SEVERE MESENTERIC VASCULITIS IN A PATIENT WITH SYSTEMIC LUPUS ERYTHEMATOSUS AND LUPUS NEPHRITIS, HOSPITAL DE BASE, SÃO JOSÉ DO RIO PRETO, SÃO PAULO STATE, BRAZIL

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ABSTRACT

Systemic lupus erythematosus (SLE) is a complex autoimmune inflammation that can affect several organs and the mesenteric vasculitis is uncommon among gastrointestinal complications. In this work, we present a case of a 61-year-old male patient with Lupus mesenteric vasculitis (LMV) and demonstrate the lack of data in Brazil.

KEYWORDS: Lupus mesenteric, transmural hemorrhage, autoimmune disease, vasculopathy.

1. INTRODUCTION

Systemic lupus erythematosus (SLE) is an autoimmune chronic disease with an extremely complex pathogenesis and the genetic predisposition can be induced by multiple stress factors involving epigenetic mechanisms and are under the influence of the innate immune system[1]. The primary pathological findings in patients with SLE are those related to inflammation, such as vasculitis, immune complex deposition, and vasculopathy[2], affecting several organs such as kidneys, joints and gastrointestinal system[3]. In fact, gastrointestinal manifestations are common in patients with SLE, and the symptoms may range from oral ulcers, dysphasia, nausea and vomiting, to more severe cases, such as hepatitis, pancreatitis, and lupus mesenteric vasculitis (LMV)[4]. However, mesenteric vasculitis is uncommon among gastrointestinal complications (2.2-9.7%)[5]. In a Chinese retrospective cohort study with 3,823 systemic lupus erythematosus patients, the LMV prevalence was 2.5%, including 13 who died due of serious complications[6]. Surgical treatment is indicated in cases associated with advanced ischemia, peritonitis, sepsis or pneumatosis[7]. The aim of this study was to describe a case of severe mesenteric vasculitis in an SLE patient attending the Hospital de Base, São José do Rio Preto, Brazil, and demonstrate the lack of data in Brazil.

2. CASE REPORT

A 61-year-old male patient, attending an outpatient clinic due to SLE with a type IV, dialytic, nephropathy, taking azathioprine and prednisone, smoker, seeks urgent care with complaint of abdominal pain in mesogastrium, associated with nocturnal vomiting and intestinal constipation for two days before; anuria for three months before. Body temperature 35.6ºC, Blood Pressure (BP) 160x100 mm/Hg, Heart Rate (HR) 100 bpm, ECG 15, diffuse pain to abdominal palpation with signs of peritoneal irritation throughout the abdomen. Absence of stool in the rectum. Hemoglobin 12.7 g/dl; Hematocrit 40.1%, Leukocytes 10,950/mm^3, Segmented 8360/mm^3, Typical lymphocytes 1,780/mm^3, Monocytes 770/mm^3, Platelets 500,000/m^3, Coagulogram: uncoagulable, Protein C reactive 11.3 mg/dl, Creatinine 10 mg/dl , Urea 69 mg/dl, Arterial gas: pH 7.41, PO_2 118.3 mmHg, PCO_2 18.4 MMOL/L, BIC 11.4 MMOL/L, Lactate 1.0 MMOL/L. Reactive antibodies anti-FAN and anti-nucleus and non-reactive anti-nucleolar, anti-cytoplasm, anti-mitotic plate and anti-metaphase apparatus. Normal serum supplement (C4: 14 mg/dl, C3: 70 mg/dl). X-ray in acute abdomen did not evidenced pathological alterations. The patient was submitted to exploratory laparotomy on the same day, evidencing the presence of a moderate amount of blood in the abdominal cavity, with an area of dark purple-red small intestine without vitality initiating from 70 cm of the angle of Treitz and extending for 120 cm. A segmental enterectomy of approximately 50 cm was performed with primary anastomosis in two planes. The biopsy revealed multifocal ischemic necrosis associated with edema and transmural hemorrhage in the intestinal segment. The patient died three days after admission due to a cardiogenic shock and exacerbation of Chronic Renal Insufficiency.

3. DISCUSSION

This case reported an adult individual with a confirmed diagnosis of SLE, who sought the hospital services due to symptoms of abdominal pain associated with vomiting, but the severity of the condition due to LMV, prevented his survival. Mesenteric vasculitis is an extremely serious complication and is highly lethal (50%) during the course of SLE[6]. Probably due the late admission, with SLE and mesenteric ischemia as consequence of vasculitis and necrosis, early diagnosis and intervention could have contributed to the longer survival. Although the severe and potentially fatal complication of SLE, which causes acute abdominal pain, the diagnosis and management remain a great challenge[8]. Contrary to the findings in the present study,[9] Anand *et al*., 2016, when administered high doses of steroids and cyclophosphamide were able to reverse mesenteric vasculitis in a patient with a class IV lupus nephritis. The vasculitis in patients with SLE may present different clinical forms based on the organ involved and the size of the vessel affected[10].

A lack of studies was observed in the Brazilian scientific literature[7,4,11,12] regarding the occurrence of LMV in individuals with SLE, according to a survey conducted in the PubMed, Scielo and Lilacs databases. Christmann *et al.,* 2003[11], describe a 28-year-old female patient diagnosed with SLE, who suddenly presented severe abdominal pain with absence of other manifestations, while Carvalho, 2010[4], described a case of a 45-year-old female patient, with symptoms of fatigue, fever, alopecia, facial edema, Raynaud's phenomenon, malar rash, polyarthritis in the great and small joints, leukopenia and lymphopenia. Albuquerque-Netto *et al*., 2013[7], in a 28-year observation series verified the occurrence of LMV and SLE in 0.4% of the patients. In a male teenager with diffuse and acute abdominal pain, nausea, bilious vomiting, abdominal distension, rebound tenderness, and abdominal muscle guarding, intestinal resection was performed, as well as in the study conducted by Palma *et al*, 1996[12], with a 24 year-old woman. Both of them had an immunosuppressive therapy and satisfactory evolution.

The inflammatory aspect of the disease, as evidenced in this patient, with high white blood cell count, antinuclear antibodies, among other factors, are in agreement with the findings of Chen *et al*, 2009[13] and Mok and Lau, 2003[2], in which antinuclear antibodies are most characteristic and present in more than 95% of patients. However, B cell hyperactivity, which leads to the generation of a multitude of different autoantibodies that are directed not only against nuclear antigens[1]. Considering that approximately 11 to 33% of SLE patients may be affected by disorders due to vasculitis[10], in which they usually present acute abdominal pain with sudden onset, severe intensity and diffuse localization[6], and that mesenteric vasculitis is an extremely serious, highly lethal complication during the course of SLE. Mesenteric vasculitis is a serious complication of systemic lupus erythematosus (SLE), since it is considered an important cause of morbidity and mortality in this disease. It is also a diagnostic and therapeutic concern, and its early diagnosis and intervention affect directly the prognosis. Thus, we emphasize the need to establish an early diagnosis in order to avoid complications and a worse evolution[11].

4. CONCLUSION

Considering that lupus is a disease with several clinical manifestations that make diagnosis difficult, and this patient had a late admission to the health care and uncommon involvement, as mesenteric vasculitis and necrosis, an early diagnosis and intervention could have promote a longer survival for the patient.

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Conflicts of interest
The authors declare no conflicts of interest.