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CASE REPORT

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Neutrophilic dermatoses in a seronegative rheumatoid arthritis patient: A case report

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Abstract

Rheumatoid arthritis (RA) is a chronic inflammatory disease, characterized by symmetric and destructive polyarthritis with a broad-spectrum clinical manifestation of various organs. RND is an unusual distinctive manifestation of RA and typically develops in severe RA. This report aims to present an unusual and a rare neutrophilic skin condition, in a seronegative RA Sudanese patient. A 51-year-old woman was diagnosed with RA three years ago and a history of bilateral polyarthritis, presented with a skin rash involving her extremities and abdomen. Clinical examination of her skin revealed the presence of maculopapular lesions affecting the extensor surfaces of the lower extremities and the lower part of the abdomen with hyperpigmentation. Hand X-ray demonstrated periarticular osteopenia, and laboratory and immunological studies that include C-reactive protein, erythrocyte sedimentation rate (ESR), rheumatoid factor (RF), anticitrullinated peptide antibodies (ACPAs), and antinuclear factor in addition to skin biopsy were all suggested a diagnosis of neutrophilic dermatosis. The patient received steroids for the skin lesion still no significant improvement was seen, and then, cyclosporin 100 mg was administrated twice/ day with close monitoring, and two weeks later marked improvement was shown.

KEYWORDS

cyclosporine, RND, seronegative RA, skin lesions, steroids

1 | INTRODUCTION

Rheumatoid arthritis (RA) is a common inflammatory synovial disorder characterized by symmetrical polyarthritis and associated with joint destruction.[1,2] RA has a broad-spectrum clinical manifestation of various organs. Extra-articular findings and lesions due to skin involvement may also coexist, which include vasculitis, pyoderma gangrenosum, rheumatoid nodules, and leg ulcers.[3,4] In addition to these findings, palmar erythema, urticaria, vitiligo, neutrophilic lobular panniculitis, and RND may less commonly occur.[5]

Rheumatoid neutrophilic dermatitis (RND) typically progresses in severe RA several years after the diagnosis of RA and manifests as symmetric erythematous and urticarial lesions over the extensor surfaces of the extremities.[6] Dermatological involvement tends to occur in patients with more severe or active RA.[7,8] In this study, we present a case of RND, a rare neutrophilic skin condition, in a seronegative RA Sudanese patient treated with cyclosporine.

2 | CASE PRESENTATION

A 51-year-old Sudanese woman was diagnosed with RA in 2018 based on the EULAR classification of RA. She had a history of bilateral symmetrical polyarthritis with involvement of small joints of the hands, both wrists, and shoulders; they are also associated with morning stiffness lasting for two hours. She was treated successfully with methotrexate and prednisolone, and her disease remains in remission. A year ago, she started to develop an itchy skin rash with pus involving her lower extremities and abdomen. She had no fever, weight loss, skin rashes, bowel symptoms, aphthous ulcers, or hair loss. At present, she has no joint pain or extra-articular manifestations. Her systemic review was unremarkable. The patient had hypertension for 5 years, and her family history revealed no history of connective tissue diseases. In terms of drug history, she is on amlodipine 5 mg for hypertension. Her musculoskeletal examination showed no synovitis, and her skin examination showed nodulopapular lesions affecting the extensor surfaces of the lower extremities particularly on thighs and the lower part of the abdomen with hyperpigmentation as shown in Figure 1. Laboratory tests revealed normal liver function test and renal function test, C-reactive protein 12.9 (normal range below 5 mg/L), erythrocyte sedimentation rate (ESR) is 70 mm/h (normal range is 1–13 mm/h for men and 1–20 mm/h for women), normal urine analysis, and blood sugar level 98 mg/dl (normal range 70–99 mg/dl). With regard to immunological studies, the following values, namely, rheumatoid factor (RF) <8 (negative), anticitrullinated peptide antibodies (ACPAs) <4 (<20, negative), and antinuclear factor <1/80 (negative), are obtained. Hand X-ray demonstrated periarticular osteopenia, and the chest x-ray was normal. Skin lesion biopsy showed multiple foci of necrosis, mixed inflammatory infiltrates composed mainly of neutrophils, hyperkeratosis, and acanthosis, and no evidence of vasculitis. The biopsy findings were consistent with a diagnosis of neutrophilic dermatosis (Figure 2). The patient received topical and systemic steroids for the skin lesion; however, no significant improvement was noticed, and then, she was started on cyclosporin 100 mg twice a day with close monitoring of the serum creatinine and blood pressure, and a few weeks later her skin lesions and general condition showed marked improvement.

3 | DISCUSSION

Dermatological complications in patients with RA can be specific or related to skin disease due to the treatment taken by a patient.[9] RA-specific skin lesions are seen only in patients with RA and include accelerated rheumatoid nodules, rheumatoid vasculitis, and RND.[6] Rheumatoid arthritis-related skin diseases include dermatoses that are associated with RA.[10,11] This group of skin disorders includes pyoderma gangrenosum, Sweet's syndrome, Raynaud's phenomenon, and erythema elevatum diutinum (EED). RND was first described by Ackerman in the last century. It is a rare and distinct manifestation of RA with high frequencies in women compared to men; only less than 20 cases had been described until now.[12,13]

The prevalence of RND is less than 2% among all patients with RA.[14] The exact pathophysiology of the condition is unknown; however, it is believed to be an immune complex-mediated disease.[15] RND commonly appears in

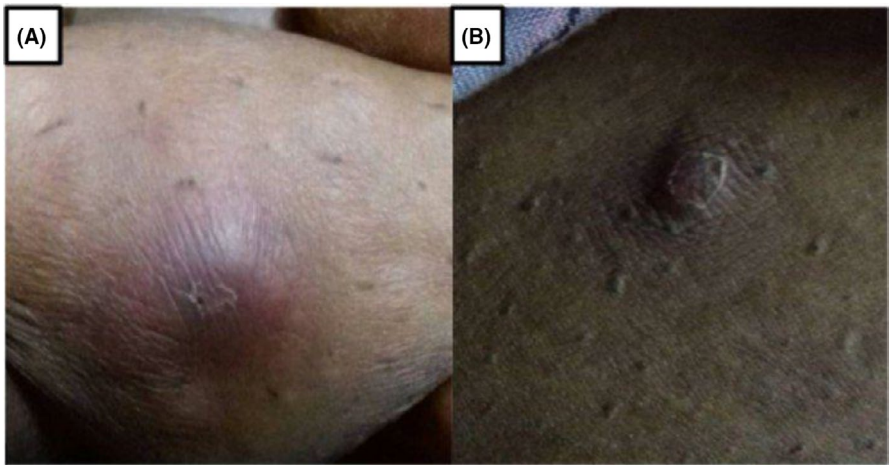

FIGURE 1 (A) Shows a large nodule on the knee, and (B) shows small papules and large crusted nodule

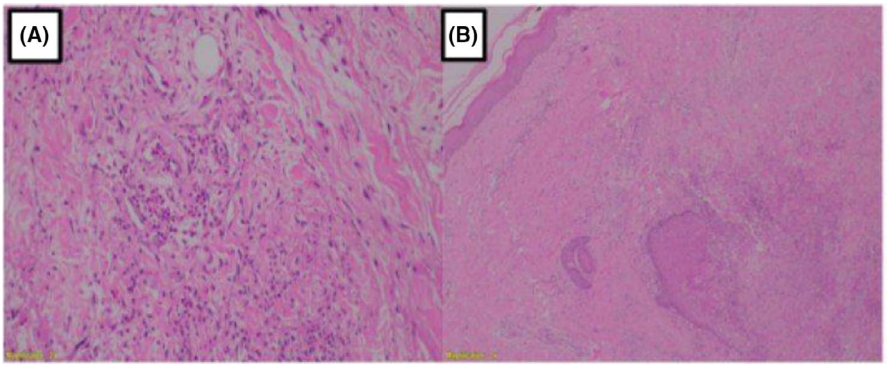

FIGURE 2 (A) Skin biopsy shows multiple foci of necrosis and mixed inflammatory infiltrate composed mainly of neutrophils. No evidence of vasculitis. (B) Skin biopsy shows hyperkeratosis and acanthosis

patients with severe, chronic seropositive arthritis and clinically manifests as asymptomatic nodules, erythematous papules, and urticaria-like lesions.[6,16] The dermatological lesion in RND can be either palpable purpura or plaque or nodule in the extensor surfaces of extremities, and joints are favored locations as in our presenting patient who showed nodulopapular lesions in the thighs and the lower part of the abdomen.[16,17] The RND skin biopsy under the microscope will show a neutrophilic infiltrate,[18] and this is the same histopathological feature of the skin biopsy revealed in our patient, as it showed mixed inflammatory infiltrate with no evidence of vasculitis.

The differential diagnosis was considered and ruled out, which includes dermatitis herpetiformis ruled out by the absence of bullous lesions, a drug eruption was also ruled out because the lesions persisted and worsened in spite of cessation of medications, and Sweet's syndrome was also excluded as the patient was not on any of the medications that can cause a Sweet's-like drug hypersensitivity such as azathioprine and nonsteroidal anti-inflammatory drugs.[19] Behcet's disease, bowel-associated dermatosis arthritis syndrome, and pyoderma gangrenosum were ruled out by history and the absence of ulcerative or vesiculopustular lesions. Erythema elevatum diutinum was ruled out by the absence of vasculitis.[20] RND may be treated with topical or systemic corticosteroids; however, in case of no improvement, cyclophosphamide will provide better improvement.[21]

In conclusion, a middle-aged Sudanese woman with a three-year history of RA presented with a skin rash, and examination revealed the presence of maculopapular lesions affecting the extensor surfaces of the lower extremities and the lower part of the abdomen with hyperpigmentation. Laboratory studies and skin biopsy have suggested a diagnosis of neutrophilic dermatosis. The patient received cyclosporin 100 mg and was well improved.

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CONFLICT OF INTEREST

None.

AUTHOR CONTRIBUTIONS

The case was diagnosed and managed by EME, histopathology was performed by AMA; treatment and follow-up were performed by AAIR and AGAM; and writing, editing, and draft were made by AAIR, MEAE, and AEAB.

ETHICAL APPROVAL

Obtained.

CONSENT

Written informed consent was obtained from the patient to publish this report in accordance with the journal's patient consent policy.

DATA AVAILABILITY STATEMENT

All the data used in the study are available from the first and corresponding author on reasonable request.

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