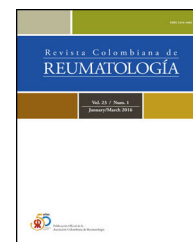




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Case report

Pseudo-obstruction, an infrequent Lupus manifestation: A case report



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ABSTRACT

Gastrointestinal involvement in SLE has been reported in up to 50%, generally secondary to the adverse effects of treatment. Intestinal pseudo-obstruction is caused by hypomotility related to ineffective propulsion. The case of a 51-year-old patient with intestinal obstruction is presented. She was taken to surgical management due to suspicion of adhesions, with a stationary clinical course; the control tomography documented loop dilation and bilateral hydronephrosis, associated with markers of lupus activity. It was managed as an intestinal pseudo-obstruction due to SLE with resolution of her symptoms. High diagnostic suspicion results in timely treatment and the reduction of complications.

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Pseudoobstrucción intestinal, una infrecuente manifestación del lupus: reporte de caso

RESUMEN

El compromiso gastrointestinal en lupus eritematoso sistémico (LES) ha sido reportado hasta en un 50%, generalmente secundario a los efectos adversos del tratamiento. La pseudoobstrucción intestinal es causada por hipomotilidad relacionada con una propulsión inefectiva. Se presenta el caso de una paciente de 51 años, con obstrucción intestinal por sospecha de bridas, que fue llevada a manejo quirúrgico y tuvo una evolución clínica estacionaria. La tomografía de control documentó dilatación de asas e hidronefrosis bilateral, en tanto que los paraclínicos mostraron actividad lúpica. Se manejó como una

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pseudoobstrucción intestinal por LES con resolución del cuadro. La alta sospecha diagnóstica favorece el tratamiento oportuno y la disminución de las complicaciones.

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Introduction

Systemic lupus erythematosus (SLE) is an autoimmune disease of the connective tissue that presents a wide range of systemic manifestations, including gastrointestinal; it is observed in up to 50% of cases^{1,2}; mostly attributed to adverse drug effects^{3,4}. Those related to the disease are less frequent, including intestinal pseudo-obstruction, a disorder frequently associated with changes in the urinary tract^{1,5,6}. A case of a patient with long-standing SLE who consulted for postprandial vomiting and abdominal distension of several months of evolution is described. Initially, it was managed as intestinal obstruction due to flanges, and later, intestinal pseudo-obstruction due to lupus was diagnosed.

Case report

A 51-year-old female patient with a diagnosis of SLE since the age of 17, with cutaneous and pleuropericardial involvement (pericarditis and recurrent pleuritis that required pleurodesis), who was treated with prednisolone 5 mg/day, without rheumatology follow-up. The patient consulted due to symptoms lasting eight months, with weight loss of approximately 30 kg, chronic diarrhea that was neither dysenteric nor lienenteric, abdominal distension, postprandial vomiting, asthenia, and adynamia. In the 15 days before the consultation, she presented an increase in bloating, nausea, and vomiting 30 min after meals, as well as generalized abdominal pain. The surgery group treated it as an intestinal obstruction since her abdominal contrast tomography showed dilation of all the loops of the small intestine, with no signs of distress up to the terminal ileum, which collapsed together with the cecum.

The diagnosis of congenital bands was raised since the patient had no history of abdominal surgery or intestinal stenosis due to vasculitis. In the exploratory laparotomy, multiple flanges were documented that were released; how-

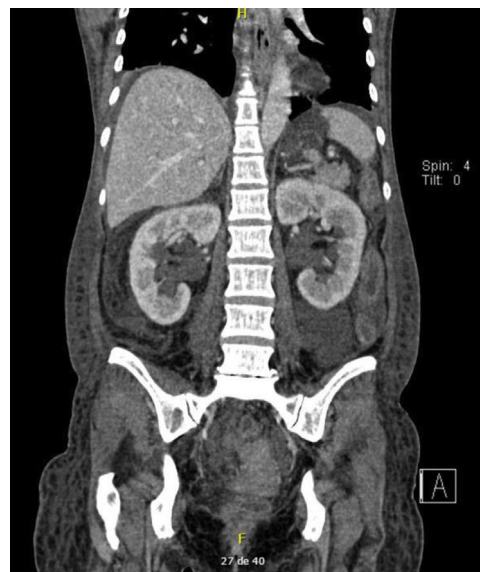


Fig. 2 – Bilateral dilatation of the ureters is observed.

ever, there was no favorable clinical response. The control abdominal contrasted CT, requested with the phase of contrast excretion for an adequate evaluation of the urinary tract, showed persistent dilation of intestinal loops and bilateral hydroureteronephrosis associated with inflammatory changes of the bladder (Figs. 1 and 2), for which intestinal pseudo-obstruction due to SLE was suspected.

Laboratory studies were compatible with lupus activity: hypocomplementemia, lymphopenia, and positive anti-dsDNA, without renal involvement (Table 1). Additionally, on admission, hypokalemia developed, but during hospitalization, potassium replacement was instituted, and on the fifth day normal values were achieved, although the symptoms persisted (Table 2). Immunosuppression was started with pulses of methylprednisolone and cyclophosphamide

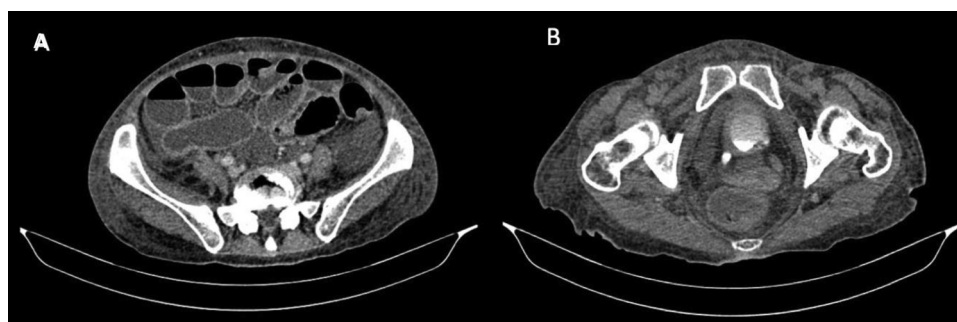


Fig. 1 – A) Dilation of the small intestine with air-fluid levels and absence of the “halo” sign was observed. B) Thickened bladder wall with inflammatory changes.

Table 1 – Initial paraclinical and immunological profile.

Paraclinical	Value	Reference value
ALT	14 U/L	0–55
AST	20 U/L	5–34
Total bilirubin	0.24 mg/dL	0.2–1.2
Direct bilirubin	0.06 mg/dL	0.1–0.5
Calcium	7.4 mg/dL	8.4–10.2
Sodium	131 mmol/L	136–145
Potassium	2.94 mmol/L	3.5–5.1
Magnesium	1.66 mg/dL	1.6–2.6
Albumin	2 g/dL	3.5–5
Creatinine	0.57 mg/dL	0.6–1.1
Blood urea nitrogen	15.5 mg/dL	9.8–20.1
C-reactive protein	3.01 mg/dL	0.01–0.82
Erythrocyte sedimentation rate	3.01 mm/h	0–20
Hemoglobin	10.3 g/dL	12–15
Medium corpuscular volume	77 fL	80–98
Leukocytes	3700 mm ³	4500–11,000
Neutrophils	2500 mm ³	1500–6600
Lymphocytes	600 mm ³	1500–3500
Platelets	239,000 mm ³	150,000–450,000
C3	52 mg/dL	83–193
C4	9 mg/dL	15–45
24-hour proteinuria	0.7 g/24 h	
Immunologic profile		
Anti-DNA	1:320	
Anti-RNP	155 - Positive	<15
Anti-Sm	26.5 - Positive	<15
Anti-Ro	5.2 - Negative	<15
Anti-La	2.7 - Negative	<15

500 mg/m², after which the patient presented improvement in her symptoms, with the disappearance of vomiting, diarrhea, and abdominal distension. The flanges described in the surgical procedure, although there was no reported background, were probably secondary to episodes of subclinical serositis.

Discussion

A case of intestinal pseudo-obstruction due to SLE is presented in a patient with diarrhea, abdominal distension, weight loss, and intractable vomiting. Gastrointestinal manifestations can be as common as 50% of patients^{1,2}, mostly related to adverse drug effects or infectious complications associated with immunosuppression, but more cases linked to the baseline disease have been described⁴.

Gastrointestinal tract involvement in SLE is variable and can impact the mouth to the rectum, with mucosal affection being more frequent, especially oral². Following these, the manifestations most described in the literature are protein-losing enteropathy, liver involvement, pancreatitis, and intestinal pseudo-obstruction (Table 3)^{2,7}.

Intestinal pseudo-obstruction is defined as a process of intestinal hypomotility related to ineffective propulsion, which produces the classic symptoms of intestinal obstruc-

Table 3 – Gastrointestinal manifestations in SLE.

Manifestation	Prevalence
Protein-losing enteropathy	60%
Lupus hepatitis	30%
Pancreatitis	23%
Intestinal pseudo-obstruction	53%–69%
Lupus enteritis	0.2%–5.8%; reports up to 47%

tion, but without an anatomical cause^{1,8}. The symptoms of this condition are usually postprandial, start acutely, and are a reason for early emergency consultation; however, their duration from three days to three years has been reported, with the possibility of chronic intestinal obstruction, malnutrition, and weight loss^{1,6,7}.

In the largest systematic review carried out to date, published in 2017, which included 150 patients with intestinal obstruction, the most frequent symptoms were reported as abdominal pain (85%), nausea and vomiting (82%), being less frequent abdominal distension (62%), diarrhea, and constipation⁷. Other case reports report similar frequencies of symptoms, with abdominal pain always being more prevalent⁹.

In addition to the symptoms related to intestinal pseudo-obstruction, visceral muscular dysmotility syndrome is described, which occurs in 3/4 of the patients, in which hydronephrosis and dilation of the biliary tract are observed, which produces symptoms related to interstitial cystitis^{1,4,6,7}.

The pathogenesis of intestinal pseudo-obstruction is still not fully clarified. Processes related to the involvement of visceral smooth muscle, enteric nerves, the autonomic nervous system, and vasculitis of the intestinal wall have been described^{2,4,6,10}. Histopathological studies have found repeatedly affection of the small intestine, such as involvement mainly of the muscular layer, with the presence of edema and intestinal inflammation, infiltration of inflammatory cells (predominantly eosinophils), myocyte necrosis related to the underlying inflammation, atrophy, and fibrosis^{4,11,12}. These findings suggest that the pathophysiological process could have the myocyte as its primary target, rather than being related to vasculitis due to SLE^{11,13}.

The diagnosis of intestinal pseudo-obstruction is based on the symptoms and the background, since it generally occurs in patients with a previous diagnosis of SLE, although less frequently it can appear at disease onset². Lupus activity should always be evaluated; the most common association is hematological involvement, which occurs in 75% of the cases⁷.

An abdominal x-ray in a standing position is usually the first diagnostic aid in which the classic signs of intestinal obstruction are evident, such as dilated loops and air-fluid levels^{6,12}. Contrast-enhanced abdominal tomography makes it possible to define the absence of an anatomical cause related

Table 2 – Evolution of potassium values during hospitalization.

	On admission	Day 2	Day 3	Day 4	Day 5	Day 14
Potassium	2.94 mmol/L	2.83 mmol/L	3.02 mmol/L	3.31 mmol/L	3.83 mmol/L	4.81 mmol/L

to the symptoms and confirms the presence of dilated loops, thickening of the wall, and if present, hydronephrosis and dilated biliary tract⁶. The requested paraclinical tests should be the same used as the approach to patients with lupus, searching for activity and organ involvement, since no antibody specifically associated with intestinal pseudo-obstruction has been found, although a high prevalence of antiRo/SSA has been observed, and, in some reports, antiLa/SSB^{7,10,12,14}. In this patient, these antibodies were negative.

Treatment in these patients has been based on support measures and immunosuppression. On the one hand, symptomatic management includes prokinetics, abdominal decompression with a nasogastric tube, and total parenteral nutrition (if necessary), especially in subjects with chronic obstruction^{4,6,7}. In some cases, the use of broad-spectrum antibiotic therapy has been considered as part of management, to avoid the risk of bacterial overgrowth^{4,6}. Regarding immunosuppressive management, the most used and has been shown to achieve remission in isolated cases, are methylprednisolone pulses followed by oral prednisolone, associated with cyclophosphamide; some response has also been demonstrated with azathioprine, cyclosporine A, and tacrolimus^{2,4,7,9}.

The prognosis is generally favorable, with response rates of up to 93%, sustained remission of up to 76%⁷, and greater relapse in patients who were initially surgically treated or with a delay in diagnosis^{9,14}. Mortality ranges from 6.99% to 18%, especially due to infections, cerebral or meningeal hemorrhage, or lupus involvement of other organs^{1,4,7}.

In conclusion, we present the case of a patient who presented with abdominal distension, persistent vomiting, diarrhea, and weight loss, who was diagnosed with intestinal pseudo-obstruction secondary to baseline SLE, and who had a favorable response to immunosuppressive and supportive treatment. This disorder, which is rare and of which few cases have been published, is important to recognize for the administration of adequate and prompt treatment.

Ethical responsibilities

The authors declare that no experiments on humans or animals have been performed for this research. They also state that no patient data appear in this article and that all patients signed the habeas data on admission to the hospital.

Conflict of interests

The authors declare that they have no conflict of interest.

REFERENCES

1. Jin P, Ji X, Zhi H, Song X, Du H, Zhang K, et al. A review of 42 cases of intestinal pseudo-obstruction in patients with systemic lupus erythematosus based on case reports. *Hum Immunol.* 2015;76:695–700, <http://dx.doi.org/10.1016/j.humimm.2015.09.022>.
2. Chng H, Tan B, Teh C, Lian T. Major gastrointestinal manifestations in lupus patients in Asia: lupus enteritis, intestinal pseudo-obstruction, and protein-losing gastroenteropathy. *Lupus.* 2010;19:1404–13, <http://dx.doi.org/10.1177/0961203310374337>.
3. Takeno M, Ishigatsubo Y. Intestinal manifestations in systemic lupus erythematosus. *Intern Med.* 2006;45:41–2, <http://dx.doi.org/10.2169/internalmedicine.45.0136>.
4. Tian XP. Gastrointestinal involvement in systemic lupus erythematosus: insight into pathogenesis, diagnosis and treatment. *World J Gastroenterol.* 2010;16:2971, <http://dx.doi.org/10.3748/wjg.v16.i24.2971>.
5. Park FD, Lee JK, Madduri GD, Ghosh P. Generalized megaviscera of lupus: refractory intestinal pseudo-obstruction, ureterohydronephrosis and megacholedochus. *World J Gastroenterol.* 2009;15:3555, <http://dx.doi.org/10.3748/wjg.15.3555>.
6. Chen YQ, Xue Q, Wang NS. Visceral muscle dysmotility syndrome in systemic lupus erythematosus: case report and review of the literature. *Rheumatol Int.* 2012;32:1701–3, <http://dx.doi.org/10.1007/s00296-011-1856-4>.
7. Li Z, Xu D, Wang Z, Wang Y, Zhang S, Li M, et al. Gastrointestinal system involvement in systemic lupus erythematosus. *Lupus.* 2017;26:1127–38, <http://dx.doi.org/10.1177/0961203317707825>.
8. Nguyen H, Khanna N. Intestinal pseudo-obstruction as a presenting manifestation of systemic lupus erythematosus: case report and review of the literature. *South Med J.* 2004;97:186–9, <http://dx.doi.org/10.1097/01.SMJ.0000087197.59817.AF>.
9. Wang J, Liu G, Liu T, Wei J. Intestinal pseudo-obstruction in systemic lupus erythematosus: a case report and review of the literature. *Medicine (Baltimore).* 2014;93:e248, <http://dx.doi.org/10.1097/MD.0000000000000248>.
10. Khairullah S, Jasmin R, Yahya F, Cheah T, Ng C, Sockalingam S. Chronic intestinal pseudo-obstruction: a rare first manifestation of systemic lupus erythematosus. *Lupus.* 2013;22:957–60, <http://dx.doi.org/10.1177/0961203313492873>.
11. Adler BL, Timlin H, Birnbaum J. Lupus intestinal pseudo-obstruction and hydronephrosis: case report. *Medicine (Baltimore).* 2019;98:e16178, <http://dx.doi.org/10.1097/MD.00000000000016178>.
12. Zhang J, Fang M, Wang Y, Mao J, Sun X. Intestinal pseudo-obstruction syndrome in systemic lupus erythematosus. *Lupus.* 2011;20:1324–8, <http://dx.doi.org/10.1177/0961203311405702>.
13. Yang C, Chen S, Gaut JP, Dehner LP. Smooth-muscle myopathy in systemic lupus erythematosus presenting with intestinal pseudo-obstruction. *Am J Gastroenterol.* 2016;111:1501–2, <http://dx.doi.org/10.1038/ajg.2016.328>.
14. Mok M, Wong RWS, Lau CS. Intestinal pseudo-obstruction in systemic lupus erythematosus: an uncommon but important clinical manifestation. *Lupus.* 2000;9:11–8, <http://dx.doi.org/10.1177/09612033000900104>.

1. Jin P, Ji X, Zhi H, Song X, Du H, Zhang K, et al. A review of 42 cases of intestinal pseudo-obstruction in patients with systemic lupus erythematosus based on case reports. *Hum*