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ANCA POSITIVE LEUKOCYTOCLASTIC VASCULITIS: A CASE REPORT

Rheumatology

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ABSTRACT

Leukocytoclastic vasculitis or Hypersensitivity vasculitis, is characterized by inflammation of small vessels that mainly affects the venules, mediated by the deposition of immune complexes. Its aetiology varies from idiopathic to secondary to medications, infections, collagenosis or neoplasms. The diagnosis is confirmed by skin biopsy, which is characterized by an angiocentric inflammatory process associated with leukocytoclasia. Brazilian authors describe a case of cutaneous leukocytoclastic vasculitis with positive C-ANCA, difficult to diagnose and successful in clinical management.

KEYWORDS

vasculitis, leukocytoclastic, cutaneous, systemic, biopsy

INTRODUCTION

Vasculitis is a heterogeneous disease, characterized by an inflammatory process that attacks the wall of blood vessels, which can lead to stenosis, occlusion, formation of aneurysms or haemorrhages. Affects large, medium and/or small vessels. Its severity varies according to the clinical repercussions, being mild and localized or leading to multiple organ failure. [1, 2]

Leukocytoclastic vasculitis, or Hypersensitivity vasculitis, is characterized by inflammation of small vessels that mainly affects the venules, mediated by the deposition of immune complexes. Although cutaneous involvement is the most common clinical presentation, systemic involvement can occur in up to 57% of cases, such as: renal, joint, pulmonary, muscular, cardiac, gastrointestinal and/or peripheral nerves. [1]

Regarding its aetiology, it varies from idiopathic, due the use of medications, infections, collagenosis or neoplasms. Its clinical presentation begins with palpable purpura, which can ulcerate as the condition progresses. The diagnosis of leukocytoclastic vasculitis is confirmed by skin biopsy, which is characterized by an angiocentric inflammatory process associated with leukocytoclasia, characterized by fragmentation of neutrophil nuclei, oedema of endothelial cells, extravasation of red blood cells and fibrinoid necrosis. [2, 3]

Brazilian authors describe a case of cutaneous leukocytoclastic vasculitis with positive C-ANCA. As she was a patient with polycomorbidities, including Chronic Obstructive Pulmonary Disease (COPD) and extensive panlobular pulmonary emphysema, she did not have a clinical status for invasive interventions.

CASE REPORT

Female patient, 69 years old, active smoker with a history of 54 pack-years, with COPD, Coronary Artery Disease (CAD) and Chronic Venous Insufficiency, using Diosmin and Hesperidin.

She presented bullous, painful skin lesions, with the presence of ulcers and crusts with a foul odor on the lower limbs (Figure 1) that had been developing for 3 years. She was admitted to the emergency room at Hospital São José de Jaraguá do Sul (HSJ) due to the significant increase in these injuries and symptoms characteristic of COPD exacerbation.

Upon admission, additional laboratory tests were performed. She presented an increase in erythrocyte sedimentation rate (ESR) and C-reactive protein (CRP); mild leukocytosis without deviation; preserved kidney function and normal urine test; rheumatological panel with non-reactive rheumatoid factor (RF), non -reactive anti-nucleus factor (ANA), non -reactive Anti-SSA / Ro, non-reactive Anti-SSB /La, non-reactive Anti-DNA , non-reactive ASO, Anti-MPO non-reactive, non-reactive P-ANCA and reactive C-ANCA; non-reactive serologies for Hepatitis B, C, HIV 1 and 2 and non-reactive Veneral Disease Research Laboratory (VDRL). On imaging examination, chest tomography showed extensive panlobular pulmonary emphysema, thickening of interlobular septa in the middle and lower lung fields, suggesting interstitial fibrosis with extensive architectural distortion, presence of consolidation with air bronchogram in the lateral segment of the middle lobe and absence of findings suggestive of alveolar haemorrhage.

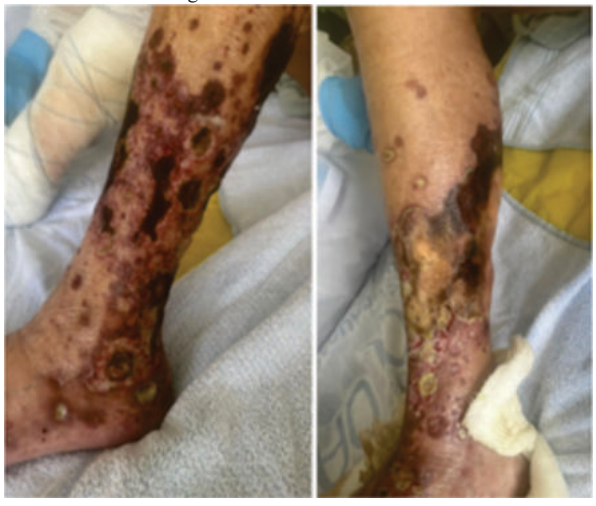

Figure 1. Bullous Lesions With Ulcers And Crusts On The Left Lower Limb.

A skin biopsy was performed showing blood vessels with swollen endothelium permeated by neutrophils, and more rarely lymphocytes, surrounded by a small amount of nuclear debris and mono- and polymorphonuclear inflammatory cells, with a result compatible with leukocytoclastic vasculitis.

Initially, the patient used broad-spectrum antibiotic therapy, due to bacterial pneumonia and skin infection, associated with colchicine 1 mg/day, with improvement in infectious criteria and weaning from supplemental oxygen. Subsequently, she presented an increase in skin ulcers and foci of necrosis, requiring chemical and surgical debridement. Due to the subsequent worsening of the condition, with hemoptoic sputum and melena with a positive faecal occult blood test, we opted for pulse therapy with methylprednisolone for three days, with excellent clinical response and significant remission of the lesions.

After clinical stabilization, a drop in inflammatory parameters and progressive tolerance to corticosteroid withdrawal, the patient was discharged from the hospital and followed up on an outpatient basis with a rheumatologist, pulmonologist and angiologist.

DISCUSSION

Vasculitis is classified according to the predominant cell type in the inflammatory infiltrate of the vascular process, which can be neutrophilic, lymphocytic or granulomatous. They can affect small, medium or large vessels. [4]

Although cutaneous involvement is the most common, systemic involvement occurs in around 57% of cases, with: renal, pulmonary, muscular, cardiac, gastrointestinal or neurological. [1] In the clinical case described, the patient presented predominantly cutaneous and gastrointestinal manifestations.

Histopathological characteristics depend on the location and time of evolution of the lesion. [4] The classic findings of cutaneous small vessel vasculitis include: postcapillary venules, fibrinoid deposits, endothelial oedema, and red blood cell extravasation. It is extremely important to emphasize that this pattern is not exclusive to a specific diagnosis. [5]

Cutaneous small vessel vasculitis affects both sexes equally and patients of all ages. A recent broad global study estimated the incidence at 4.5 per 100,000 inhabitants/year. Half of the cases had an idiopathic cause, which may have been drug-induced or post-infectious. [6]

Although there is no global consensus on the classification of vasculitis, it is believed that the causal agents can be divided into 4 categories, namely: infections (bacterial, viral, protozoa and helminths); drugs (hormonal contraceptives, vaccines, acetylsalicylic acid, antibiotics, thiazide diuretics); chemical products (insecticides and petroleum derivatives) and food allergens (milk protein and gluten); in association with other diseases such as neoplasms and rheumatological diseases. [two]

Leukocytoclastic vasculitis, also called Hypersensitivity vasculitis, is characterized by the involvement of small vessels, being more common in venules. [3] It is characterized by an immune-mediated vascular inflammatory process that occurs in the structural damage of vessels. [4] Its pathophysiological mechanism consists of the deposition of immune complexes, generally involving IgG and IgM immunoglobulins that activate the complement cascade and expression of adhesion molecules. [3,4]

Within the subclassification of leukocytoclastic vasculitis there are 2 mechanisms for ANCA positivity. In the first proposed mechanism, ANCA binds to a natural killer cell that is activated, releasing cytotoxic mediators against the target cell. In the second mechanism, molecules from the complement system (specifically the C5-C9 complex) bind to ANCA, which activates these molecules and exerts a lytic effect on the target cell. [7.8]

Regarding the characteristics of skin lesions, in the acute form of the disease, mainly haemorrhagic, purpuric and necrotic lesions are observed. In the subacute phase, the lesions may be purpuric, erythematous -macular or urticarial, with nodules and small necrosis possible. In the chronic form, erythematous macules predominate. Purpuric lesions usually disappear within three to four weeks, eventually remaining a hyperpigmented or atrophic lesion. These manifestations may recur or become chronic. [3]. Complementary tests, such as blood count, tests of inflammatory activity, serum complement levels, serology and antibody panel, are not specific for diagnosis, but help in the assessment of organic dysfunctions. [3]

In the clinical case in question, it was not possible to accurately define the precipitating factor for the worsening of the disease, since the patient had symptoms for approximately three years. According to Fiorentino, the aetiology of cutaneous vasculitis can be idiopathic in up to 55% of cases. In the patient under study, infectious exacerbation of COPD and smoking may have contributed to the activation of the disease. [9]

The treatment currently used varies according to the severity and extent of the disease. The gold standard treatment proposed in the literature is the use of corticosteroids at a dose of 1 mg/kg/day and/or cyclophosphamide 2 mg/kg/day, generally maintained for 6 months, with progressive reduction according to tolerance. In cases of limited skin disease, colchicine at a dose of 0.5 mg/day can be used. [1,6,7]

CONCLUSIONS

Leukocytoclastic vasculitis is characterized by inflammation of small vessels, mediated by the deposition of immune complexes. It is known that this pathology is associated with ANCA, through two mechanisms. According to a study carried out at Istanbul Training and Research Hospital, only 2.4% of patients diagnosed with leukocytoclastic vasculitis were ANCA positive. [10]

Despite the diagnostic complexity of the case and the presence of possible factors activating the disease, there was no definitive etiological elucidation, even though we achieved a successful therapeutic response using gold standard treatment medications. In this way, contributing to the increase in the diagnosis of leukocytoclastic vasculitis.

It is important to highlight that the current literature on leukocytoclastic vasculitis and its association with ANCA remains scarce, suggesting the need for new studies on the topic.

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