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Case Report

# A Case of Rheumatoid Neutrophilic Dermatosis

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**Key Words:** rheumatoid neutrophilic dermatosis, rheumatoid arthritis

**Abstract**

**Observations:** Rheumatoid neutrophilic dermatosis (RND) is a rare and distinct manifestation of rheumatoid arthritis. It is more common amongst women and patients who have severely seropositive rheumatoid arthritis (RA). 57 years old man with RA was seen and diagnosis of RND was established. Because of its rare occurrence and tendency to be misdiagnosed as other pustular dermatosis cases, we considered to present this case.

## Introduction

Rheumatoid arthritis (RA) is a chronic inflammatory synovial disease characterized with symmetrical poliarthritis and associated with joint destruction. Extra-articular findings and lesions due to skin involvement may also co-exist [1, 2, 3]. Most common skin manifestations of RA include rheumatoid nodules, vasculitis, leg ulcers and pyoderma gangrenosum. In addition to these findings, urticaria, palmar erythema, vitiligo, interstitial granulomatous dermatitis, neutrophilic lobular panniculitis and rheumatoid neutrophilic dermatosis (RND) may less commonly occur [1].

57 years old man with RA was seen and diagnosis of RND was established. Because of its rare occurrence and tendency to be misdiagnosed as other types of pustular dermatosis, we considered to present this case.

## Case Report

57 years old man was admitted to our outpatient clinic 1.5 months ago with a number of small, pustular lesions and red dots resembling pimples. Patient's history revealed diagnosis of RA (16 years ago) for which he was taking tablets of prednisolon (10 mg daily) and leflunomide (100 mg daily). No other significant finding was retrieved from his history. On dermatologic examination there were generalized pustules which were 1 mm in diameter with an erythematous base. These pustules were remarkably present on upper limbs, trunk, lower limbs and antecubital fossa sparing the face, hands and feet. Some of these pustulas were drained and revealed plaques and papules associated with peripheric desquamation (**Figure 1**). No lymphadenopathies were observed. On routine laboratory examination, leukocyte count of 16580/mm$^3$ (a percentage of 88 being neutrophiles), erythrocytes sedimentation rate of 47 mm/hour, C-reactive protein level of 122 mg/L (0-5 mg/L), alanine transaminase level of 53 U/L (0-41 U/L) and rheumatoid factor level of 503 (less than 10 normally) revealed as abnormal results. Cysts of

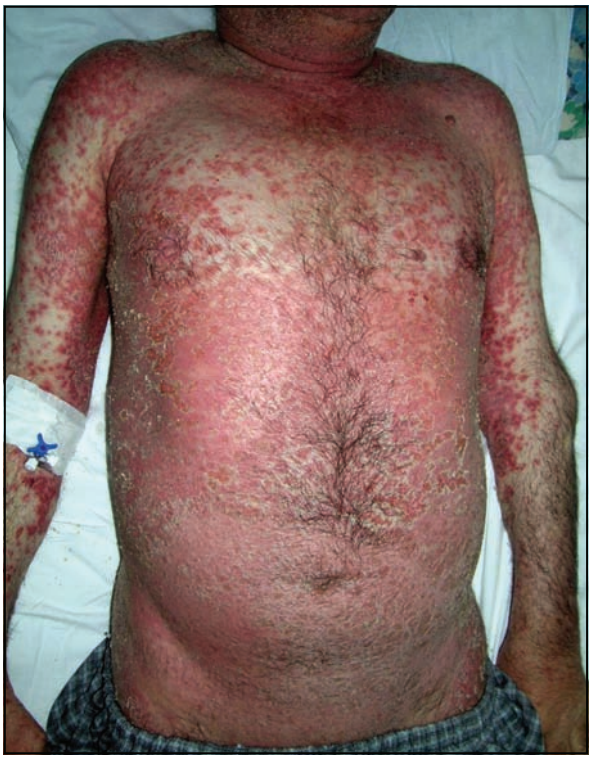

**Figure 1.** Generalized pustulas which are 1 mm in diameter and are located on erythematous background. Except for face, hands and feet, these pustulas were remarkably present on upper limbs, trunk ,lower limbs and antecubital fossa. Some of these pustulas were drained and revealed plaques and papules associated with peripheric desquamation.

*Blastocystis hominis* were abundant on stool. HAV IgG, anti-HBs and anti-HBc IgG markers of hepatitis were positive.

Patient had fever. No radiologic abnormality of either the lungs or sinus (*Waters* view) was revealed. Titer of antibody to *Streptolysin O* was 48 and was in normal range. There were no additional abnormalities in other laboratory measurements. Skin biopsy sample, when evaluated histopathologically, revealed abundant neutrophilic infiltration foci on dermis and epidermis. Formation of abscesses which are composed of neutrophiles were also observed (**Figure 2**).

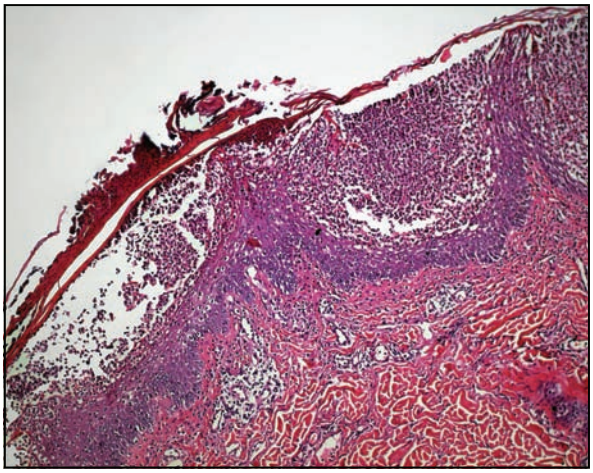

**Figure 2.** In histopathologic examination, revealed abundant neutrophilic infiltration foci on dermis and epidermis. Formation of abscesses which are composed of neutrophiles were also observed. *(Hematoxylin and Eosin*, × 10)

Clarithromycin 500 mg (twice daily), methotrexate 25 mg/per week, folic acid 5 mg (once daily), chlobetasol propionate topical (once daily) and cream containing 4% concentration of urea were added to the treatment. At the third week of treatment, a prominent regression of lesions appeared and the lesions resolved at the end of the fourth week.

## Discussion

RND was first described by *Ackerman* in 1978. It is a rare and distinct manifestation of RA. Its frequency is unknown, but only 14 cases had been reported up to day. This manifestation mostly occurs amongst patients who have strong positivity of rheumatoid factor [**1, 2, 4**]. However, occurrence amongst sero-negative patients was also reported [**1, 4**]. It usually involves middle-aged women [**1**]. Our patient had a high level of rheumatoid factor and clinic state was consistent with RA.

In RND, the presenting lesion is either a papule, or palpable purpura, or plaque or nodule which usually resembles urticaria and characteristically has a yellowish-red color. Annular, ulcerative, pustular, vesicular and/or crusted lesions may also occur less commonly. Trunk, shoulders, neck and extensor faces of limbs are the most commonly involved sites of body. Lesions tend to be located symmetrically [**1, 2, 4**]. In our case, except for face hands and feet, these lesions were present on upper limbs, trunk, lower limbs and antecubital fossa being significantly incident on latter. Some of these

pustules were drained and revealed plaques and papules associated with peripheral desquamation.

Histopathologically, on the background of dense and neutrophilic infiltration there are usually leukocytoclasis and endothelial swelling without vasculitis. Features such as spongiosis, formation of intraepidermal neutrophilic abscess, subepidermal edema and papillary dermal neutrophilic micro-abscess which are also observed in our case may be shown [**1, 2, 4**].

*Sweet*'s Syndrome, erythema elevatum diutinum, urticarial vasculitis, neutrophilic urticaria, pyoderma gangrenosum and Behçet's disease should be considered as differential diagnosis [**1, 4**]. *Sweet*'s syndrome usually follows a non-specific gastrointestinal or upper respiratory tract infection and involves face and limbs. Association with fever, fatigue and/or arthralgia is common. However RND is usually asymptomatic. Pyoderma gangrenosum, *Behçet*'s disease or *Sweet*'s syndrome may be associated with manifestations such as oral aphthae, genital ulcers and iritis [**4**]. In our case the lesions were located symmetrically and pustules were scattered throughout trunk and limbs. Absence of necrotic ulcers, oral aphthae, genital ulcers and iritis, erythematous papules and nodules on face and limbs were the distinguishing features from pyoderma gangrenosum, *Behçet*'s disease and *Sweet*'s syndrome respectively. Since clinical similarity with pustular psoriasis exists, this condition should be considered, as well. However, there were no personal or family history of psoriasis, rheumatoid factor positivity or parakeratosis and papillomatosis histopathologically.

Certain treatment of RND still remains unclear. It may either resolve spontaneously or follow after resolution RA [**2**]. Systemic and/or topical steroids, systemic hydroxychloroquine, dapsone, cyclophosphamide, colchicine and etretinate are the options of therapy [**1, 2, 4**]. In our case, complete resolution was established in four weeks using topical and systemic steroid and methotrexate.

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