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## Case report

### Rheumatoid vasculitis – Case report<sup>☆</sup>



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#### ABSTRACT

Rheumatoid arthritis (RA) is a chronic systemic inflammatory autoimmune disease and its main manifestation is persistent synovitis affecting peripheral joints symmetrically, in spite of its destructive potential, the evolution of RA is highly variable. Some patients may have only a short-term process oligoarticular with minimum lesion, while others suffer a polyarthritis evolving with progressive and continuous involvement of other organ systems such as skin, heart, lungs, muscles and blood vessels rarely leading to rheumatoid vasculitis. The aim of this study was to describe a case of rheumatoid vasculitis a rare and severe condition.

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#### Vasculite reumatoide – Relato de caso

##### RESUMO

A artrite reumatoide (AR) é uma doença crônica autoimune inflamatória sistêmica e sua principal manifestação é a sinovite persistente, que compromete articulações periféricas de forma simétrica. Apesar do seu potencial destrutivo, a evolução da AR é muito variável. Alguns pacientes podem ter apenas um processo de curta duração oligoarticular com lesão mínima, enquanto outros sofrem uma poliartrite progressiva e contínua e evoluem com acometimento de outros órgãos e sistemas, como pele, coração, pulmões, músculos e mais raramente vasos sanguíneos, que leva à vasculite reumatoide. O objetivo deste estudo foi descrever um caso de vasculite reumatoide, uma condição rara e grave.

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##### Palavras-chave:

Vasculite

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## Introduction

Systemic vasculitis has been a diagnostic challenge in the areas of clinical medicine and rheumatology for many years. The rheumatoid vasculitis, rare and severe complication of rheumatoid arthritis, is one of them. The most common cutaneous presentation of rheumatoid vasculitis is the development of palpable purpura in the lower extremities that if not treated early enough can evolve into a multi-organ systemic vasculitis.<sup>1</sup>

## Case report

64-year old, non-Caucasian, female patient, with RA, without treatment for 25 years. Six years ago, developed edema in her left lower limb, erythema and hard lumps that ulcerated in the lateral leg region with moderate serosanguineous secretion. She was treated with multiple topical and systemic antibiotics, but showed no improvement. The size of the lesion evolved until it involved the distal two-thirds of the left lower limb. Two years later, she presented similar ulceration in the right leg, for which she received several topical, unspecific treatments, but did not obtain a good response. Due to the impairment of her general condition and worsening of the lesions, she sought a health care unit, where she was evaluated and referred for hospital treatment. Upon admission to the hospital, the patient was in regular general condition – emaciated, presenting asthenia and anemia, claiming morning stiffness for over an hour, showing ulnar deviation of the fingers in symmetrical hands with bony prominences both in the metacarpophalangeal and the proximal interphalangeal joints (Fig. 1), impairment of bilateral elbows and knees. She also presented shallow-edged ulcerated lesions with moderate serosanguineous secretion in the distal two-thirds of her lower limbs (Fig. 2).



**Fig. 1 – Hand deformities. Ulnar deviation of the fingers with bony prominences both in the metacarpophalangeal and the proximal interphalangeal joints, secondary to rheumatoid arthritis.**



**Fig. 2 – Vasculitis in lower limbs. Shallow-edged ulcerated vasculitic lesions with moderate serosanguineous secretion in the distal two-thirds of the lower limbs, secondary to rheumatoid arthritis.**

She was diagnosed with seropositive RA according to the criteria of the 1987 American College of Rheumatology, presenting over four criteria (morning stiffness, arthritis of 3 or more joint areas, arthritis of hand joints, symmetric arthritis and positive rheumatoid factor) and scoring greater than 6 in the new classification criteria for rheumatoid arthritis, 2010 ACR/EULAR (joint involvement score 5, serology score 3, duration of symptoms score 1), DAS disease activity score<sub>28</sub>: 4.83 (moderate activity), complicated by rheumatoid vasculitis. The diagnostic was made according to clinical evaluation of the rheumatology service. She was treated with methotrexate 15 mg/week after systemic antibiotics and topical treatment of the ulcers, showing significant improvement of the lesions in the lower limbs and of her general condition. She was discharged from hospital to continue the treatment as out-patient.

## Laboratory tests

CBC: RBC 3.81 million/mL, hemoglobin 7.8g; hematocrit 24.6%; mean corpuscular volume 64.6 fL; mean corpuscular hemoglobin 20.5 pg; mean corpuscular hemoglobin concentration 31.7 g/dL; anisocytosis index 20.4%; leukocytes 2950; segmented 44%; lymphocytes 39%; monocytes 8%; platelets 404 thousand. Hematoscopy: microcytic erythrocytes; intense hypochromia; numerous red blood cells in target and rare elliptocytes; presence of Rouleaux; serum iron 29 mcg/dL (reference 60–160 mcg/dL); transferrin saturation index 18.13% (reference 20–25%); erythrocyte sedimentation rate 35 mm; creatinine 0.5 mg/dL; urea 15 mg/dL; rheumatoid factor 94 UI/mL (reference: until 20 UI/mL, turbidimetry, repeated and confirmed); non-reactive anti HIV 1–2 antibodies; anti HBs, reagent (193.6 mUI/mL). Reference: non-reagent less than 10 mUI/mL; non-reagent HbsAg; non-reagent anti-HCV. Total abdominal ultrasonography: without alterations, with normal spleen volume and homogeneous texture.

## Discussion

Rheumatoid arthritis (RA) is a relatively common chronic inflammatory autoimmune disease that affects men and women of all ages, with worldwide presence. It affects 0.3–2.1% of the world population, but in some, such as the black population in the Caribbean and rural population in Sub-Saharan Africa, the disease is less recurrent and less severe.<sup>2</sup> In Brazil, it is estimated that around 1.3 million people are affected, being more common in the age group between 30 and 50 years old, mostly women than man, in a proportion of 3:1.<sup>3</sup> Its clinical presentation is characterized by the joint involvement, in an insidious process that often results in deformities. However, it is important to acknowledge that RA is a systemic disease. Because of that, in addition to joint manifestations, several organs and specific systems can be affected, enabling the onset of various extra-articular manifestations.<sup>4</sup> These manifestations are usually observed in individuals with high titers of rheumatoid factor and anti-CCP.<sup>5</sup> Our patient presented rheumatoid factor of 94 UI/mL (reference limit up to 20 UI/mL), three times greater than the upper limit of normal.

Rheumatoid vasculitis is a form of cutaneous presentation with annual incidence of 12.5/1 million that typically affects small and medium-sized vessels, with associated peripheral neuropathy (often motor), digital gangrene, nail bed infarcts and palpable purpura.<sup>6</sup> The spectrum of clinical lesions reported in the rheumatoid vasculitis is broad and varies with the size and the location of the affected vessels and the extent of the disease.<sup>7</sup> This is a rare and severe complication that must be diagnosed early enough, given the large interference in the evolution, treatment and prognosis of the underlying disease. Our patient did not undergo an anatomopathological examination (biopsy) as it is not easily accessible in the institution. The diagnosis was based on the clinical presentation and the experience of the Hospital's rheumatology service.

Due to the presence of neutropenia, we investigated the possibility of Felty's syndrome (rheumatoid arthritis, neutropenia and splenomegaly). However, the patient did not show signs of splenomegaly in the physical examination or in the abdominal ultrasound, removing this hypothesis.

The patient presented microcytic, hypochromic anemia with low iron levels and high RDW, low transferrin saturation index compatible with iron-deficiency anemia, associated to low ingestion of specific foods, according to dietary recall, associated to anemia of chronic disease.

One of the aspects that make this case particularly interesting is the fact that this disease evolved for many years without any kind of treatment for the underlying disease (rheumatoid arthritis) or the vasculitis, which highlights the importance of proper diagnosis and early treatment of the disease.

## Conclusion

Despite being a rare condition, it is important to be aware of the development of rheumatoid vasculitis in the clinical course of rheumatoid arthritis so the appropriate treatment can be promptly instituted, thus avoiding the progression of lesions and possible associated complications.

## Conflicts of interest

The authors declare no conflict of interest.

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