

Images in Clinical Rheumatology

An atypical cause of intestinal occlusion and hidronephrosis

Íleo e hidronefrosis de causa atípica

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Introduction

We describe a patient who developed an intestinal ileus as the debut feature of systemic lupus erythematosus (SLE), with good response to corticosteroid treatment.

Case report

We present the case of a 56-year-old male with a history of psoriasis and occasional arthritis treated with NSAIDs and corticosteroids. Onset was days before admission, presenting poor general conditions, symmetrical arthritis (elbows, wrists and interphalangeal joints) and sustained fever of 38.5 °C accompanied by progressive dyspnea at rest, no cough, expectoration or chest pain. The exploration observed widespread psoriasis lesions, arthritis of the elbows and wrists, and hypoventilation of the base of the lungs, with rhythmic heart sounds and a pleural rub. Analytically, the patient presented hemoglobin of 11.3 g/dl, WBC $14.8 \times 10^3/\mu\text{l}$, neutrophils $13.3 \times 10^3/\mu\text{l}$, lymphocytes $1 \times 10^3/\mu\text{l}$, platelets $298 \times 10^3/\mu\text{l}$, prothrombin time 58%, activated partial thromboplastin time: 57.9 s, C-reactive protein > 9 mg/dl, ESR 83 mm/1st hour and cardiac enzymes within normal values. The electrocardiogram showed sinus rhythm at 100 beats per minute, with an axis at +30°, with no evidence of the repolarization abnormalities or signs of ischemia. The chest x-ray showed grade III/V cardiac enlargement and pleural effusion. The echocardiogram showed moderate pericardial effusion without signs of tamponade or segmental hypomotility. A chest scan was requested, which confirmed the existence of pleural and pericardial effusion (Figure 1). Suspecting sepsis of respiratory origin, empirical antibiotic therapy with ceftriaxone was begun. Blood urine and sputum

cultures for bacteria and Koch bacilli were serially taken and resulted repeatedly negative. The patient had an unfavorable clinical course, with persistent fever and intestinal ileus presented as abdominal distension, diffuse pain on palpation and fecaloid vomiting, with a progressive increase of acute phase reactants. Abdominal scan was performed (Figure 2) and significant expansion of the intestine, from sigma to the stomach, was observed without an evident cause of gastric obstruction. The right kidney showed dilated caliceal groups, with grade III hydronephrosis and dilation of the initial portion of the ureter. The colonoscopy was not objective because of stenosis.



Figure 1. Pleural and pericardial effusion.

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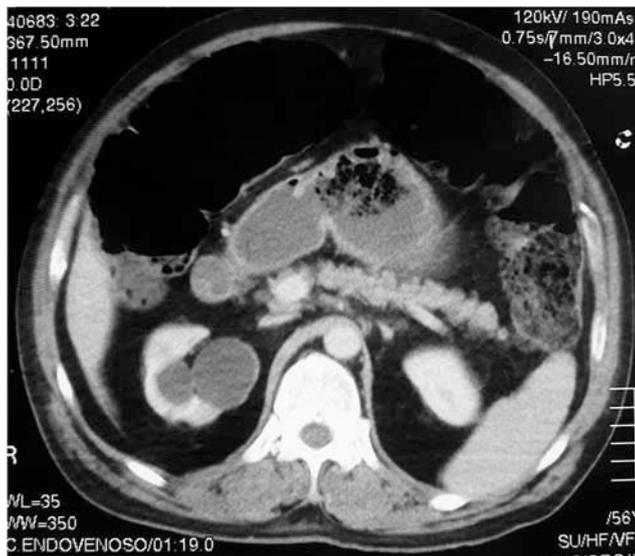


Figure 2. Ileum and hydronephrosis.

At the moment we received autoimmunity related laboratory results showing antinuclear antibody (ANA) 1/320, with lymphopenia, arthritis and polyserositis making the patient meets the criteria of the 1997 American College of Rheumatology (ACR) for SLE. We started the patient on high-dose steroids (methylprednisolone 1 g every 24 hours for 5 days). The patient improved clinically with defervescence, resolution of the ileus, polyserositis, arthritis, and laboratory measurements. In follow up visits we found the patient barely symptomatic, with episodes of occasional arthritis.

Discussion

SLE is a multisystem autoimmune disease of unknown etiology. Although gastrointestinal manifestations occur during the course of the disease in over 50% of patients, these are under evaluated.¹ The

digestive symptoms are varied and can affect any organ, although the majority of complications occur in the small and large intestine. The most common process at this level is vasculitis.¹ Pseudo-obstruction related to SLE has recently been described,² and there are fewer than 30 cases published.² Intestinal pseudo-obstruction can occur as an initial manifestation of SLE in around 40%² of cases, as our patient presented (with the consequent difficulty of diagnosis), or as an outbreak of disease after years of evolution.³ The clinical manifestations are vomiting, abdominal pain and diarrhea with bacterial overgrowth when the small bowel is involved or constipation if it acts on the large intestine. Intestinal pseudo obstruction is accompanied by ureterohydronephrosis in up to 63.6% of cases.⁴ The mechanism producing this is unknown. The association with ureterohydronephrosis may be explained by the presence of antibodies against smooth muscle¹ or possible vasculitic involvement producing hypomotility and muscle damage,² leading to atrophy and degeneration of the media with fibrinoid necrosis of vessel wall and infiltration of the lamina propria.⁵ Treatment consists of high doses of steroids and/or immunosuppressive drugs and prokinetics. It has a poor prognosis, with mortality around 18% at 6 months after onset of the pseudo-obstruction.² Importantly, a high degree of clinical suspicion is crucial to avoid unnecessary surgery.

Disclosures

The authors have no disclosures to make.

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