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Rheumatoid Vasculitis Complicating Rheumatoid Arthritis: A Case Report

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Abstract

Introduction: Among the chronic and feared complications of this disease, rheumatoid vasculitis stands out as one of the most severe, albeit rare. The most frequently affected sites by rheumatoid vasculitis are the skin and the peripheral nervous system. We report a case of rheumatoid vasculitis complicating a 30-year history of untreated rheumatoid arthritis. **Case Report:** The patient is a 75-year-old male with a 30-year history of deforming and erosive seropositive rheumatoid arthritis. He presented with polyarthritis and digital necrosis. Physical examination revealed peripheral joint syndrome with characteristic deformities of rheumatoid arthritis. Additionally, there was well-demarcated dry gangrene affecting the first and second digits of the right hand. Laboratory findings indicated an inflammatory syndrome. Tests for antinuclear antibodies (ANA) and anti-neutrophil cytoplasmic antibodies (ANCA) to investigate other causes of vasculitis were negative. The patient received a corticosteroid bolus. After two weeks, there was a marked reduction in pain and an improvement in the general condition. The dry gangrene remained stable. **Conclusion:** Rheumatoid vasculitis remains a serious and chronic complication of rheumatoid arthritis, associated with significant mortality. This case highlights the crucial importance of early diagnosis and effective management of rheumatoid arthritis.

Keywords

Rheumatoid Arthritis, Vasculitis, Necrosis, Burkina

1. Introduction

Rheumatoid arthritis (RA) remains the most common chronic inflammatory rheumatism and is an autoimmune disease characterized by a variety of articular and extra-articular symptoms [1]. Among the chronic and feared complications of this condition, rheumatoid vasculitis stands out as one of the most severe, albeit rare [1]. It involves inflammation of small and medium-sized blood vessels, adding further complexity to the management of RA [2]. The most frequently affected sites in rheumatoid vasculitis are the skin and peripheral nervous system. The pathophysiology of rheumatoid vasculitis is based on a type III hypersensitivity reaction, where immune complexes activate the complement system, leading to chemotactic recruitment of neutrophils and resulting in endothelial damage to blood vessels [3]. Small vessel rheumatoid vasculitis is generally a leukocytoclastic vasculitis, characterized by neutrophilic and eosinophilic infiltrates, with red blood cell extravasation and fibrinoid necrosis of the vessel walls [4]. The prevalence of RA associated vasculitis is estimated to be between 1% and 5% [2]. To date of our knowledge, no case of rheumatoid vasculitis has been described in Burkina Faso literature, as the diagnosis of rheumatoid vasculitis remains a significant challenge, requiring rigorous exclusion of other causes of vasculitis, a step that is often not achieved in many medical settings like ours. We report a case of rheumatoid vasculitis complicating a 30-year history of untreated RA.

2. Case Report

A 75-year-old male farmer with seropositive rheumatoid arthritis (rheumatoid factor at 170 IU and anti-citrullinated peptide antibodies at 59 U/ml), deforming and erosive. The RA symptoms had been progressing for approximately 30 years. The diagnosis was made 3 months ago, and a treatment regimen of methotrexate 15 mg and folic acid 10 mg was initiated, albeit with irregular follow-up. He presented to the rheumatology department with general deterioration, polyarthritis, and digital necrosis, which was preceded by a month-long history of burning-type paresthesia in the fingers. Physical examination revealed a general health status classified as stage 3 according to the World Health Organization, with 26 painful joints, 2 swollen joints, and 5 deformed joints, including: Z-thumb deformity of both thumbs, ulnar drift of the hands, and mallet deformity of the fifth digit of the right hand. Additionally, there was well-demarcated dry gangrene of the first and second digits of the right hand (**Figure 1**), accompanied by paresthesia at their tips. Peripheral pulses were palpable, and the examination of other systems was unremarkable. Laboratory tests revealed an inflammatory syndrome with a C-reactive protein level of 76.73 mg/L and microcytic hypochromic anemia with a hemoglobin level of 10 g/dL. There was no leukocytosis, and renal and hepatic functions were normal, as were muscle enzymes. Tests for antinuclear antibodies (ANA) and anti-neutrophil cytoplasmic antibodies (ANCA) to investigate other causes of vasculitis were negative. HIV serology and hepatitis serologies

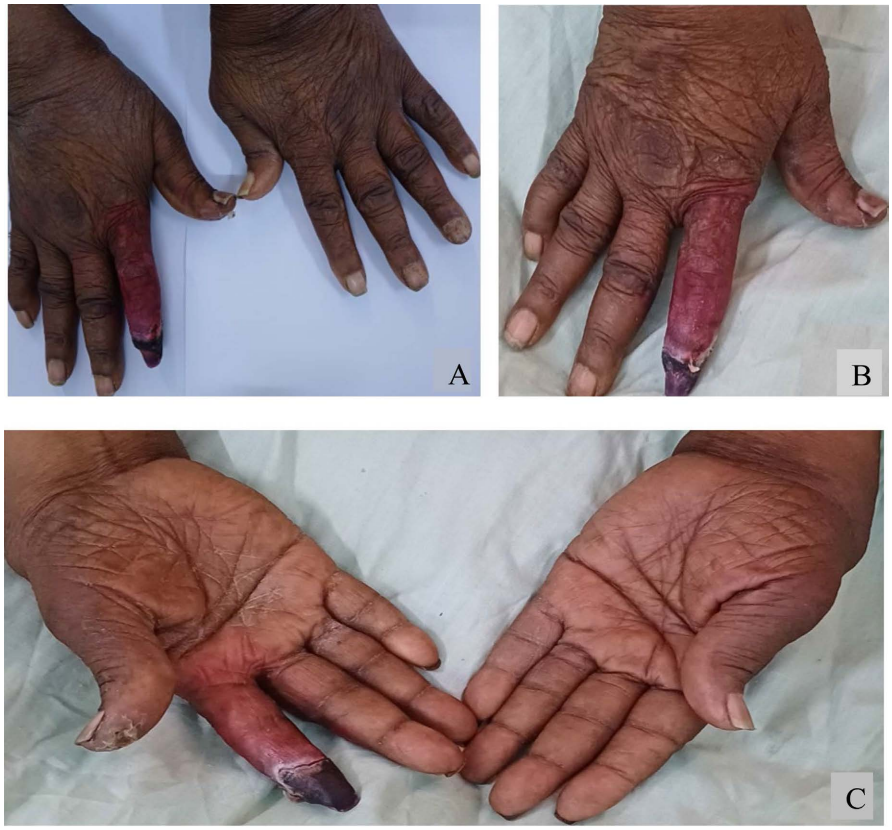

Figure 1. (A) Dorsal view of the hands showing dry gangrene of the 1st and 2nd fingers; (B) Dorsal view of the right hand clearly displaying necrosis of the first two fingers, with more pronounced involvement of the 2nd digit; (C) Palmar view of the hands highlighting more pronounced dry gangrene on the pulp of the 2nd finger of the right hand.

were normal. Radiographs of the hands and feet showed erosion of the head of the fifth metatarsal, fusion of the carpal bones, and deformities. A CT angiography of the right upper limb was normal. Based on these clinical and paraclinical findings, a diagnosis of rheumatoid vasculitis complicating a flare of rheumatoid arthritis was made. Digital disarticulation was indicated, but the patient did not consent. He received 3 infusions of 240 mg methylprednisolone, followed by an oral prednisolone regimen at 0.5 mg/kg/day, with a progressive tapering of 10% every 10 days over 6 months. The RA treatment, particularly methotrexate, was continued. After two weeks, the patient experienced a reduction in pain and an improvement in general condition. The dry gangrene remained stable.

3. Discussion

Rheumatoid vasculitis typically emerges at an advanced stage of rheumatoid arthritis (RA), generally after more than 10 years of disease progression [1]. It is an inflammatory condition affecting small and medium-sized vessels, primarily seen in a subset of patients with progressive RA. In our patient, vasculitis manifested after 30 years of untreated RA, during which time the patient had relied on traditional remedies. The diagnosis was established three months ago, but there has

been poor adherence to therapy.

The understanding of rheumatoid vasculitis began to take shape in the 1960s [3]. Since then, dozens of isolated cases have been documented in the global literature [3]. Multiple studies have analyzed risk factors for developing extra-articular manifestations in RA. These include high levels of rheumatoid factor, the presence of anti-citrullinated peptide antibodies, and low complement levels [4] [5]. In a study by Makol *et al.* involving 86 patients with rheumatoid vasculitis and 172 controls, several risk factors were identified, such as younger age at RA diagnosis, smoking, vasculopathy, severe RA with erosions, and the use of biologic therapies and other disease-modifying anti-rheumatic drugs (DMARDs) in addition to hydroxychloroquine and methotrexate [2]. In our patient, the identified risk factors were age, and more significantly, the severity of RA, which was erosive and deforming due to a prolonged diagnostic delay (over 30 years) and nonadherence to treatment. Identifying specific risk factors is crucial for the prevention and management of rheumatoid vasculitis in RA patients.

The diagnosis of rheumatoid vasculitis is typically based on dermatological manifestations, such as purpura or digital necrosis, in most cases [6]. In numerous series, the skin or peripheral nerves are affected in over 90% of patients [6]. However, rheumatoid vasculitis can involve virtually any organ in addition to the skin and nervous system, including the heart, kidneys, lungs, and eyes [2]. In our patient, rheumatoid vasculitis manifested as dermatological signs, specifically digital necrosis, preceded by neurological symptoms, such as finger paresthesias. The diagnosis of vasculitis was confirmed by excluding other causes of vasculitis, particularly connective tissue diseases, drug-induced causes, and ANCA-associated vasculitis [7].

A biopsy of the necrotic tissue to identify characteristic lesions could not be performed due to the patient's refusal. Furthermore, the presence of anti-CCP2 antibodies in RA patients is associated with progressive joint damage as well as severe extra-articular manifestations [8].

The management of rheumatoid vasculitis is not well-established. In all cases, DMARDs should be continued to stabilize RA activity. Clinical practices are largely based on personal experience, as only two open-label studies recommend the use of cyclophosphamide and high doses of glucocorticoids in patients with rheumatoid vasculitis [8]. Biologic therapies have not demonstrated efficacy [9]. Amputation is often indicated in cases of massive necrosis, as in our patient, at advanced stages, highlighting the need for timely management to improve patient outcomes.

Isolated skin involvement without other organ involvement has a better prognosis in rheumatoid vasculitis. Several studies have reported high morbidity and mortality rates, ranging from 33% to 43%, with infection being the leading cause [3] [10].

4. Conclusion

Rheumatoid vasculitis remains a serious and chronic complication of rheumatoid

arthritis (RA) and is associated with significant mortality. This case highlights the crucial importance of early diagnosis and effective management of RA to minimize severe extra-articular complications, such as rheumatoid vasculitis. Furthermore, early recognition of vascular complications in RA allows for effective management of rheumatoid vasculitis to ensure a favorable prognosis for patients.

Conflicts of Interest

The authors declare no conflicts of interest regarding the publication of this paper.

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