Ascites due to lupus peritonitis: a rare form of onset of systemic lupus erythematosus

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ABSTRACT

Serositis is commonly seen in systemic lupus erythematosus (SLE). Approximately 16% of patients with SLE have pleural or pericardial involvement. However, peritoneal involvement is extremely rare, and clinically seen in a small group of patients. This is the case report of a 47-year old female with discoid lupus who evolved with systemic manifestations of the disease, characterized by significant abdominal distension and pain, asthenia, weight loss, signs of ascites, and acute non-invasive diarrhea. Exhaustive diagnostic investigation was performed and included laboratory and imaging tests, colonoscopy, and analysis of the ascitic fluid. Besides ruling out the possibility of an infectious, neoplastic, and hemodynamic etiology, the investigation also allowed the confirmation of SLE. Thus, the hypothesis of lupus peritonitis with ascites became viable. The patient was treated with prednisone and chloroquine, with substantial improvement of her condition.

Keywords: systemic lupus erythematosus, serositis, ascites, colitis.

INTRODUCTION

Systemic lupus erythematosus (SLE) is a chronic multisystem autoimmune disease, whose most striking characteristic, from the clinical and pathologic point of view, is the development of inflammatory reactions in several tissues and organs.[1,2]

The disease alternates periods of exacerbation of the inflammatory process with periods of relative improvement, or even inactivity, and affects predominantly skin, joints, serosa, glomeruli, and central nervous system. Serositis is a common finding among the wide range of manifestations of SLE patients. Approximately 16% of SLE patients have pleuritis and/or pericarditis, but the effusion rarely causes ventilatory or circulatory repercussion. However, peritoneal serositis with ascites (known as lupus peritonitis) is an extremely rare manifestation.[3]

This is the report of a patient initially diagnosed with discoid lupus who evolved with systemic manifestations, such as chronic peritoneal serositis and colitis.

CASE REPORT

The patient is a 47-year-old Caucasian female, complaining of progressive increase of her abdominal volume associated with asthenia and diffuse heavy abdominal pain for two months, which got significantly worse in the last week. She denied any pattern of pain irradiation, association with food, dysuria, or fever. She also reported weight loss of approximately 7 kg in the last three months, and small amounts of watery diarrhea free from food residues, mucus, pus, or blood in the past week. When specifically questioned, the patient reported being diagnosed with discoid lupus one year before, and treated with chloroquine. She denied any recent exacerbation of the underlying disease. She also denied alcoholism and illicit drug use, but reported smoking half a pack of cigarettes per day for 30 years.

On physical exam, her general state of health was regular and she was pale, acyanotic, anicteric, febrile (37.5 °C), eupneic, and thin. Cardiovascular and respiratory examinations

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were within the normal range. Her abdomen was distended, and diffusely tender, with neither palpable masses nor collateral circulation. Signs of ascites were present. Liver and spleen were non-palpable. Digital rectal examination did not reveal feces, blood, or mass in the rectal ampulla. Mild, cold, painless, pitting edema was observed in both lower limbs. The osteoarticular system showed no changes. Lace-like, purplish discolorations of the skin of the trunk and dorsum, compatible with livedo reticularis, were observed. There were also malar erythema and diffuse discoid lesions in her trunk and limbs.

The patient had no previous laboratory tests. On admission, her initial laboratory tests were as follows: fine speckled antinuclear antibodies (ANA), 1:160; reactive anti-Ro; and normal cryoglobulins and serum complement. The patient was non-reactive for the following antibodies: anti-La; anti-DNA; anti-cardiolipin; lupus anticoagulant; anti-SM; anti-RNP; anti-SCL-70; and anticentromere. Serologies for HIV, viral hepatitis, and VDRL were negative, and ALT, ASG, GGT, and LDH levels were normal. CBC showed normocytic normochromic anemia with negative direct Coombs test, and her leukogram and platelets were normal. BUN was normal with slight creatinine elevation (1.86 mg/dL); 24-h proteinuria: 800 mg/24 h; and serum albumin: 3.68–2.54–2.20 g/dL. Fresh stool test and stool culture showed no abnormalities. Urinalysis evidenced 1:25,000 erythrocytes, and proteinuria +++.

Puncture of the ascitic fluid showed the following: total leukocyte count of 135 (differential: lymphocytes, 94%; neutrophils, 3%; and eosinophils, 3%); frequent mesothelial cells; LDH, 136 U/L; and amylase, 9 U/L. The following tests were negative: LE cells; bacterioscopy; Koch bacillus; and oncotic cytology. Serum-ascites albumin gradient (SAAG) of 0.85.

Plain chest X-ray showed no changes. Abdominal ultrasound revealed only moderate ascites with no other alterations, and transvaginal ultrasound evidenced neither uterine, nor adnexal changes. Abdominal CT confirmed the ultrasound findings and showed discrete thickening of the colonic wall.

Esophagogastroduodenoscopy showed moderate enanthematous pangastritis and mild bulbar duodenitis, negative for the urease test. Colonoscopy showed enanthematous lesions in the terminal ileum and ascending colon, and no bleeding. During this procedure biopsy was performed, and the histopathology showed moderate non-specific chronic ileitis with areas of erosion and moderate active chronic colitis with ulceration and sparse foci of acute cryptitis.

Systemic lupus erythematosus was suspected based on the American College of Rheumatology criteria:[2] malar rash; discoid lupus; photosensitivity; persistent proteinuria > 500 mg/dL; serositis; and ANA. Prednisone (60 mg/day) was introduced, and chloroquine (250 mg/day) was maintained. The patient showed improvement of her general state of health, a reduction in abdominal circumference, and weight gain, and initiated outpatient clinic follow-up.

Three months after beginning treatment, the patient has gained 4 kg. She is currently followed up at the outpatient clinic, remains asymptomatic, with no ascites, and has regular intestinal rhythm (once or twice a day) with soft stools and no bleeding.

DISCUSSION

Systemic lupus erythematosus is an autoimmune disorder with clinical manifestations that can affect any organ or system.[1] Inflammation of the pleural and pericardial serous membranes, although non-specific, is relatively common, constituting one of the 11 American College of Rheumatology criteria for the classification of SLE.[2] Inflammation of serous membranes, including pericardium, pleura, and peritoneum, can cause pain, fluid accumulation, adherence, and even fibrosis.[3] In a recent prospective study with 1,000 European patients with SLE followed-up for ten years, the frequency of serositis was 16%.[3] Peritoneal serositis with ascites (known as lupus peritonitis) is extremely rare.[4]

The gastrointestinal tract can be involved in SLE.[5,6] However, gastrointestinal manifestations are, in most cases, caused by infections or adverse effects of medications. Symptoms related to the disease itself are not as common as the involvement of other organs, such as in lupus nephritis. On the other hand, the incidence of gastrointestinal manifestations can be clinically underestimated, since some are non-specific and abdominal symptoms can be absent.[5,6]

Acute diarrhea is a common clinical manifestation of several diseases. Its causes can be grouped into two major categories: infectious and non-infectious. More than 90% of the cases of acute diarrhea are caused by infectious agents and are frequently accompanied by vomiting, fever, and abdominal pain. In the present case, initially the diarrhea associated with ascites left no doubt regarding a possible acute, non-invasive diarrhea syndrome of infectious etiology. However, lack of improvement after 48 hours and SLE activity motivated closer investigation. The absence of changes in laboratory tests suggested the diagnosis of diarrhea as a manifestation of SLE. When gastrointestinal involvement is associated with SLE activity, the causes to be considered include mesenteric vasculitis and protein-losing enteropathy.[5,6] The hypoproteinemia of the patient could be justified by both proteinuria and protein-losing enteropathy. In addition, the intestinal involvement might have

been secondary to mesenteric vasculitis, since it responded promptly to treatment of the underlying condition.

In a post-mortem study, 60%–70% of the patients with SLE had peritonitis, while only approximately 10% of them had been clinically diagnosed.[5] In the case reported here, the patient had moderate ascites of insidious-onset as her initial SLE manifestation. In clinical practice, when facing a patient with visible ascites, correctly diagnosing the cause of the ascites is the first essential step for a successful treatment. Ascites can be acute or chronic, with or without pain, and two factors could be implicated: portal hypertension or peritoneal diseases. These factors trigger a wider cyclic process that justifies and perpetuates the constant accumulation of abdominal fluid. This process is due to the activation of the renin-angiotensin-aldosterone system, sodium and water retention, increased plasma volume, and extravasation into the ascitic fluid.[7]

Specific diagnostic approach is initiated by the distinction between those two triggering factors by use of diagnostic paracentesis. Thus, in portal hypertension, ascites is formed by a mechanism involving only the increase in vascular hydrostatic pressure, classically resulting in a transudate. However, in the damage of the peritoneal tissue (inflammatory or neoplastic), vessel permeability is altered, originating a protein-rich exudative fluid.[7]

Still little understood, ascites probably involves a multifactorial mechanism. Two triggering factors have been suggested: in one, self-reactive B lymphocytes produce autoantibodies that bind circulating antigens, forming immune complexes that deposit on the peritoneum, triggering a local inflammatory reaction; in the other, vasculitis of peritoneal vessels or the serous membrane of abdominal organs is observed. Regardless of the triggering factor, ascites in SLE is a peritoneal condition, classically resulting in an exudate.

Lupus peritonitis should be considered an exclusion diagnosis, demanding an extensive investigation for the most common causes of exudative ascites, such as peritoneal carcinomatosis, primary mesothelioma, peritoneal pseudomyxoma, hepatocellular carcinoma, peritoneal tuberculosis, fungal peritonitis, HIV-associated peritonitis, nephrotic syndrome, protein-losing enteropathy, and severe malnutrition.[7]

The prognosis of SLE peritonitis is usually good, if specific treatment is promptly instituted.[3,4,8] Since it is a disease with an inflammatory etiopathogenic substrate, current therapeutic schedules are based on non-steroidal anti-inflammatory drugs and corticosteroids.[3,8,9] Although lupus serositis usually responds to non-steroidal anti-inflammatory drugs or corticosteroids, refractory cases that lead to persistent fluid accumulation in the serosal cavity have been reported.[3] In these cases, immunomodulators or immunosuppressors, as well as surgical procedures, such as pleurodesis and pericardial fenestration, could be necessary.[3,9]

CONCLUSIONS

Serositis is a common characteristic of SLE, but lupus peritonitis is rare, which is most likely due to its clinical underestimation. Even though, lupus peritonitis should be considered an exclusion diagnosis, requiring extensive clinical evaluation in search for alternative causes of exudative ascites. Prognosis is usually good, and treatment is based on the use of non-steroidal anti-inflammatory drugs and corticosteroids, with good response. For refractory cases, individualized alternative measures are indicated.

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