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Case reports

An unusual case of intestinal pseudo-obstruction presenting in an adolescent with juvenile-onset systemic lupus erythematosus: A diagnostic challenge

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ABSTRACT

Introduction: Systemic lupus erythematosus (SLE) is an autoimmune disease with multiple systemic manifestations. The disease itself typically causes only 2–30% of SLE-associated gastrointestinal conditions.

Case report: We present the case of a 16-year-old male with history of SLE diagnosed 5 months prior to admission. Patient was non-compliant to medical treatment. He presented with 20 days of cough, mucopurulent and blood-tinged sputum, progressive shortness-of-breath and abdominal bloating. Patient was found to have multiple organ dysfunction due to an active lupus flare that developed during hospitalization, and required treatment with high doses of corticosteroids and close observation in an intensive care unit. Despite initial improvement of symptoms, he continued with abdominal pain, bloating, abolished bowel sounds and poor food intake. An abdominal X-ray showed signs of intestinal obstruction, establishing the diagnosis of intestinal pseudo-obstruction (IpsO). Patient then displayed a marked improvement of his gastrointestinal condition following treatment with 400 mg/kg/day of intravenous immunoglobulin (IVIgG) for 5 days.

Conclusion: Intestinal pseudo-obstruction is an unusual clinical manifestation of SLE and may represent a diagnostic challenge. We underscore the importance of a prompt and precise recognition of this condition, which is likely to have a positive impact on clinical outcomes. IpsO is caused by a non-mechanical obstructive bowel injury. Evidence points towards the use of IgG and steroid for five days as the mainstay of therapy for patients with IpsO.

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1. Introduction

Systemic Lupus Erythematosus (SLE) is an inflammatory autoimmune disease with multiple systemic manifestations [1]. Gastrointestinal involvement has been similarly reported in adult and juvenile-onset SLE patients [2]. Among the associated gastrointestinal complaints, only 2–30% are typical of the disease, the remainders are caused by adverse reactions to medications. In general, 50% of patients with SLE suffer from anorexia, nausea, vomiting, diarrhea, esophageal dysmotility, protein-losing

enteropathy and mesenteric vasculitis as well as intestinal pseudo-obstruction (IpsO) in up to 23% [3] and ascitis in 35% [4]. The latter is known to be the rarest manifestation of SLE and is more frequently encountered among 30-year-old female patients [5–10]. This article describes a case of intestinal pseudo-obstruction in a young male suffering from a relapse of SLE disease activity.

2. Case report

We present the case of a 16-year-old male from a rural area in the south of Huila, Colombia, diagnosed with SLE five months prior to admission at a specialist outpatient clinic due to polyarthritis, malar rash and positive antinuclear antibodies (ANAs) and anti-double stranded deoxyribonucleic acid (ds-DNA) antibodies [11].

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The patient presented with 20 days of coughing with mucopurulent and blood tinged sputum. Patient was discharged with medication for symptomatic relief, however due to the persistence of his symptoms, functional respiratory decline and abdominal distension, the patient returned to the local hospital where bilateral pleural effusions were found on a chest X-ray. A bilateral tube thoracostomy was performed, removing 1800 mL of serohematic fluid. During the course of his hospitalization the patient experienced a severe anaemic syndrome, requiring a blood transfusion and referral to a higher-level facility. On admission he was afebrile with tachycardia, tachypnea, signs of dehydration and generalized mucocutaneous pallor. Besides heart rate, cardiac examination was unremarkable. Respiratory sounds were diminished. Chest tubes contained blood clots. Abdomen was soft, distended, with a positive fluid wave test indicating ascites. No guarding or rebound was present and no collateral circulation was found. The rest of the physical examination was within normal limits. The case report conforms to the 1995 Helsinki ethical declaration. Informed consent was obtained from the patient.

Laboratory test results (Table 1) were indicative of an active lupus flare and systemic management began with immunomodulators and steroids. In the following days the patient experienced diarrhoea and his temperature peaked at 102.2 °F. A lung infection was considered. The patient was transferred to the high dependency unit where an X-ray and a computed tomography (CT) scan of the abdomen showed signs of intestinal obstruction (Figs. 1 and 2). Despite initial improvement of symptoms, he continued with abdominal pain, bloating, abolished bowel sounds and poor food intake; general surgeons considered the patient to have an acute abdomen that required surgical management once lung function was stabilized.

On the patient's eighth day of hospitalization, pleurectomy/decortication surgery was intended for patient's right lung due to pleural effusion. During anaesthetic induction the patient suffered cardiac arrest, was resuscitated and the blood clots were removed from his thorax. The surgery was cancelled and the patient was transferred to the intensive care unit in isolation where, due to his symptoms, medical history and imaging studies, was considered

Table 1

Immunological profile and other laboratory test findings in a male patient with intestinal pseudo-obstruction and systemic lupus erythematosus.

Finding	Initial labs.		Hospitalization				
	30/03	02/04	Day 1	Day 2	Day 8	Day 19	Day 50
<i>Immunological profile</i>							
Anti-dsDNA	1:640		1: 40				
<i>homogeneous pattern</i>	1:640						
<i>ribosomal pattern</i>	1:160						
Anti-phospholipids							
IgM		24/10					
IgG		42/10					
C3 mg/dL			11.3		10	19.5	
C4 mg/dL			<5		6	4	
Anti-Sm				78.03			
Anti-RNP				68.44			
SS-A (anti-La)				1.67			
SS-B (anti-Ro)				6.75			
<i>Renal function</i>							
Creatinine (mg/dL)	0.8		2.17	3	2.93	1.39	0.96
BUN (mg/dL)			35.3	42.2	72.77	33.18	

Ds-DNA: anti-double stranded deoxyribonucleic acid; Ig: immunoglobulins; Anti-Sm: anti-Smith; C: complement; RNP: ribonucleoprotein; SS: Sjögren's syndrome, BUN: blood urea nitrogen.

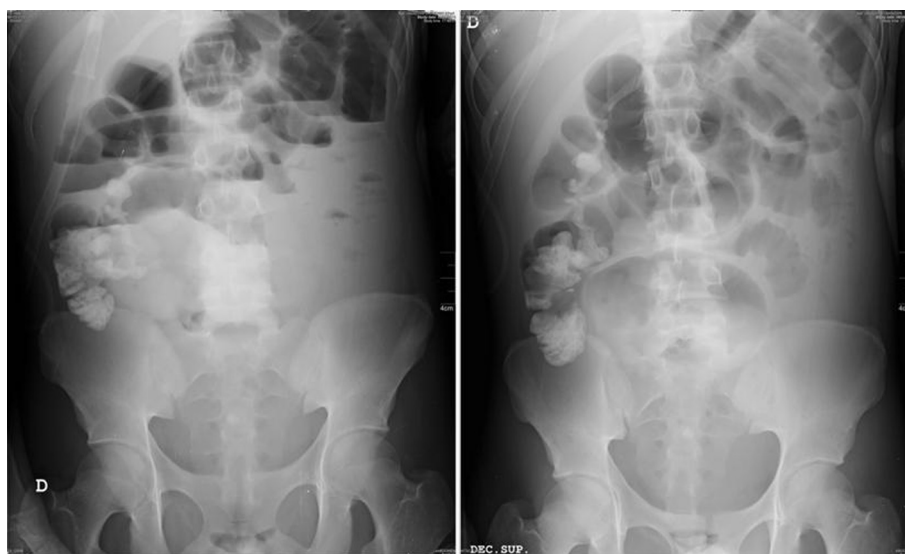


Fig. 1. Abdominal X-ray in a male patient with intestinal pseudo-obstruction and systemic lupus erythematosus. Gaseous distension of the bowel and hydro-aerial levels can be seen.

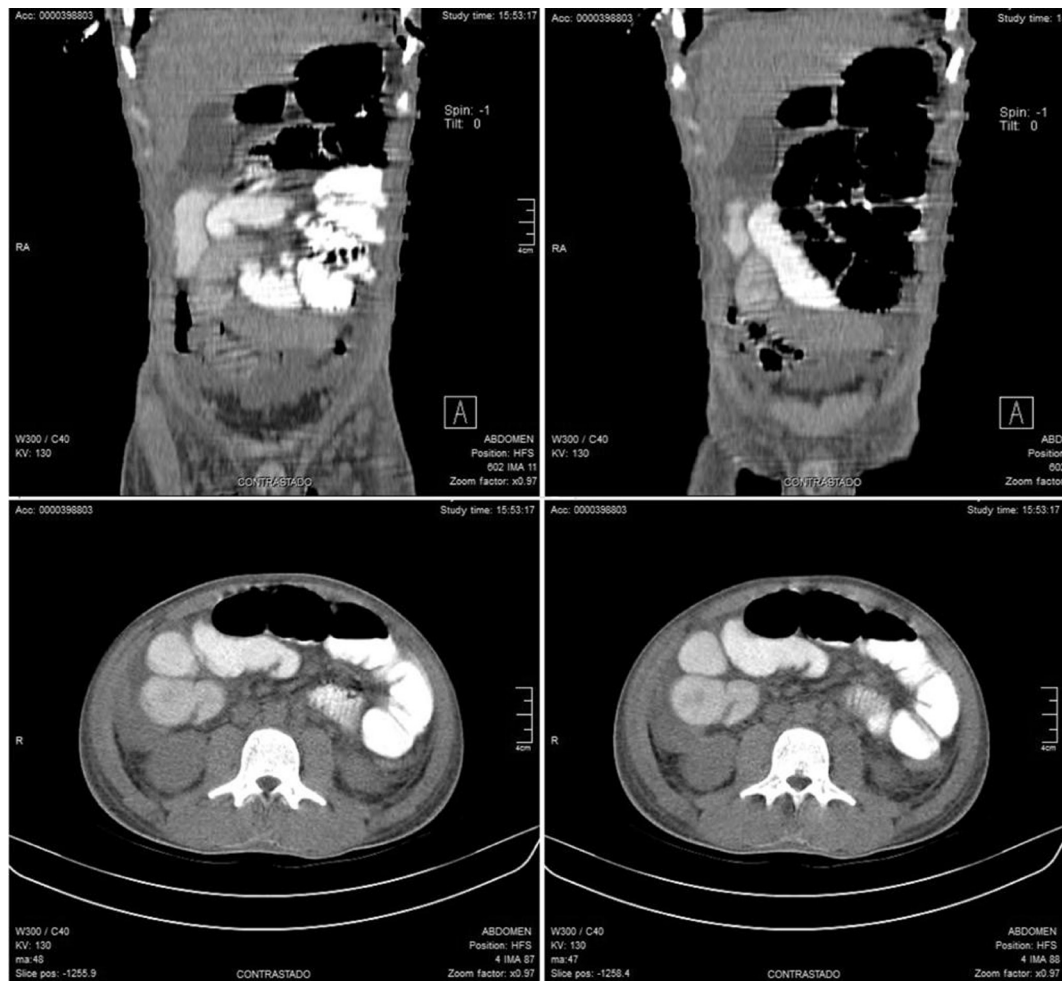


Fig. 2. Abdominal CT scan with contrast in a male patient with intestinal pseudo-obstruction and systemic lupus erythematosus. Free fluid can be seen in the peritoneal cavity. The stomach and intestinal loop are swollen and an abnormal gas and fluid distribution pattern can also be seen.

to be suffering from intestinal pseudo-obstruction due to SLE. The patient displayed marked improvement of his abdominal condition following treatment with 400 mg/kg/day of intravenous immunoglobulin (IVIgG) for 5 days and his SLE went into clinical remission as well.

3. Discussion

Intestinal pseudo-obstruction is a rare clinical syndrome of SLE and may represent a diagnostic challenge in most clinical settings. We underscore the importance of a prompt and precise recognition of this condition, which are likely to have a positive impact on morbidity and mortality outcomes [7]. IpsO presents typically as an initial manifestation of SLE or may arise as a complication of the disease or medications within the first few months after its diagnosis. It is characterised by being a non-mechanical obstructive lesion of the intestine, which is accompanied by diarrhoea, constipation, vomiting, abdominal pain, bloating, absence of bowel sounds and weight loss [7,9,12,13]. The underlying pathophysiology is not fully well understood, however, a few small cohort studies have suggested that the alteration is caused by mesenteric vasculitis which causes damage to the smooth muscle of the intestine, leading to hypomotility [7,9,14]. Other studies have proposed that, similar to glomerular compromise, intestinal damage may be caused by the deposition of immune complexes of anti-ds-DNA antibodies and C1q among others, which have receptors in both locations and may cause local inflammation, eventually leading

to formation of antibodies against the smooth muscle and tissue destruction. However, this remains a hypothesis and has not yet been verified [1,14,15].

Diagnosis is based on abnormal findings on abdominal x-rays and/or CT scans, such as: a grossly dilated bowel; abnormal bowel gas patterns; thickening of the intestinal walls, signs of peritonitis, pyelocaliectasis and ureterohydronephrosis (present in up to 60% of cases) are observed [9,16]. Systemic disease activity is usually present, so haematological and immune alterations are found in the majority of cases, remarkable mainly by leukopenia, decreased serum levels of complement, positive titers of anti-dsDNA, Sjögren's syndrome type A antibodies (anti-Ro) and anti-ribonucleoprotein (RNP) antibody and negative Sjögren's syndrome type B antibodies (anti-La) [17–19].

Treatment with prokinetic agents and high doses of steroids may suffice depending the degree of obstruction and the promptness of diagnosis [5,18,19]. In cases in which obstruction is more severe and/or the diagnosis is established in a delayed manner, surgical intervention may be necessary. Various studies suggest that the combined use of intravenous steroids along with a course of azathioprine, methotrexate, cyclophosphamide or even IVIgG for 5 days may lead to clinical remission and resolution of Intestinal pseudo-obstruction through means of immunosuppression and immunomodulation, reducing the need for surgical intervention and therefore reducing the risks of surgical complications [5,18,19].

The patient exhibited all of the typical symptoms associated with IpsO a few months after being diagnosed with SLE. On

laboratory tests, leukocytes were in normal ranges prior to admission, and serum complement levels were decreased at the time of discharge. Anti-dsDNA and anti-RNP autoantibodies were positive and anti-Ro and anti-La autoantibodies were negative. Abdominal imaging studies carried out on the patient revealed several abnormal findings that are characteristic of IpsO, such as abnormal gas distribution patterns (multiple hydro-aerial levels), the presence of liquid in the peritoneal cavity and a grossly-dilated bowel. Surgical management of the abdominal clinical picture was initially proposed, however, was not possible due to the associated respiratory condition. Combined use of high doses of corticosteroids and IVIgG was chosen as therapy for this case; cyclophosphamide was not used due to the high risk of infection and complications [3,5].

As a conclusion, from our review of medical literature and clinical experience, we recommend the combined use of steroids and IVIgG whenever IpsO is suspected and avoidance of gastrointestinal surgery is preferred. We also underscore the importance of maintaining a high clinical suspicion in order to achieve the prompt and precise diagnosis of this condition that is necessary to initiate a proper treatment [5,12,18,20].

Conflict of interest

None

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