

# Febrile pancytopenia, hematochezia and hematemesis as the presenting manifestations of systemic lupus erythematosus

## Abstract

Systemic lupus erythematosus (SLE) may extremely rarely present with gastrointestinal symptoms. Gastrointestinal involvement in SLE is not uncommon, but more than half of these manifestations are attributed to adverse reactions to lupus medications and viral or bacterial infections, which occur more commonly in these immunosuppressed patients. Herein, we describe a female patient who presented with fever of unknown origin and eventually with hematochezia and hematemesis, without any abdominal pain.

**Keywords:** pancytopenia, hematochezia, hematemesis, hematochezia

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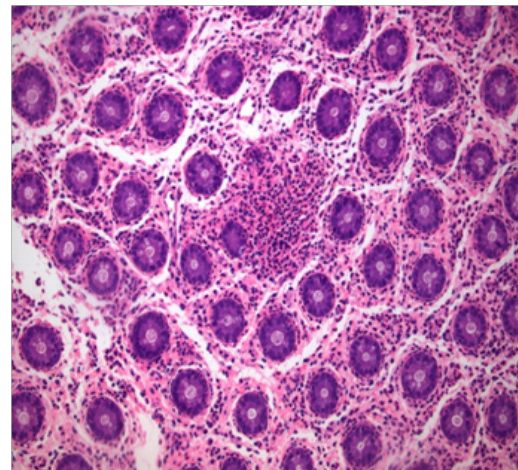
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## Introduction

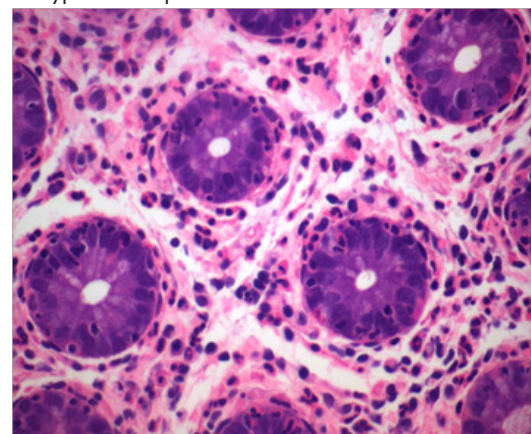
Gastrointestinal involvement per se in SLE usually presents with abdominal pain due to vasculitis, pseudo-obstruction, pancreatitis or peritonitis. Presentation with gastrointestinal hemorrhage is extremely rare. We describe a patient who was admitted to our hospital due to febrile pancytopenia and who developed hematochezia and hematemesis, which were attributed to gastrointestinal involvement of SLE.

## Case presentation

A twentyyears old female patient presented to the Emergency Department of our Hospital due to febrile pancytopenia. From her past medical history, the patient had autoimmune thyroiditis. Fever had started twenty days before and reached 40°C. Apart from fever, the patient complained for anorexia, but had no other specific symptoms. On examination, she had a mildly enlarged liver and spleen. Laboratory findings included anemia (Hb=8.4g/dl), leukocytopenia (WBC=2.350mm<sup>3</sup>) and thrombocytopenia (PLT's=49.000mm<sup>3</sup>). Computed tomography (CT) of the thorax revealed a small left pleuritic effusion, while abdominal CT confirmed an enlarged liver and spleen. The patient underwent bone biopsy, which showed only a reactive bone marrow. ANA were positive in a title >1/640 and anti-dsDNA were positive (60 U/mL), while C4 serum levels were very low (5,46mg/dL). Seven days after her admission to the hospital, the patient had hematochezia. A sigmoidoscopy revealed the presence of petechial mucus in the rectus and the sigmoid colon and biopsies were taken. Histopathology of these petechiae was suggestive of colitis due to autoimmune disease (Figures 1 & 2). Due to the occurrence of hematemesis as well, an upper gastrointestinal endoscopy was performed, which showed the presence of petechiae in the duodenum, too. The patient was administered 1g methylprednisolone intravenously (iv) for three days and afterwards she was treated with prednisone 1mg/Kg iv, after which the fever resolved, she experienced no other incidence of hematochezia nor hematemesis and her hematological parameters returned to normal.



**Figure 1** Preserved crypt architecture, increased lamina propria inflammation, moderate crypt mucin depletion.



**Figure 2** Apoptotic activity, intraepithelial lymphocytes and neutrophils.

## Discussion

Gastrointestinal involvement in SLE is common, but more than half of these manifestations are attributed to adverse reactions to lupus medications and viral or bacterial infections, which occur more commonly in these immunocompromised patients.<sup>1-4</sup> However, gastrointestinal involvement per se is not as often as other more common manifestations of SLE, such as lupus serositis or nephritis.<sup>5</sup> It is noteworthy that the clinical manifestations of the gastrointestinal system, when implicated in SLE patients may vary from life-threatening to mild, even insidious.<sup>6-9</sup>

Gastrointestinal vasculitis, with or without infarction, is one of the most serious complications of SLE. Because of the paucity of cases of lupus enteritis, no randomized trials have come out with the best treatment options. Many cases of successful treatment of intestinal vasculitis with high dose prednisolone, (1mg/kg/day), alone have been reported. For corticosteroid-resistant GI vasculitis, there have reports of successful outcomes with intravenous methylprednisolone and cyclophosphamide.<sup>4-11</sup> In cases of perforation, the importance of early laparotomy must be emphasized, because of the otherwise high mortality. In general, the outcome in patients with perforation is poor, with death occurring in more than two thirds of cases.<sup>10,11</sup>

Hematochezia and hematemesis are very rare manifestations of SLE gastrointestinal involvement.<sup>5-9</sup> This is the first case of SLE presenting with hematemesis and hematochezia. Another case has been described in 1999 and presented with fever, rash and massive hematemesis with maleness, but no lower gastrointestinal involvement, as our patient did.<sup>12</sup>

## Conclusion

Lupus mesenteric vasculitis is the most common cause of gastrointestinal involvement in SLE, followed by protein-losing enteropathy, intestinal pseudo-obstruction, acute pancreatitis, acute hepatitis and complications, such as celiac disease and inflammatory bowel diseases. The presentation of our patient with fever, pancytopenia, hematochezia and hematemesis is indicative of the multifaceted nature of this systemic disease.

## Acknowledgments

None.

## Conflicts of interest

Author declares that there is no conflicts of interest.

## References

1. Takeno M, Ishigatsubo Y. Intestinal manifestations in systemic lupus erythematosus. *Intern Med.* 2006;45(2):41–42.
2. Endo H, Kondo Y, Kawagoe K, et al. Lupus enteritis detected by capsule endoscopy. *Intern Med.* 2007;46(18):1621–1622.
3. Prouse PJ, Thompson EM, Gumpel JM. Systemic lupus erythematosus and abdominal pain. *Br J Rheumatol.* 1983;22:172–175.
4. Lian TY, Edwards CJ, Chan SP, et al. Reversible acute gastrointestinal syndrome associated with active systemic lupus erythematosus in patients admitted to hospital. *Lupus.* 2003;12(8):612–616.
5. Xin-Ping Tian, Xuan Zhang. Gastrointestinal involvement in systemic lupus erythematosus: Insight into pathogenesis, diagnosis and Treatment. *World J Gastroenterol.* 2010;16(24):2971–2977.
6. Lee CK, Ahn MS, Lee EY, et al. Acute abdominal pain in systemic lupus erythematosus: focus on lupus enteritis (gastrointestinal vasculitis). *Ann Rheum Dis.* 2002;61(6):547–550.
7. Grimbacher B, Huber M, Kempis J, et al. Successful treatment of gastrointestinal vasculitis due to systemic lupus erythematosus with intravenous pulse cyclophosphamide: a clinical case report and review of the literature. *Br J Rheumatol.* 1998;37(9):1023–1028.
8. Bert J, Gertner E. Lupus Gastrointestinal Tract Vasculopathy: Lupus “Enteritis” Involving the Entire Gastrointestinal Tract from Esophagus to Rectum. *Case Rep Gastroenterol.* 2017;11(1):48–53.
9. Yuasa S, Suwa A, Hirakata M, et al. A case of systemic lupus erythematosus presenting with rectal ulcers as the initial clinical manifestation of disease. *Clin Exp Rheumatol.* 2002;20(3):407–410.
10. Medina F, Ayala A, Jara LJ, et al. Acute abdomen in systemic lupus erythematosus: the importance of early laparotomy. *Am J Med.* 1997;103(2):100–105.
11. Drenkard C, Villa AR, Reyes E, et al. Vasculitis in systemic lupus erythematosus. *Lupus.* 1997;6(3):235–242.
12. Hiraishi H, Konishi T, Ota S, et al. Massive gastrointestinal hemorrhage in systemic lupus erythematosus: successful treatment with corticosteroid pulse therapy. *Am J Gastroenterol.* 1999;94(11):3349–3353.