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Case report

Ileal perforation as an initial manifestation of systemic lupus erythematosus: A case report

Bibek Man Shrestha [a,*], Suraj Shrestha [a], Sanjeev Kharel [a], Ajay K.C. [b], Sujan Shrestha [b], Sumita Pradhan [b], Ramesh Singh Bhandari [b]

[a] Maharajgunj Medical Campus, Institute of Medicine, Kathmandu, Nepal
[b] Department of GI and Hepatobilliary Surgery, Tribhuvan University Teaching Hospital, Kathmandu, Nepal

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ABSTRACT

Introduction and importance: Lupus enteritis is uncommon in patients with SLE and usually presents with anorexia, vomiting, and abdominal pain. Intestinal perforation as an initial manifestation of SLE is rare and can have a grave prognosis if not timely diagnosed.

Case history: We report an unusual case of a 22-year-old regularly menstruating female who presented with features of perforation peritonitis as an initial manifestation of lupus enteritis. Intraoperatively, a gangrenous ileal segment with multiple perforations was present. Thus, with an intraoperative diagnosis of perforation peritonitis, a gangrenous segment of the small bowel was resected and a double-barrel jejuno-ileostomy was created.

Discussion: Lupus enteritis manifesting initially as bowel perforation can be an uncommon cause of acute abdomen. A plain chest X-ray can show gas under the diaphragm suggesting bowel perforation. A contrast-enhanced CT scan of the abdomen is the gold standard in diagnosing lupus enteritis with a good prognosis on steroids.

Conclusion: Primary closure, resection, and anastomosis of small gut or diverting stoma are required for management of perforation. A high degree of clinical suspicion is required for early diagnosis thus preventing the grave prognosis of such an entity.

1. Introduction

Systemic lupus erythematosus (SLE) is a multisystem disorder mostly affecting women. Any part of the gastrointestinal tract can be affected with varying manifestations and more than 50% of SLE patients can have gastrointestinal symptoms in their disease course [1]. Most common symptoms include nausea, vomiting, and anorexia while symptoms like abdominal pain, diarrhea, and abdominal distension can be a manifestation of serious underlying GI involvement, infections, and/or treatment complications [2]. Lupus enteritis occurs in only about 0.2% to 5.8% of patients with SLE and is the vasculitis or inflammation of the small bowel [3]. Though Lupus enteritis has an excellent prognosis, however, intestinal necrosis leading to intestinal perforation can develop if not managed timely which is potentially fatal [1,4].

Here, we report a case of SLE presenting initially with ileal perforation managed by exploratory laparotomy and resection of a perforated portion. This case has been reported in line with SCARE criteria [5].

2. Case presentation

A 22-year $P_{2+1}L_1$ Mongolian female regularly menstruating, non-alcoholic and non-smoker without any prior surgical history or any family history of malignancy presented to our center with complaints of continuous mild non-radiating pain over the periumbilical region for 12 days with associated symptoms such as nausea, vomiting, fever and abdominal distension for the last 2 days. She denied constipation or obstipation, decreased appetite, cough, hematochezia/melena, significant weight loss, and trauma to the abdomen. The patient had a normal bowel and bladder habit and no active tuberculosis.

On examination, she was ill-looking with a blood pressure (BP) of 100/60 mm Hg, pulse rate of 100 bpm, respiratory rate of 24breaths per minute, and oxygen saturation of 94% in the room air. On per abdomen examination, the whole of the abdomen was distended with tender and generalized rigidity on palpation suggesting peritonitis. On per rectal examination, the rectum was filled with stool normal in contour with no

* Corresponding author.
E-mail address: bibekmanstha7@gmail.com (B.M. Shrestha).

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blood. She was anemic (hemoglobin-8.2 g/dl, PCV-29 g %) with raised total leukocyte count (TC-14000/mm^3) and normal platelet count of 200,000/mm^3. A plain chest X-ray revealed gas under the diaphragm. Thus, the diagnosis of bowel perforation, and peritonitis was made and proceeded for Emergency Laparotomy.

The peritoneal cavity was accessed via a midline incision. Intraoperatively, a gangrenous segment of the small bowel was noted 120 cm distal to duodenojejunal flexure to 15 cm proximal from the ileocolic junction with 2 perforations of size 1.5 cm each on the on the antimesenteric border of gangrenous segment located approximately 45 and 50 cm proximal to ileocolic junction. There was approximately 500 ml purulent fluid in the peritoneal cavity along with inter-loop adhesions of ileum (Figs. 1 and 2). Although the cause of perforation was noted to be bowel gangrene, the cause of ischemia leading to gangrene could not be established intraoperatively. There were no bands, adhesions, volvulus that could lead to segmental ischemia of bowel. Similarly, there were no tubercular deposits, bowel or mesenteric thickening, mesenteric lymphadenopathy that would suggest tuberculosis or serosal fat wrapping that would suggest Crohn's disease as a possible cause of perforation. The gangrenous segment of the small bowel was resected and double barrel jejuno-ileostomy was created because of the severe peritoneal contamination, edematous bowel, low albumin level (24 g/l) and threatened viability of the resected margins. One unit of whole blood was transfused intraoperatively. Otherwise, the operative procedure was uneventful and patient remained hemodynamically stable throughout the procedure.

Postoperatively, she was clinically stable, the stoma was healthy and functioning well, and was tolerating a soft diet orally. However, on the 7th postoperative day, she suddenly developed bluish-blackish discoloration of fingertips and toes suggestive of the Raynaud phenomenon. The case was then evaluated by the rheumatology team. Thromboembolic etiologies were ruled out after normal findings on echocardiography, Doppler USG, and CT angiography. However, anti-ds DNA, hsCRP, anti-CCP antibody, and anticardiolipin antibody were positive suggesting SLE as the likely cause of vasculitis. She had also subsequently developed autoimmune hemolytic anemia evident by anemia, incompatibility on blood cross-matching, and positive Direct Coombs test. Later, histopathology also supported segmental small intestine gangrene with perforation due to thrombotic phenomenon. Following findings were noted on histopathology: Perforated sites showed inflammatory granulation tissue with plenty of acute and chronic inflammatory cells. Gangrenous areas had necrosis, congestion and dilated blood vessels. Occluded vessels with thrombus were noted on mesenteric sections.

Although the cause of gangrene and perforation was not evident initially, the clinical course on the postoperative period was suggestive of SLE with lupus vasculitis as the most likely cause of segmental small bowel gangrene leading to perforation. Hence, the final diagnosis of active SLE with Ileal perforation due to lupus vasculitis was established. She was then started on steroids. She had gradual clinical improvement and was discharged on the 21st postoperative day with plans to start immunosuppressants on follow-up and ileostomy reversal after 2 months. Since the patient hailed from the remote region of Nepal, further follow-up was advised at the regional health facility. At discharge, she was clinically stable, tolerating a normal diet, and stoma output was controlled with daily wound care.

3. Discussion

The commonly found gastrointestinal disorders associated with SLE patients are protein-losing enteropathy, lupus mesenteric vasculitis, acute pancreatitis, intestinal pseudo-obstruction, inflammatory bowel disease, and celiac disease [6]. Lupus enteritis as the sole initial manifestation of active SLE is rarely found only in 0.2–5.8% of patients

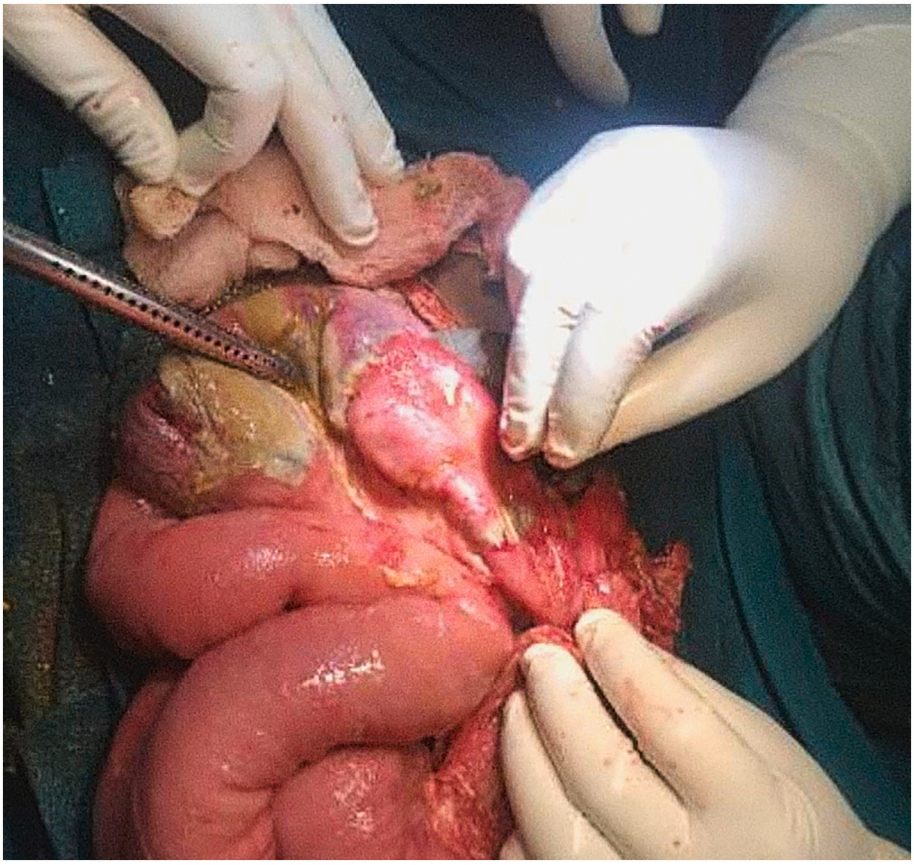

Fig. 1. Shows gangrenous ileal segment.

Fig. 2. Shows resected gangrenous ileal specimen with perforations.

without previous diagnosis [7].

The cause behind lupus mesenteric vasculitis is the formation of immune complexes deposition in blood vessels by circulation autoantibodies leading to thrombosis and inflammation of vessels supplying the intestine. Thus, lack of blood supply to the area of the intestine can cause ulceration, infarction, and eventually perforation [8,9]. In our case, the patient was an unknown case of SLE diagnosed only after surgery and further evaluation. Lupus enteritis complicating to infarction and finally, intestinal perforation is the most probable reason for the above-mentioned complaints. Non-specific symptoms like abdominal pain (97%), ascites (78%), nausea (49%), vomiting (42%), diarrhea (32%), and fever (20%) are common symptoms with lupus enteritis [10]. Similar vague symptoms also occurred in our patient. Lupus nephritis is an additional concern in patients with lupus enteritis present in about 65% of cases [10,11]. Signs of lupus nephritis were not present in our patient.

Under laboratory investigation as reported in previous cases, positive ANA is seen in 100%, positive ds-DNA in 80%, low complement levels in 70%, and positive anti-Smith antibodies in 20% of cases. Lymphopenia, hypocomplementemia, and normal C reactive protein are some of the other findings [12]. In contrast, our patient had negative ANA and high levels of C reactive protein. In about 5% of cases, negative ANA can be found. Such patients have clinical manifestations like skin rashes, photosensitivity, Reynaud phenomenon, and serositis [13]. This finding is similar to our patient. A contrast-enhanced CT (CECT) scan of the abdomen is the gold standard technique for diagnosing lupus enteritis [10,11]. Submucosal edema of the jejunum and ileum, leading to classic findings of circumferential bowel wall thickening (target sign) and dilation of intestinal segments, and engorgement of mesenteric vessels (comb sign) are seen in lupus enteritis [14]. Since our patient presented with features of perforation peritonitis, she was rushed to emergency operation theatre as CECT abdomen is not done in cases suspected of perforation peritonitis.

The management of ileal perforations includes primary closure, resection, and anastomosis of small gut or diverting stoma, depending on the site and number of perforations, severity of peritonitis, and condition of the patient [15]. Lupus enteritis can have multiple or singular forms of lesions so attention should be given during exploration [13]. In our patient also similar procedure was done and a double barrel ileostomy was made. SLE patients are more prone to the risk of surgical intervention compared with those without SLE [16]. So, proper attention is required for such patients.

Lupus enteritis is found to have a good prognosis with steroids but complications like perforations can cause mortality up to 2.7% [10]. Takashi et al. found half of the patients (6 of 11 patients) with SLE had intestinal perforation as a complication and died [17]. Though the

prognosis of SLE patients with intestinal perforation is poor but early diagnosis and surgical treatment are useful for the management of the disease [18].

4. Conclusion

Intestinal perforation can be an uncommon presentation of mesenteric vasculitis in a patient with SLE which can have a grave prognosis if there is a delay in diagnosis and management. Hence a high clinical suspicion can aid in early diagnosis in patients with acute abdomen and associated clinical features of SLE. It rarely can be the initial manifestation of SLE.

Consent for publication

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

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Ramesh Singh Bhandari (RSB), Sumita Pradhan (SP) and Sujan Shrestha (SS) = Study concept, Data collection, and surgical therapy for the patient
Bibek Man Shrestha (BMS), Sanjeev Kharel (SK), Suraj Shrestha (SS) and Ajay K.C. (AK) = Writing - original draft preparation
BMS, SS and SK = Editing and writing
RSB and SP = senior author and manuscript reviewer.

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Declaration of competing interest

None.

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