CASE REPORT

# MESENTERIC VASCULITIS IN LUPUS

## • *case report and literature review* •

***Orivaldo Alves Barbosa*, Jose Gerardo Araujo Paiva**, Max Victor Carioca Freitas**, Romano Bezerra Brasileiro***, Leilane da Conceição Almeida Guimarães****, José Walter Correia********

* Professor of Medicine Unichristus Internal Medicine Service ,Hospital Cesar Cals – orivaldo.alves.barbosa@gmail.com

** Rheumatology Service - Hospital Cesar Cals

*** Resident of Rheumatology Service - Hospital Cesar Cals

**** Academic of Medicine

***** Doctor in Medicine, Internal Medicine Service ,Hospital Cesar Cals

## Abstract

Systemic lupus erythematosus often presents with involvement of the gastrointestinal tract of several causes. The lupus mesenteric vasculitis, a well-described entity, is rare, affecting between 1-2% of patients with lupus and abdominal pain, leading to severe and potentially fatal disease. We report a case of a young woman, without prior diagnosis, presenting with severe abdominal pain and fever with final diagnosis of lupus mesenteric vasculitis successfully treated with immunosuppression .We present a brief review of literature.

*Keywords:* Lúpus enteritidis; Mesenteric vasculitis; Abdominal pain in lupus

## INTRODUCTION

Systemic lupus erythematosus (SLE), a disease characterized by the production of autoantibodies, complement activation and multisystem damage often presents itself with involvement of the gastrointestinal tract of several causes, ranging from adverse drug effects and infections related to immunosuppression, to pancreatic diseases, lupoid hepatitis, peritoneal serositis associated disease (celiac disease, inflammatory bowel disease) and ischemia.[1-7]

The lupus mesenteric vasculitis (VML) is infrequent. It affects between 1-2% of patients with lupus and abdominal pain, leading to severe and potentially fatal disease if not recognized and promptly treated with immunosuppression.[2]

## CASE REPORT

FV woman, 22 years old, previously healthy, approximately one month prior to admission started an intense diffuse abdominal pain, accompanied by nausea, vomiting, diarrhea (three daily episodes, worsening with feeding;

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unyielding with fasting without food debris, mucus or blood) and high fever, which had intensified in the last five days with progressive abdominal distension. Sought medical assistance at another service, was hospitalized with acute surgical abdomen hypothesis, was referred to a rheumatology service of Hospital Cesar Cals.

On admission, the patient presented impaired general health status with respiratory distress and had an erythematous rash on the cheek. The patient presented distended abdomen, with reduced noise, this mobile dullness and extremely painful to superficial palpation with signs of peritoneal irritation.

In the initial tests, hypoalbuminemia was evidenced with proteinuria (1g in 24 hours) and chest radiography with bilateral pleural effusion and ultrasound with mild ascites.

With the possibility of SLE, intravenous methylprednisolone at a dose of 50 mg/day and bowel rest were initiated, with significant improvement in pain symptoms in the early days. In the fourth days of hospitalization, the patient developed worsening respiratory distress and severe abdominal pain. Thoracentesis has been carried out for relief and diagnostic. An abdominal CT with contrast was performed (Fig. 1), showing dilatation and diffuse thickening of the bowel (duodenum, jejunum and ileum), wall marked edema (signal "target"), without signs of perforation,compatible with lupus mesenteric vasculitis. Pulse therapy with methylprednisolone 500mg for three days was indicated. The patient evolved with a good clinical response, resuming oral diet. As the abdominal CT, seven days after pulse therapy, showed significant radiological improvement (Fig. 2), we started pulse cyclophosphamide 1g, monthly, without recurrence. We have programmed with six cycles of cyclophosphamide and corticosteroid weaning at a household level.

**Figure 1 –** Abdominal contrast-enhanced CT - marked wall thickening of the duodenum and jejunum generating the classic target sign, caused by the edema of the small vessel

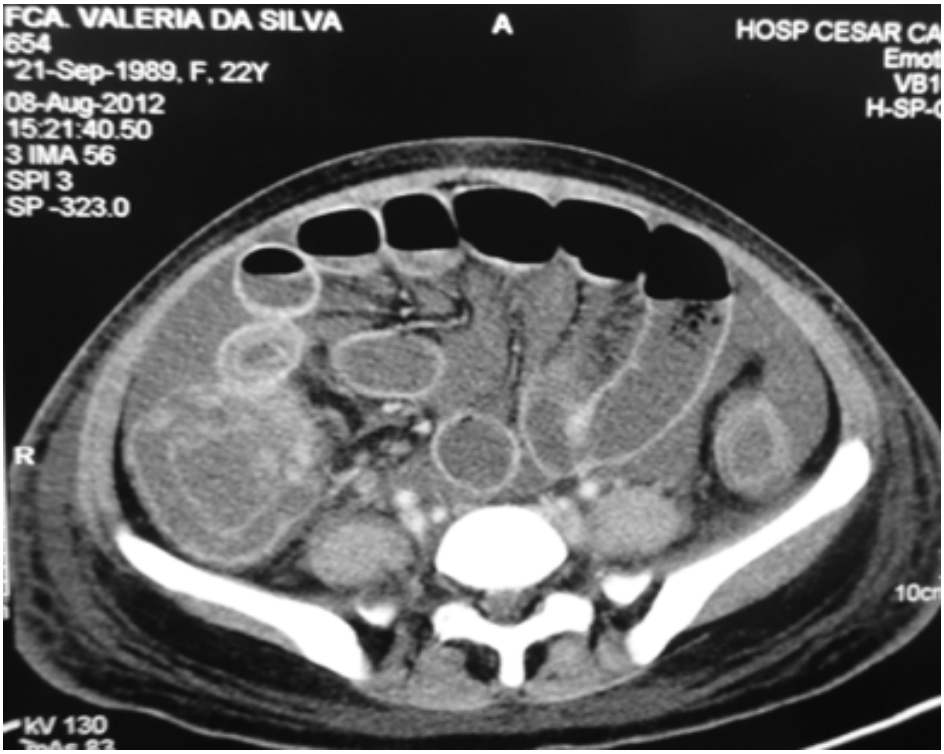
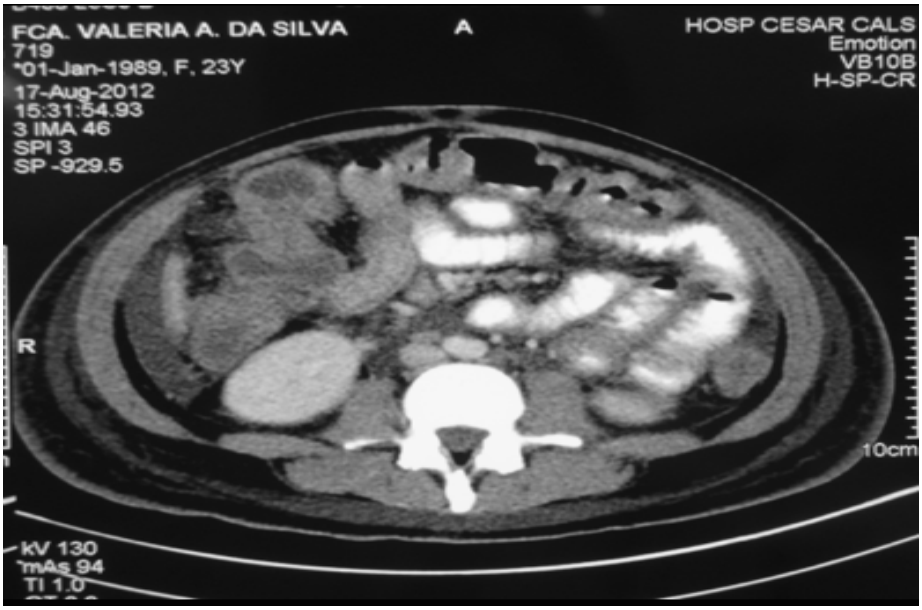


**Figure 2 –** Abdominal TC with contrast 7 days after treatment - significant improvement both in duodenum and small bowel edema.




## DISCUSSION

We describe the case of a patient without a previous diagnosis, starting lupus frame with hypocomplementemic refractory mesenteric vasculitis to initial therapy with corticosteroids.

Vascular involvement in lupus can be classified into two types: inflammatory (capillaritis, vasculitis and leucoaggregation) and thrombotic (venous or arterial, often associated with antiphospholipid antibody syndrome)[1-3]. In our patient, both abdominal image and the complement consumption and a lack of compatible APS antibodies (Table 1) facilitated the diagnosis of inflammation.

**Table 1 –** Laboratory Data

(continued)

| Exame | Reference | Results |
|---|---|---|
| Haemoglobin | 11.3 a 16.3 g/dl | 11 g/dl |
| Leukocytes | 4000 a 10000 mm3 | 4120 mm 3 |
| Lymphocytes | 1500 a 4000 mm3 | 769 mm3 |
| Platelets | 150.000 a 450000 mm3 | 450.000 mm3 |
| C -reactive protein | Under 5,6 mg/l | 7,8 mg/l |
| Lipase | 10-140 ui/l | 126 u/l |
| Albumin | Above 3,5 g/dl | 2,6 g /dl |
| Sodium | 135 -145 meq/l | 138 meq/l |
| Potassium | 3,5 -5,5 meq/l | 3,0 meq /l |
| Creatinine | 0,5-1,3 mg/dl | 0,6 mg /dl |

**Table 1 –** Laboratory Data

(conclusion)

| Exame | Reference | Results |
|---|---|---|
| Amilase | 80-180 ui/dl | 202 ui/l |
| Complement (C3) | 84-160 mg/dl | 34,9 mg/dl |
| Complement (C4) | 12 -36 mg/dl | 12 mg/dl |
| Complement (CH50) | Maior que 60 ui/CAE | 12 ui/CAE |
| Antinuclear Antibody | Não Reagente | 1:2560 Speckled |
| Anti-Smith antibody | under 15 U/mL | 23 U/mL |
| Anti-DNA Antibody | Negative | Positive 1:320 |
| AntiCardiolipin IgM | Under 7,0 MPL/ml | 3,0 Mpl/ml |
| AntiCardiolipin IgG | Under 10 GPL/ml | 2,6 GPL/ml |

The VML, also called as lupus arthritis, lupus enteritis or acute gastrointestinal syndrome, may be considered an infrequent complication of the underlying disease, which affects between 0 to 9.7% of patients with lupus symptoms and TGI [1-8] (Table 2). Considering only patients with diffuse severe abdominal pain, VML incidence ranges between 29-65%.

**Table 2 -** Sumary of studies with prevalence of mesenteric vasculitis in lupic patients.

| Study | Country | Number of Patients with Enteritis/ number of patients |
|---|---|---|
| S Shapeero et al (1974) | USA | 9/141 (6,4%) |
| Zizic et al (1982)[1] | USA | 9/140 (6,4%) |
| Sultan et al (1999)[8] | England | 1/255 (0,4%) |
| Lee et al (2002)[4] | South Korea | 17/175 (9,7%) |
| Lian et al (2003)[2] | Singapore | 33/1500 (2,2) |
| Ramos-Casals et al (2006)[3] | Spain | 0/670 (0%) |
| Kwok et al (2007)[4] | South Korea | 41/706 (5,8%) |

Physiopathologically, vascular inflammation occurs in small vessels mediated by immune complexes and complement deposition, with possible thrombosis (both associated with antiphospholipid syndrome, such as through mutual worsening of vasculitis and thrombosis). Clinically, patients present more commonly with diffuse abdominal pain, nausea and vomiting, anorexia and diarrhea, ranging from mild cases to gastrointestinal bleeding and possibly intestinal necrosis with perforation.

In suspected cases, the diagnosis should be made as soon as possible, with early institution of therapy, seeking improvement in survival. In this

context, the most appropriate test for the evaluation is computed tomography with intravenous contrast. The image findings are characterized by a classic diffuse edema with thickening of the bowel, causing a typical contrast uptake described as "target lesion". You can also view diffuse gaseous distension, ascites, and engorgement of mesenteric vessels [9].

Treatment is a rapid implementation of immunosuppression with high doses of methylprednisolone and bowel rest, obtaining clinical response in most cases. In refractory and recurrent cases, should be used alternative immunosuppression, especially with cyclophosphamide as pulse therapy. Cases without clinical improvement should be evaluated for intestinal perforation[11].

The prognosis varies according to the quick implementation of treatment. However, some authors reported mortality of up to 50%[12].

## CONCLUSION

Due to its high morbidity and mortality, lupus mesenteric vasculitis should always be considered in the differential diagnosis of acute abdominal pain, especially in young women. Computed tomography helps in diagnosis and the "target sign", found frequently, has good specificity for the diagnosis.

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