on entry to the anal canal. The rectal mucosa was erythematous and friable with diffuse submucosal haemorrhage up to 10 cm from the anal verge (figure 1). Normal mucosa was observed beyond 10 cm. Rectal biopsies showed ulceration, crypt architectural distortion and reactive fibrosis in the lamina propria, but no evidence of granuloma, viral inclusion, dysplasia or neoplasia (figure 2). In addition, AFB smear, immunoperoxidase stain, viral PCR and Periodic acid–Schiff–diastase (PAS-D) stain of rectal biopsies were negative for *Mycobacterium*, CMV, herpes simplex virus and parasites.

Infectious colitis and inflammatory bowel disease were felt to be unlikely, and the rectal ulcers were believed to be a result of ischaemia secondary to mesenteric vasculitis from SLE. The patient was discharged on mesalamine suppository 1 g twice daily. However, she noticed minimal improvement and returned to the hospital 2 weeks later with an

Figure 1 Endoscopic examination of the rectum revealing multiple deep ulcers with mucosal friability and submucosal haemorrhage.

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Figure 2 Rectal biopsy demonstrating ulceration, crypt architectural distortion and reactive fibrosis in the lamina propria.

increase in rectal bleeding and bilateral pedal oedema. Repeat flexible sigmoidoscopy showed coalescing erythematous mucosal patches and a 3×4 cm full-thickness rectal ulcer. Therefore, high-dose intravenous methylprednisolone 250 mg daily was started. However, no clinical improvement was seen after 3 days and subsequently intravenous cyclophosphamide 500 mg daily was initiated. Her symptoms significantly improved and repeat flexible sigmoidoscopy performed 2 weeks later showed a significant decrease in mucosal erythema and friability as well as fibrotic healing of the previously identified rectal ulcers (figure 3). The patient was discharged on oral prednisone 25 mg twice daily and had been doing well since.

DISCUSSION

In limited published case reports, 6 of 10 patients presented with multiple rectal ulcers.^{1–10} Five cases were complicated by rectal perforation or fistulisation,^{3–7} which is associated with

significant morbidity and mortality. Our case also presented with multiple rectal ulcers but fortunately without perforation or fistulisation.

The single most unique aspect of our case is that our patient did not respond to high-dose corticosteroids. Five cases responded to either oral prednisolone or intravenous methylprednisolone.^{1 2 5 6 10} Four cases required surgery for necrosis or perforation.^{3 4 7 9} One case received both tacrolimus and high-dose corticosteroids and hence clinical improvement could be ascribed to either or both of these agents.⁸ Our patient's rectal ulcers improved only when cyclophosphamide was initiated, which is the most common second-line agent for the treatment of mesenteric vasculitis induced by SLE.¹¹

Mechanisms by which rectal ulcers develop in gastrointestinal lupus have been postulated, including mesenteric vasculitis from SLE, mesenteric thrombosis from antiphospholipid syndrome,⁸ endothelial enlargement from CMV infection,¹² and vascular sclerosis from steroid use,⁴ all of which may result in mucosal ischaemia. Histology of rectal ulcers induced by SLE typically reveals non-specific inflammation without any pathognomonic findings, and vasculitis with fibrinoid necrosis is often difficult to demonstrate.¹³

Our case illustrates that rectal ulcers are a rare but possible manifestation of an SLE flare. As rectal involvement may lead to potentially life-threatening complications, it is crucial to make a timely diagnosis and start treatment promptly.

Learning points

- ▶ Systemic lupus erythematosus may affect any part of the gastrointestinal tract but rectal involvement is rare.
- ▶ Rectal ulcers induced by systemic lupus erythematosus may lead to life-threatening complications such as perforation and fistulisation.
- ▶ Rectal ulcers induced by systemic lupus erythematosus may be refractory to corticosteroids and only respond to cyclophosphamide.

Figure 3 Repeat endoscopic examination of the rectum after cyclophosphamide treatment showing fibrotic healing of the rectal ulcers with significant decrease in mucosal friability and submucosal haemorrhage.

Competing interests None.

Patient consent Not obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

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