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Case report

Successful treatment with intravenous immunoglobulins in a patient with intestinal pseudo-obstruction associated with systemic lupus erythematosus☆

Alex A. Echeverry[a], *Andrés A. Hormaza*[a], *Juan F. Betancur*[b], *Carlos A. Cañas*[a], *Iván Posso-Osorio*[a], *Gabriel J. Tobón*[a,c,*]

[a] *Unidad de Reumatología, Fundación Valle del Lili, Universidad Icesi, Cali, Colombia*
[b] *Departamento de Medicina Interna, Facultad de Medicina, Universidad CES, Medellín, Colombia*
[c] *Laboratorio de Inmunología, Fundación Valle del Lili, Cali, Colombia*

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ABSTRACT

Gastrointestinal involvement in patients with systemic lupus erythematosus (SLE) is very diverse, and the frequency of occurrence and location along the digestive tract varies widely. Inflammatory processes secondary to immune complex deposits or vascular events may cause this involvement. One of the most characteristic gastrointestinal manifestations in these patients is the intestinal pseudo-obstruction, which is defined as the ineffective intestinal propulsion that occurs in the absence of mechanical or obstructive factors. This is, however, a rare and poorly understood complication of SLE. The case is presented of a male SLE patient presenting with intestinal pseudo-obstruction, and was successfully treated with steroids and intravenous immunoglobulin. A complete review of the literature and a proposal for the pathophysiology of intestinal pseudo-obstruction are presented.

Tratamiento exitoso con inmunoglobulinas intravenosas en un paciente con pseudoobstrucción intestinal asociada a lupus eritematoso sistémico

Palabras clave:
Pseudoobstrucción intestinal
Lupus eritematoso sistémico

RESUMEN

El compromiso gastrointestinal en pacientes con lupus eritematoso sistémico (LES) es muy diverso. Su frecuencia y ubicación a lo largo del tracto digestivo varían ampliamente. Los procesos inflamatorios secundarios a los depósitos de complejos inmunes o eventos

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* *Corresponding author.*
E-mail address: gtobon1@yahoo.com (G.J. Tobón).

Hidronefrosis
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vasculares pueden ser los causantes de este compromiso. Una de las manifestaciones gastrointestinales características en los pacientes con LES es la pseudoobstrucción intestinal, que se define como la propulsión intestinal ineficaz que se produce en ausencia de factores mecánicos u obstructivos. Esta es, sin embargo, una complicación rara y poco entendida del LES. En este artículo, reportamos el caso de un paciente masculino con diagnóstico de LES y pseudoobstrucción intestinal, que fue tratado exitosamente con esteroides e inmunoglobulinas intravenosas. Se presenta una revisión completa de la literatura y una propuesta de la fisiopatología de la manifestación.

Introduction

Gastrointestinal involvement in patients with systemic lupus erythematosus (SLE) is very diverse. Its frequency and location along the digestive tract vary widely. Inflammatory processes secondary to deposits of immune complexes or vascular events may be the causes of the commitment.[1] Oral lesions, esophageal dysmotility, mesenteric vasculitis and protein-losing enteropathy are the most common manifestations.[2]

Intestinal pseudo-obstruction (IPO) is defined as the ineffective intestinal propulsion resulting from impaired functioning of the visceral smooth muscle, the enteric nerves or the visceral autonomic nervous system.[3] In many cases, the origin is primary, but secondary causes may occur, including certain neurological, endocrine and connective tissue diseases. The causes included in the latter group had been reported, mainly, in patients with systemic sclerosis.[4] In contrast, the association with SLE has been reported in few cases in the literature in English language around the world. This entity has been recently recognized as a rare and poorly understood complication of SLE.[1,5] In this article is presented the case of a male patient with SLE who had IPO and was successfully treated with steroids and intravenous immunoglobulin.

Case report

We report the case of a 28-year-old man with a history of SLE, diagnosed 3 months prior to admission because a systemic clinical picture characterized by acute abdomen secondary to appendicitis treated with an emergency laparotomy, with development of postoperative ascites and edematous syndrome with evidence of type IV lupus nephritis confirmed by renal biopsy, with arthritis and neurological involvement manifested as a convulsive episode without any other etiology. The immunological tests confirmed the diagnosis of SLE (positive antinuclear antibodies, positive anti-Sm and anti-double stranded DNA antibodies and hypocomplementemia). At this time the patient was treated extra-institutionally with 500 mg of intravenous cyclophosphamide every 2 weeks and, subsequently, with 2 g of mycophenolate mofetil every 24 h, 50 mg of losartan every 12 h, 30 mg of prednisolone every 24 h and 250 mg of chloroquine every 24 h. The patient was referred to our institution because of persistence of the abdominal pain in the lower hemiabdomen, diarrhea without dysentery and dysuria.

Upon admission to our institution, the patient was stable, with the following positive findings on physical examination: hypoventilation on the pulmonary auscultation of both bases and diffuse pain with abdominal palpation, with no evidence of signs of peritoneal irritation. The admission laboratory tests are shown in Table 1.

It was requested a renal ultrasound which showed diffuse bilateral enlargement, with bilateral pyelocaliceal dilatation and diffuse urothelial thickening. A magnetic resonance imaging of the brain was performed, which evidenced 2 hyperintense lesions located in the left frontal lobe in the FLAIR and T2 sequences, with diffusion restriction.

The transthoracic echocardiogram showed concentric hypertrophy of the left ventricle with an ejection fraction of 50–55% and a global pericardial effusion without hemodynamic repercussion. The colonoscopy evidenced mucosal edema in the descending colon with negative histopathological findings. The Systemic lupus erythematosus disease activity index (SLEDAI) at admission was high (value: 16). Due to the high activity of the SLE and the multisystemic involvement, the patient required management with pulses of methylprednisolone (1000 mg every 24 h) for 3 days, with initial clinical improvement. The evolution of the patient became torpid due to the presence of lower urinary tract symptoms, with a cystoscopy which evidenced trabeculation of the posterior wall of the bladder, with a bladder neck posterior lesion and a diverticulum in the left ureteral meatus.

Since the patient presented recurrent emesis, abdominal distention and, subsequently, constipation and severe abdominal pain, an endoscopy of the upper gastrointestinal tract was carried out, which showed gastric contents with retention of bile (800 cc). These findings were interpreted as adynamic ileus.

The abdominal CT scan with contrast (Fig. 1) showed hepatomegaly and splenomegaly, ascites, signs of intestinal vasculitis, mild bilateral hydronephrosis and an image suggestive of thrombosis of the right renal vein, which was verified by Doppler echography. Because of this, anticoagulation was started and it was administered a fourth dose of cyclophosphamide. Despite these treatments, the patient continued with poor clinical response, with increased abdominal pain, constipation and the need of high doses of opioids to