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.
Systemic lupus erythematosus is an autoimmune disorder with clinical manifestations that can affect any organ or system. 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. Inflammation of serous membranes, including pericardium, pleura, and peritoneum, can cause pain, fluid accumulation, adherence, and even fibrosis. In a recent prospective study with 1,000 European patients with SLE followed-up for ten years, the frequency of serositis was 16%. Peritoneal serositis with ascites (known as lupus peritonitis) is extremely rare.

The gastrointestinal tract can be involved in SLE. 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.

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.

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. 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.3,5,9 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.
REFERENCES

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