A case of systemic lupus erythematosus presenting with rectal ulcers as the initial clinical manifestation of disease

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Abstract

Gastrointestinal involvement is often seen in patients with systemic lupus erythematosus (SLE). All parts of the gastrointestinal tract may be affected. However, rectal involvement at onset is rare. We describe here a case of SLE in which rectal ulcers due to vasculitis occurred as the initial manifestation of the disease without involvement of any other organ. The ulcers worsened, along with the appearance of lupus nephritis 5 years later. When steroid therapy was initiated, there was rapid clinical and radiographic improvement. Our case suggests that rectal ulcer is a rare but important complication of SLE and can represent the initial and sole clinical manifestation of the disease.

Introduction

Patients with systemic lupus erythematosus (SLE) may present with various gastrointestinal symptoms such as nausea, vomiting, dysphagia, abdominal pain, diarrhea and hemorrhage (1, 2). Some of these symptoms are known to be caused by vasculitis (1,2). Although gastrointestinal manifestations due to vasculitis are relatively uncommon, they represent an important cause of morbidity and mortality in SLE (1, 2). Here we describe a patient with SLE who first presented with rectal ulcers due to vasculitis as the initial clinical manifestation of disease.

Case report

In June 1995, a 27-year-old man first presented with fever and abdominal pain with constipation, and was treated with nonsteroidal anti-inflammatory drugs. In July 1996 he was admitted to the Japanese Foundation for Cancer Research Hospital with a two-month history of constipation, fever and bloody stool. Laboratory investigations revealed a erythrocyte sedimentation rate of 5 mm/h (Westergren); white blood cell count, 3,300/mm³ with mild lymphopenia (702/mm³); C-reactive protein, 0.27 mg/dl (normal < 0.4); and immunoglobulin (Ig) G, 2,690 mg/dl. Antinuclear antibody and anti-SS-A/Ro antibodies were positive. Anti-DNA antibody was present at 25 IU/ml (normal < 10). Colonoscopy demonstrated multiple ulcers on the rectal mucosa. A gastrografin enema revealed a fistula in the posterior wall of the rectum, and leakage of gastrografin along the sacral bone was detected. A diagnosis of perirectal abscess caused by rectal ulcers was made. Antibiotics were administered and a colostomy was performed to provide the patient with an artificial anus at the transverse colon. The patient gradually improved postoperatively. Although he remained well, angiography of the inferior mesenteric artery was performed in June 1997 to evaluate this rectal involvement of unknown origin and to attempt to close the artificial anus surgically. The superior rectal artery branch from the inferior mesenteric artery displayed irregular stenosis and an interrupted image.

In August 1999 the patient was referred to Keio University Hospital. In March 2000 he developed polyarthritis. He presented with a bloody rectal discharge and low-grade fever in April 2000. On admission, examination revealed that he was emaciated. Vital signs were as follows: blood pressure 118/68 mmHg; pulse rate 64/minute; temperature 37.0°C. Findings from the pulmonary and cardiovascular examination were normal. Musculoskeletal examination revealed mild synovitis. Laboratory findings included a erythrocyte sedimentation rate of 71 mm/h (Westergren); normal urinalysis; white blood cell count 2,400 /mm³ with 27% lymphocytes; red blood cell count 356 x 10⁹/mm³; hemoglobin 11.7 g/dl; platelet count 18 x 10⁹/mm³; fibrinogen 439 mg/dl (normal < 400); albumin 3.3 g/dl (normal > 3.8); blood urea nitrogen 11 mg/dl (normal < 23); creatinine 7 mg/l (normal < 12); lactate dehydrogenase 193 IU/L (normal < 220); glucose 73 mg/dl (normal < 110); and C-reactive protein 0.27 mg/dl. The serum concentration of IgG was elevated, whereas IgA and IgM were within the normal range. Complement C3 was 27 mg/dl (normal 60 - 120) and C4 was 6 mg/dl (normal 12 - 40). Immune complexes and cryoglobulin were detected. The antinuclear antibody was positive (1:1,280 with a diffuse and speckled pattern). An LE cell preparation was positive. Antibodies to SS-A/Ro were posi-
CASE REPORT

Rectal ulcers in SLE / S. Yuasa et al.

The anti-DNA antibody titer was elevated to 233 IU/ml. Autoantibodies to SS-B/La, U1RNP, and Sm were not detected. Anti-cardiolipin antibody and myeloperoxidase-antineutrophil cytoplasmic antibody (MPO-ANCA) were negative. Rheumatoid factor was negative. Urinalysis revealed proteinuria (0.6 g/day); the sediment contained 20 red cells and an occasional granular cast per high-power field. Stool samples were positive for occult blood. Chest and abdomen roentgenograms showed no remarkable findings. A double-contrast barium enema examination demonstrated erosive changes of the mucosa, and narrowing of the rectum with marked deformity (Fig. 1). Colonoscopy disclosed multiple round ulcers at the rectum accompanied by white fur. The surrounding mucosa was red and prone to bleeding (Fig. 2a). The rest of the colon up to the cecum was normal. The ulcer and tissue at its periphery were biopsied, and non-specific inflammatory tissue accompanied by mild mononuclear cell invasion and vascular hyperplasia were documented. There was no evidence for vasculitis.

The present patient fulfilled 5 of the 11 revised criteria for the classification of SLE. The absence of any positive evidence for infective agents and the presence of rectal ulcers were felt to be consistent with a gastrointestinal manifestation of SLE. Treatment with prednisolone (PSL) at 40 mg/day resulted in marked symptomatic improvement. Nine weeks after the initiation of steroid therapy, colonoscopy was repeated and showed healing of the ulcers (Fig. 2b). The patient has remained well for 12 months on tapering doses of PSL.

Discussion

Gastrointestinal manifestations are of-
ten seen in patients with SLE (1,2). Although the type of gastrointestinal manifestation may vary, serious complications such as perforation, hemorrhage, infarction and ulceration occur with an increasing frequency (1-3). Our case is unique in several respects, the first being that there were no other concurrent SLE manifestations when this patient first presented with rectal ulcer and that the characteristic lupus symptoms and findings first appeared 5 years later. He was followed up under a diagnosis of non-specific enteritis, although several immunological abnormalities such as a positive ANA test, positive anti-SS-A/Ro antibody, and slightly elevated anti-DNA antibody titers were seen. Gastrointestinal manifestations relapsed along with the appearance of SLE-specific symptoms and laboratory findings. Finally, he fulfilled 5 of the American College of Rheumatology criteria for the classification of SLE. These were polyarthritis, renal involvement, hematological disorder, immunological disorder (positive LE cell and anti-DNA antibody), and a positive ANA test. Therefore, he was diagnosed as having definite SLE. A review of the literature showed that there have been only a few reported cases in which the patient presented with severe gastrointestinal complications as the initial manifestation of SLE (4-7). In most cases, the time elapsed from the onset of gastrointestinal manifestations to the diagnosis of SLE was short (4-7). The second unique aspect of our case was rectal involvement in the form of ulceration, but with sparing of the rest of the colon. In most cases of SLE, the left, transverse, and right colon may be involved (1, 2). There have been only 9 case reports of SLE (including the present one) in which rectal involvement due to vasculitis was seen (Table I) (8-15). Seven of these 9 cases had lesions localized to the rectum. The most common type of lesion was ulcerous. Perforation was observed in 4 cases. Histopathological examination provided evidence for vasculitis in 6 cases. In our patient, no findings of vasculitis were obtained from a biopsy specimen. However, it was thought that the rectal ulcers were caused by vasculitis on the basis of the following facts: (i) vasculitis is difficult to prove by endoscopic biopsy; (ii) the angiography of the inferior mesenteric artery displayed an image of irregular stenosis and interruption; and (iii) the rectal ulcer responded to steroid therapy, showing impressive improvement. The presence of vasculitis was also suggested in the other 2 cases based on the clinical course and findings from angiography. It has been reported that the abdominal vasculitis which occurs in patients with SLE correlates with vasculitis in other organs (3). However, this was not necessarily true in the cases of rectal involvement. Six of 9 patients (including our case) in whom lupus nephritis developed later, had no active organ involvement at the onset of the gastrointestinal manifestations. Guy et al. described a similar case of a patient with SLE who developed isolated rectal ulcers due to vasculitis (15). In their case, the dangerous ulcer was present for 3 years. The reason why rectal involvement in SLE is rare and sometimes persists might be related to the organ’s rich, dual-source blood supply, as suggested previously (13, 15).

It is recommended to use high-dose corticosteroids for the treatment of SLE with gastrointestinal vasculitis. Regarding previous cases of rectal vasculitis, PSL was administrated in 5 cases. Five patients received surgery for perforation, tumor and necrosis. Unfortunately 4 patients died despite the above therapy. It should be noted that the prognosis of patients presenting with perforation was extremely poor. Our patient responded sufficiently to 40 mg of PSL and his ulcers improved dramatically along with the other active lupus symptoms within 9 weeks. As it has been found that some patients relapse when the corticosteroid dose is tapered, careful follow-up is required.

The present case illustrates the fact that rectal ulcer is an extremely rare but possible initial manifestation of SLE. Increased awareness of this possibility may lead to early diagnosis and prompt treatment.

References
1. Hoffman BI, Katz WA: The gastrointestinal manifestations of systemic lupus erythe-

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Table I. Reported cases of SLE complicated with rectal involvement due to vasculitis

<table>
<thead>
<tr>
<th>No.</th>
<th>Age/Sex</th>
<th>Location</th>
<th>Rectal involvement Type</th>
<th>Vasculitis</th>
<th>Activity of SLE</th>
<th>Therapy</th>
<th>Prognosis</th>
<th>Reference</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>28/F</td>
<td>R</td>
<td>Ulcer, Perforation</td>
<td>(+)*</td>
<td>(+)</td>
<td>PSL 60 mg</td>
<td>Dead</td>
<td>Tsugu et al. (1978) (8)</td>
</tr>
<tr>
<td>2</td>
<td>18/F</td>
<td>R-S</td>
<td>Ulcer (multiple)</td>
<td>(+)*</td>
<td>(+)</td>
<td>PSL 60 mg</td>
<td>Alive</td>
<td>Yagita et al. (1981) (9)</td>
</tr>
<tr>
<td>3</td>
<td>43/F</td>
<td>R</td>
<td>Tumor</td>
<td>(+)*</td>
<td>(-)</td>
<td>Operation</td>
<td>Dead</td>
<td>Palvio et al. (1987) (10)</td>
</tr>
<tr>
<td>4</td>
<td>39/F</td>
<td>R-S</td>
<td>Ulcer (multiple), perforation</td>
<td>(+)</td>
<td>(+)</td>
<td>PSL 60 mg</td>
<td>Dead</td>
<td>Iida et al. (1991) (11)</td>
</tr>
<tr>
<td>5</td>
<td>31/F</td>
<td>R</td>
<td>Perforation</td>
<td>(+)*</td>
<td>(-)</td>
<td>Operation</td>
<td>Dead</td>
<td>Igarashi et al. (1991) (12)</td>
</tr>
<tr>
<td>6</td>
<td>54/F</td>
<td>R</td>
<td>Necrosis</td>
<td>(+)*</td>
<td>(-)</td>
<td>Operation</td>
<td>Alive</td>
<td>Reissman et al. (1994) (13)</td>
</tr>
<tr>
<td>7</td>
<td>41/M</td>
<td>R</td>
<td>Ulcer, Perforation</td>
<td>(+)</td>
<td>(-)</td>
<td>Operation</td>
<td>Alive</td>
<td>Teramoto et al. (1999) (14)</td>
</tr>
<tr>
<td>8</td>
<td>34/F</td>
<td>R</td>
<td>Ulcer</td>
<td>(+)*</td>
<td>(-)</td>
<td>PSL + Operation</td>
<td>Alive</td>
<td>Guy et al. (1999) (15)</td>
</tr>
<tr>
<td>9</td>
<td>27/M</td>
<td>R</td>
<td>Ulcer (multiple)</td>
<td>(+)</td>
<td>(-)</td>
<td>PSL 40 mg</td>
<td>Alive</td>
<td>Present case</td>
</tr>
</tbody>
</table>

R: Rectum; S: Sigmoid colon; PSL: Prednisolone.
*Vasculitis was proven by histopathological examination.
CASE REPORT

Rectal ulcers in SLE / S. Yuasa et al.