CLINICAL VIGNETTE

Rheumatoid Arthritis with Lymphoplasmacytic Panniculitis: A Rare Extraarticular Manifestation

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Introduction

Rheumatoid arthritis (RA) is a chronic, systemic autoimmune disorder primarily characterized by persistent symmetric polyarthritis. While joint inflammation and damage are the hallmarks of RA, a considerable proportion of patients can develop extraarticular manifestations involving various organ systems.[1] These extraarticular complications can precede, coincide with, or follow the onset of articular disease and often contribute to increased morbidity and mortality.[1] Common extraarticular manifestations include rheumatoid nodules, pulmonary involvement, cardiovascular disease, and vasculitis.[2] The skin lesions associated with RA are diverse and can be categorized as neutrophilic dermatoses, vasculitic lesions, granulomatous lesions, and rheumatoid nodules.[3] Panniculitis, an inflammation of the subcutaneous fat, is an exceptionally rare cutaneous manifestation of RA.[4]

Case Presentation

A 58-year-old male presented with a 5-year history of joint pains, morning stiffness, and intermittent skin lesions. Physical examination revealed swelling and tenderness in the small joints of the hands and feet, suggestive of active synovitis characteristic of RA. Additionally, multiple palpable, tender, raised erythematous skin lesions were observed across the anterior chest wall and back (Figures 1 and 2).

Laboratory Findings

- The patient's laboratory results included normal complete blood count, comprehensive metabolic panel, muscle enzymes, and urinalysis. Tests for rheumatoid factor, anti-dsDNA, complements, anti-Scl-70, anti-Ro/La, anti-centromere antibodies, Jo1, Mi-2 protein, and cryoglobulins were also negative. Additionally, ANCA (antineutrophil cytoplasmic autoantibody), RPR, HIV (Human Immunodeficiency Virus), and hepatitis serologies were negative, and immunoglobulin G (IgG) subsets were normal. Abnormal findings included a positive antinuclear antibody (ANA) at a 1:1280 titer with a centromere pattern, a positive anti-cyclic citrullinated peptide (anti-CCP) antibody > 250 Units/mL, and elevated erythrocyte sedimentation rate (ESR) and C-reactive protein (CRP).

Imaging Studies

- Chest CT scan revealed cutaneous nodules without pulmonary involvement.
- Ultrasound of the joints showed active inflammatory arthritis with erosive changes

Skin Biopsy

The biopsy of one of the truncal lesions demonstrated superficial and deep lymphoplasmacytic panniculitis, with diffuse perivascular and subcutaneous chronic inflammatory infiltrates, consistent with a cutaneous manifestation of rheumatoid arthritis.

Treatment and Outcome

Upon diagnosis of lymphoplasmacytic panniculitis as an extraarticular manifestation of CCP positive active rheumatoid arthritis (RA), the patient was initiated on oral steroids and hydroxychloroquine as standard therapy for RA. However, despite this initial treatment, there was only partial improvement in the joint symptoms, and the skin lesions persisted. Recognizing the inadequate response, leflunomide, another disease-modifying antirheumatic drug (DMARD), was added to the treatment.

Despite the addition of leflunomide, the patient continued to experience active synovitis and persistent skin lesions. At this point, considering the severity of the symptoms and the refractory nature of the disease, a biologic agent was deemed necessary. Abatacept, a selective costimulation modulator, was chosen due to its mechanism of action targeting T-cell activation, which is central to the pathogenesis of RA. Anti TNF (Tumor Necrosis Factor) agent was initially considered but due to positive ANA and anti-Centromere antibody, co-existing undifferentiated connective tissue disease (UCTD) was possible.

Following abatacept therapy, the patient’s joint pains and skin lesions improved significantly. The active synovitis resolved, with reduced joint swelling and tenderness. Additionally, the erythematous skin lesions became less tender and gradually faded, indicating a favorable response to treatment.

Discussion

Panniculitis is a distinct group of inflammatory disorders primarily affecting the subcutaneous adipose tissue. The histopathological diagnosis of panniculitis involves a systematic approach, evaluating the pattern of involvement (septal vs. lobular), the presence or absence of vasculitis, and the predominant inflammatory cell type (neutrophilic vs. lymphocytic).[5,6]

Histological evidence of lymphocytic lobular panniculitis is always a great diagnostic challenge.[7,8] Possible diagnoses range from benign conditions, such as lupus panniculitis (LP), to indolent or aggressive types of T-cell lymphoproliferative disorders, such as subcutaneous panniculitis-like T-cell lymphoma. Integration between clinical, histological, and immunophenotypic features is necessary to obtain a definitive diagnosis.[7,8] In cases like our case, where lymphoplasmacytic panniculitis is observed in the context of rheumatoid arthritis, careful consideration of histopathological findings along with clinical features is needed for accurate diagnosis and appropriate management.[5]

Lobular lymphoplasmacytic panniculitis, as observed in this case, can be associated with various connective tissue diseases, including systemic lupus erythematosus and dermatomyositis.[9] However, its occurrence in rheumatoid arthritis is exceptionally rare. Erythema nodosum is the most reported form of panniculitis in patients with RA.[4] Other cutaneous manifestations of RA encompass a wide spectrum, including rheumatoid nodules, granulomatous dermatitis, neutrophilic dermatoses, and rheumatoid vasculitis.[3]

In patients with RA presenting with atypical skin lesions, it is crucial to consider and rule out potential infectious etiologies or underlying malignancies, particularly in those receiving immunosuppressive therapies. Comprehensive diagnostic evaluation, including skin biopsies and appropriate laboratory investigations, is warranted to establish the correct diagnosis and guide appropriate treatment.

Conclusion

Cutaneous findings associated with sterile inflammatory infiltrates, such as panniculitis, are uncommon in rheumatoid arthritis. However, physicians should be aware of these rare manifestations and consider them in the differential diagnosis of RA patients presenting with atypical skin lesions. Prompt recognition and appropriate management of extraarticular complications are crucial for improving overall disease outcomes in patients with rheumatoid arthritis.

Figures

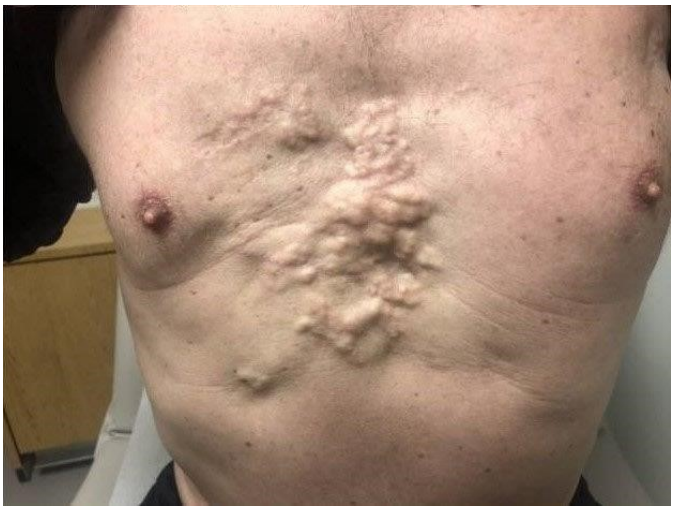

Figure 1. Lobulated skin lesion on the chest of patient with rheumatoid arthritis.

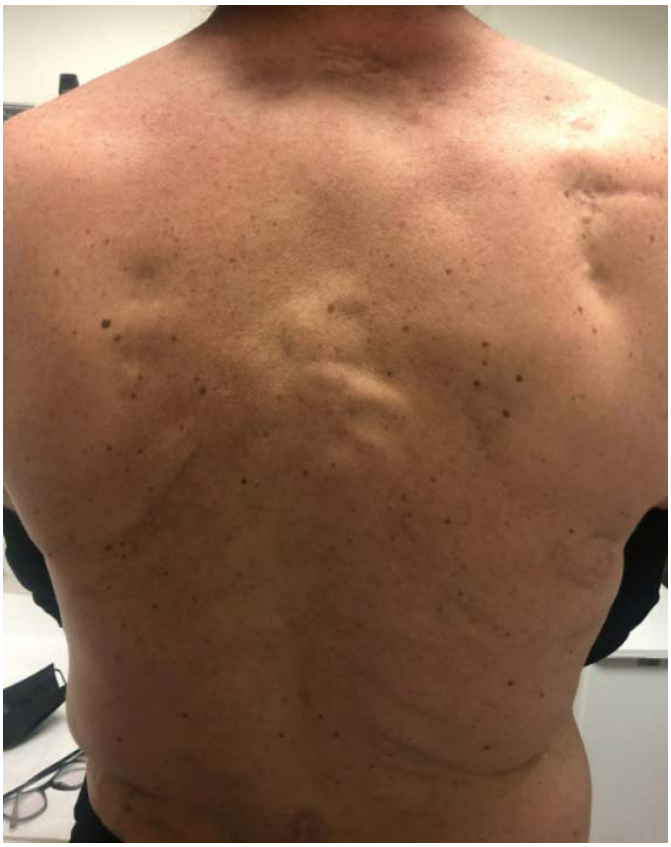

Figure 2. Lobulated skin lesion on the back of patient with rheumatoid arthritis.

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