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# Case-Based Review: Gastrointestinal Stricture Decades After Diagnosis of Lupus

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## *Abstract*

*Gastrointestinal manifestations are well described in the setting of lupus either as presenting features or more commonly as adverse effects of immunosuppressive drugs. However, gastrointestinal stricture occurring as a complication of lupus years after initial diagnosis is rare. We searched for keywords lupus, gastrointestinal, rectal, and stricture in PubMed, Medline, and Google Scholar databases. The search included articles published from January 1950 to October 2020 with no language restrictions. The search yielded 64 citations in English; 36 of the articles were excluded as they were not relevant; 5 articles were excluded since they were duplicates, leaving 23 articles. Full-text articles of the 23 citations were obtained and reviewed. Only two prior cases reported in literature and both these cases were complicated by perforation. Our patient, a 43-year-old lady, presented with abdominal pain, vomiting and loose stools of 3 months duration. CT demonstrated long segment eccentric rectosigmoid thickening with no obvious proximal dilatation. Colonoscopy and histopathology confirmed underlying stenosis and chronic inflammatory infiltrates. Tuberculosis, inflammatory bowel disease and malignancy were ruled out. Medical history was notable for diagnosis of lupus at 23 years of age and irregular treatment with steroids and lupus was detected to be active at current admission. Subsequent to treatment with steroids and cyclophosphamide, a good response was noted clinically and confirmed by endoscopy. This case highlights the need to judiciously investigate and consider lupus disease activity as etiology for gastrointestinal stricture even years after the diagnosis of lupus.*


***Keywords:*** *lupus, gastrointestinal, rectal, stricture*

## Highlighting points

1. The present case study underlined the importance of considering lupus as the aetiology for stricture in a patient presenting to the gastroenterologist years after diagnosis of lupus.

2. Extent of involvement, absence of obstructive symptoms, excellent response to steroids and cyclophosphamide and recovery without complications are reported as unique features of the current case.

3. In the absence of histopathological findings, the review of the patient's clinical and investigations and reasonably ruling out other differentials aided decision making.

## Introduction

Systemic lupus erythematosus (SLE) is a classic prototype of chronic autoimmune disease characterised by multisystem involvement. Gastrointestinal features are common in lupus; these manifestations are most often related to adverse effects of therapy or concurrent infections. Of the specific manifestations, oral ulcers are a hallmark of active lupus **[**1**]**. Though less common, GI vasculitis, protein losing enteropathy, hepatobiliary involvement and pancreatitis have been described in literature **[**1-3**]**. Inflammatory bowel disease and celiac disease have been infrequently associated with lupus **[**3**]**. Clinical examination with particular attention to other clinical features of disease activity and judicious investigations are necessary as symptoms like abdominal pain, vomiting and altered bowel habits are non-specific. Rectosigmoid stricture is an unusual manifestation of lupus. We present a case of rectosigmoid stricture presenting with chronic diarrhea, abdominal pain and weight loss in a patient with long-standing lupus.

## Clinical Case Presentation

A 43-year-old woman presented to the gastroenterology OPD of our hospital with complaints of abdominal pain, loose stools, sporadic episodes of bleeding per rectum, intermittent vomiting of 3 months duration. In the first two months of her illness, she received three courses of different antibiotics suspecting an infective aetiology. Two months after onset of her illness, due to persisting symptoms she was admitted at a regional hospital.

**Initial medical management and investigations**

A working diagnosis of infective colitis/antibiotic associated diarrhoea was considered and she received intravenous fluids and oral metronidazole. Initial work up demonstrated BUN 14.9 mmol/l, creatinine 1.23 mg/dl, C-reactive protein 201 mg/l, white cell count 24.7 × 103/dl, haemoglobin 9.2 mg/dl and negative cultures. CT abdomen and pelvis followed by sigmoidoscopy demonstrated a mucosal ulceration with luminal narrowing in the rectosigmoid region. Biopsy was inconclusive except for chronic inflammatory infiltrate.

**Subsequent workup and medical management**

One month later, she presented to the GI clinic of our centre due to persistent symptoms. She had lost 8 kg of weight since the onset of her illness; she had no history of fever, melena or hematemesis. She was re-evaluated and CT Abdomen with rectal contrast revealed long segment asymmetrical mural thickening involving sigmoid colon and rectosigmoid along with thickening of the mesorectal fascia with loss of fat planes with uterus, bilateral adnexa, cervix and the mid and distal ileal loops (Fig.1&2) Flexible sigmoidoscope could be passed only up to 10 cms due to luminal narrowing, mucosal oedema, and ulceration (Fig. 3). Stricture was traversed partially using a gastroscope and a biopsy specimen was taken from the rectosigmoid. Even the gastroscope could not be negotiated further due to tightness of the stricture. Differential diagnosis of inflammatory bowel disease, tuberculosis and malignancy were considered. Histopathology demonstrated areas of chronic colitis with presence of lymphocytes, foamy macrophages and few giant cells. Usual mucosal crypt architecture was distorted. The arteries had marked hyperplasia resulting in severe hemorrhage and congestion (Fig. 4 & 5). The pathologic findings did not confirm Crohn's disease but, rather, were suggestive of vasculitis, with inflammatory cell infiltrates in the vessel walls.

Review of history revealed multiple comorbidities. She has hypothyroidism since the last 20 years, hypertension for 5 years and diabetes mellitus for one year. SLE was diagnosed in 2001 following features of skin rash, oral ulcers, Raynaud's phenomenon and joint pain. Renal biopsy in 2015 showed features of class II nephritis. She also has paresthesia and decreased sensation of bilateral lower limbs and left upper limbs since 2015. She was advised steroids, azathioprine and hydroxychloroquine (HCQ). She discontinued azathioprine after a few months but took steroids and HCQ intermittently.

She was evaluated by a rheumatologist. She had alopecia, Raynaud's but no oral ulceration. Livedoid and healed vasculitic rash was present over soles. She had proximal weakness (4-/5) of the lower limbs bilateral along with distal sensory neuropathy of both upper limbs and lower limbs. Nerve conduction

velocity showed axonal and demyelinating asymmetric motor sensory neuropathy. Investigation revealed raised erythrocyte sedimentation rate (ESR) of 52 mm/hour and sensitivity C-reactive protein, positive dsDNA, positive antinuclear antibodies (ANA), and low C3 complement levels. She had diminished vision for 6 months. Fundus examination revealed retinal vasculitis along with exudates and bilateral post subcapsular cataract.

She was started on injectable methylprednisolone 500 mg iv followed by 0.5 mg/kg steroids in a tapering schedule along with monthly intravenous cyclophosphamide. Symptomatic relief was seen as early as one month after therapy in the form of reduced episodes of abdominal pain, normal stool formation and no fresh episodes of bleeding per rectum. Repeat sigmoidoscopy after 2 doses of cyclophosphamide revealed near complete resolution of rectosigmoid stricture with significant mucosal healing (Fig. 6). After completion of 6 monthly cyclophosphamide infusions, maintenance therapy with mycophenolate mofetil is planned.

| S. no | Author/ year | Age/Sex | GI manifestations | Duration of Gi manifestation | Rheumatology diagnosis/ duration | Past and ongoing rheumatological Rx | Antibody, complements | Scopy findings | CT/Barium | Operative fIndings | Histopath | Treatment |
|---|---|---|---|---|---|---|---|---|---|---|---|---|
| P1 | Gore RM, 1983 [5] | 37/F | nausea, vomiting, mid abdominal pain, diarrhea | 3 days | SLE-12 yr | NA | ANA+ Low complements | Mucosal ulceration,, friable mucosa | 12-cm segment of mucosal ulceration and irregularity of the midtransverse colon | ND | ND | Prednisolone 60 mg and tapered |
| | | | 6 weeks later following steroid taper crampy abdominal pain, nausea, vomiting, and guaiac-positive loose stools | | | | | | 3-cm narrowed segment of midtransverse colon, measuring 1 cm in greatest diameter, dilation and "thumbprinting" of proximal colon, thickening of valvulae conniventes of the distal ileum | 1.2-cm perforation of the caecum, focal necrosis and ulceration in transverse colon | Fibrinous peritonitis, mesenteric arteritis with organizing thrombus, acute and chronic vasculitis of the subserosal vessels in the right and transverse colon | Distal ileum, cecum, ascending and transverse colon resection and ileostomy |
| P2 | Keating, 1998, [6] | 51/F | Inc frequency of bowel movements | 3 weeks | SLE/27 years | Cyclophosphamide (past) Prednsisolone 6 mg current | NA | Tight stricture at distal 30 cm of colon | 1.5-cm, high grade stricture in the mid sigmoid colon no associated mucosal irregularity no diverticular disease | ND | ND | Steroids (dose NA) |
| | | | 18 mo later malaise and sacral pain | 3 weeks | | Steroids (dose NA) | NA | ND | supralevator horseshoe abscess, | edematous mucosa and serosal fibrosis of colon | fibrosis and chronic inflammation of the bowel wall Fat necrosis of serosa, endarteritis obliterans of medium-sized arteries | CT guided percutaneous drainage of abscess and Hartmann's procedure |

**Table 1:** Clinical presentation and histopathology findings of the previous two cases

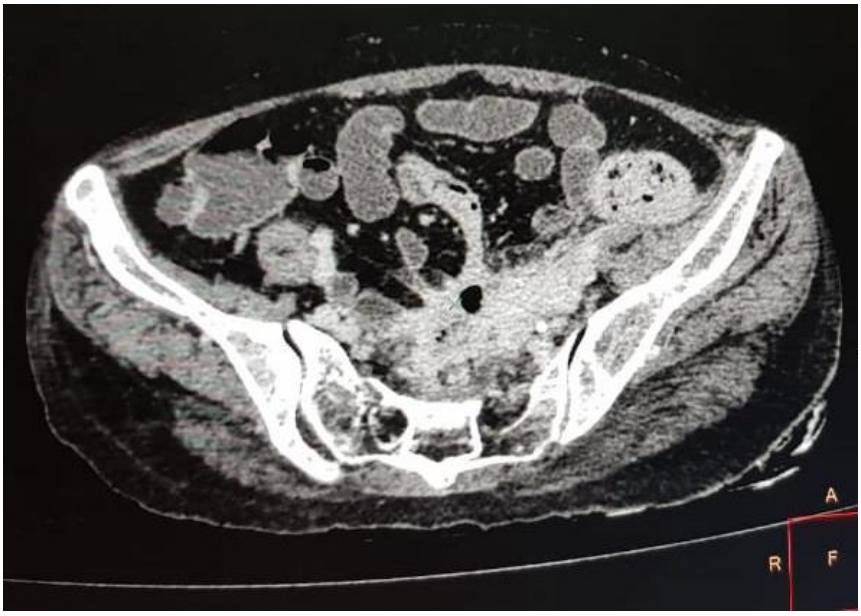


**Figure 1:** CT

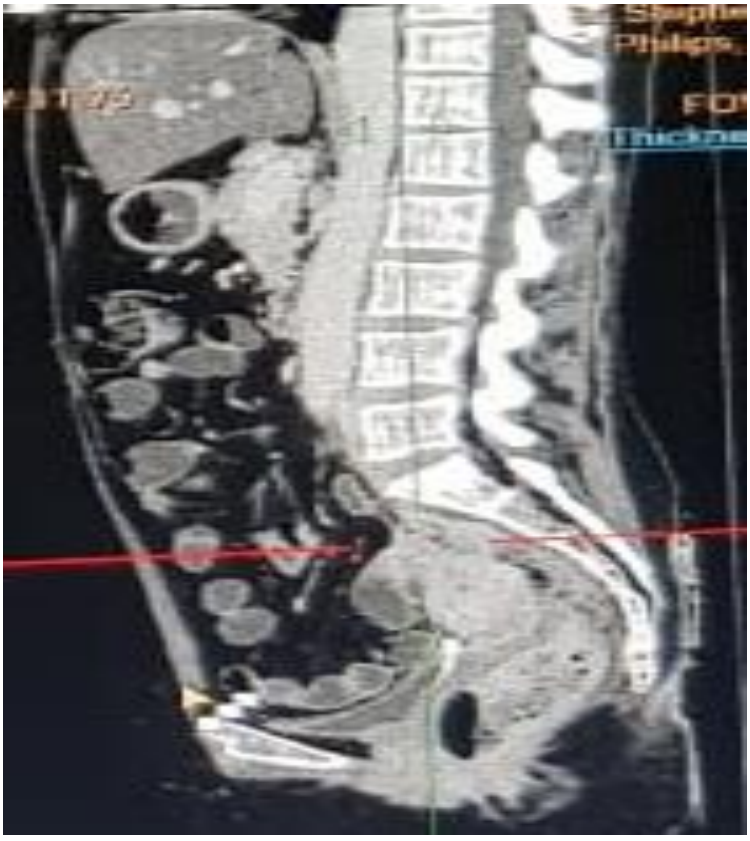

**Figure 2:** CT

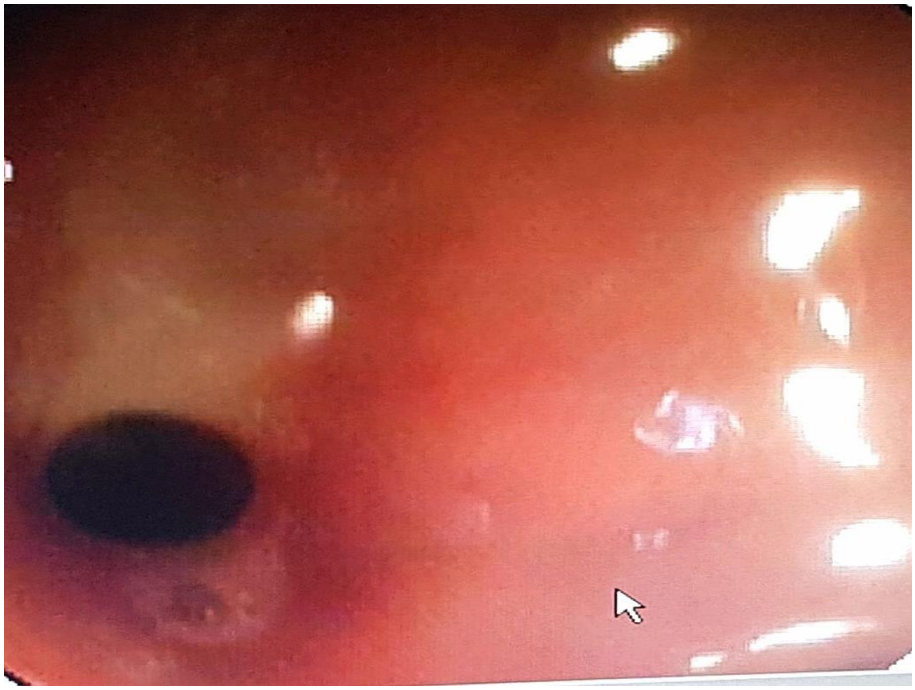

**Figure 3:** Sigmoidoscopy image

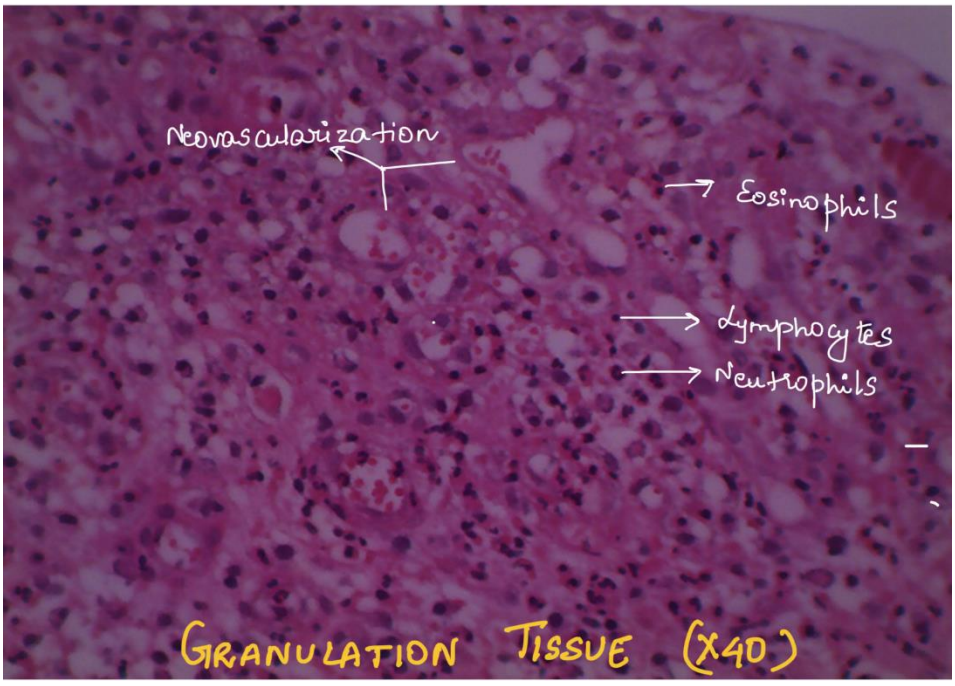


**Figure 4:** Histopathology slide

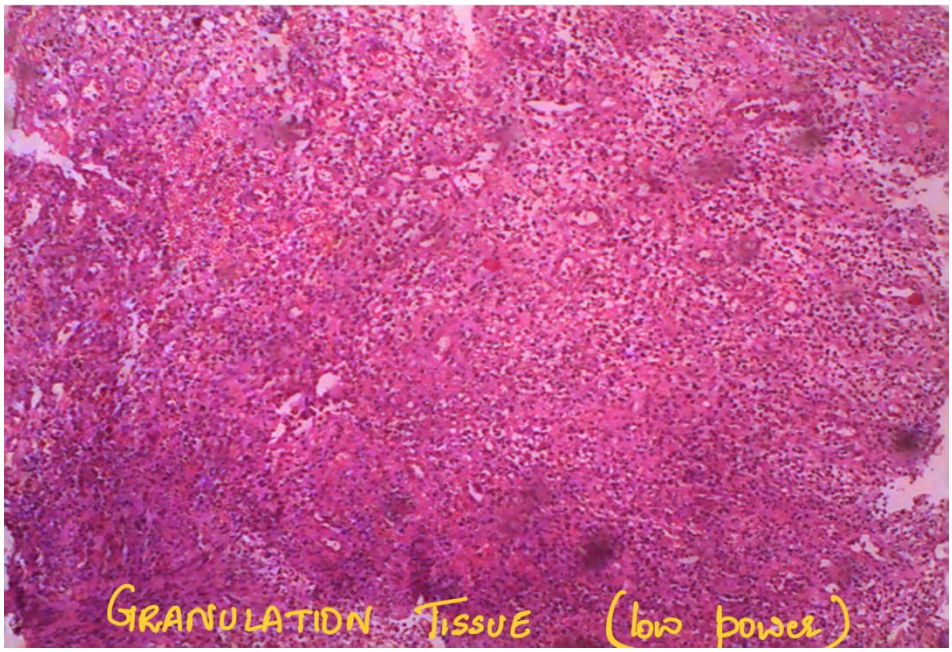


**Figure 5:** Histopathology slide

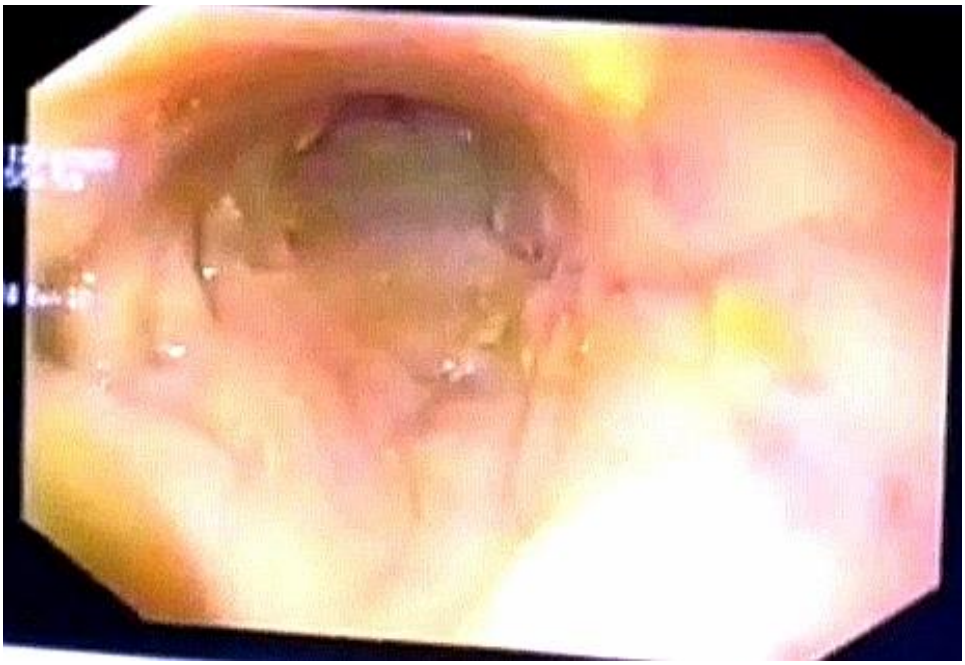

**Figure 6:** Repeat sigmoidoscopy image

## Discussion

Intestinal stricture in lupus is described rarely as a consequence of vasculitis complicating lupus. There are only two case reports of colon stricture in lupus patients in the 1980s. There is no mention of stricture in subsequent large case series and reviews of gastrointestinal manifestations of lupus **[**1-4**]**. This case is the third case described in literature so far.

The clinical presentations, endoscopy, imaging and histopathology findings of the previous two cases are detailed in Table 1. As in our patient, the previous two patients were also females with a history of long standing SLE. Similar to our patient, Raynaud's phenomenon has been described in the case report by Gore et al. **[**5**]**. In our patient lupus was not adequately controlled and she was not compliant with treatment. She had other organ involvement consistent with lupus activity such as axonal and demyelinating neuropathy, cutaneous lesions and retinal vasculitis. She had low c3 and increased anti-dsDNA. Patient P1 had cutaneous ulcers and low c3 which is evidence for active lupus **[**5**]**. P2 had clinically quiescent disease in other organs however serological reports are not available **[**6**]**. In this context, it is relevant to mention that Raynaud's phenomenon and low complements have been described as an independent predictor of gastrointestinal involvement **[**4**]**. It is likely that uncontrolled disease leading on to vasculitis would have caused stricture.

Both the patients described in literature showed initial improvement on steroids but subsequently developed perforation which necessitated surgery. The surgical specimen showed clear features of vasculitis. In our patient, though histopathology showed features of chronic inflammation, unequivocal vasculitis was not demonstrated. The rarity of presentation and elevated CRP level lead to consideration of other differentials. Histopathology was not suggestive of inflammatory bowel disease, malignancy or infective cause such as TB and CMV. Considering the clinical and imaging findings in entirety, vasculitis is the most likely aetiology and it is possible that the biopsy was not representative as it was not a full thickness post-surgical specimen. The patient was commenced on IV methylprednisolone for rapid action. IV cyclophosphamide was given as the efficacy of oral agents was doubtful in the setting of gut involvement and poor compliance. Further, addition of cyclophosphamide also facilitated faster steroid tapering. Keating et al. suggested that though immunosuppressive treatment is beneficial, established stricture in lupus requires surgery which doesn't seem to be the case in our patient **[**6**]**.

This case highlights the importance of considering lupus as the aetiology for stricture in a patient presenting to the gastroenterologist years after diagnosis of lupus. In the absence of clear-cut histopathological features, the review of the patients clinical and investigations and reasonably ruling

out other differentials aided decision making. Unique to the current case was the extent of involvement, absence of obstructive symptoms, excellent response to steroids and cyclophosphamide and recovery without complications like perforation or peritonitis.

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