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Rheumatoid Neutrophilic Dermatosis: Diagnostic Challenge in a Patient with Rheumatoid Arthritis

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Abstract **Case Report**

Rheumatoid neutrophilic dermatosis (RND) is an extremely rare cutaneous manifestation of rheumatoid arthritis (RA), with few cases reported in current medical literature. Its educational value lies in raising awareness among dermatologists about this uncommon entity, usually associated with severe forms of RA, and in highlighting the importance of early recognition for appropriate and effective management.

Keywords: Rheumatoid neutrophilic dermatosis, rheumatoid arthritis, cutaneous manifestations, neutrophilic infiltrate, histopathology.

INTRODUCTION

Rheumatoid neutrophilic dermatosis (RND) is a rare neutrophilic skin disorder associated with rheumatoid arthritis (RA). It was first described by Ackerman in 1978 and remains an underrecognized entity, with fewer than 100 cases reported in the literature to date. Histologically, it is characterized by a dense neutrophilic infiltrate in the dermis without evidence of true vasculitis. RND typically occurs in patients with long-standing, severe RA, although it has also been reported in early stages and, in rare cases, as an initial manifestation.

Clinically, the lesions present as asymptomatic erythematous papules, nodules, or plaques, mainly located on the extensor surfaces of the extremities. These findings can mimic other neutrophilic dermatoses, such as Sweet's syndrome or pyoderma gangrenosum, and even cutaneous vasculitis, making its diagnosis a challenge.

The pathogenesis of RND is not fully understood, but it is thought to involve dysregulation of the innate immune system and excessive neutrophilic response, possibly mediated by pro-inflammatory cytokines such as IL-1, IL-6, and TNF-α—molecules also central to the pathophysiology of RA.

Accurate diagnosis requires histopathologic evaluation, which helps distinguish RND from other neutrophilic or vasculitic conditions. Most cases respond favorably to treatment focused on controlling the underlying RA, with topical or systemic corticosteroids occasionally used as adjunctive therapy.

We present a case of RND in a patient with RA, emphasizing the clinical presentation, histopathologic findings, and therapeutic approach, along with a brief literature review to raise awareness of this uncommon dermatologic entity.

CASE REPORT

A 61-year-old male patient with a known history of rheumatoid arthritis presented to the dermatology clinic with erythematous skin lesions of three weeks' duration. The lesions were located on the anterior chest, inframammary folds, and right axilla. The patient denied fever, pain, or systemic symptoms, and reported no recent medication changes or infections.

On dermatological examination, multiple erythematous, infiltrated plaques with annular and circinate configurations were observed on the anterior thorax and inframammary areas. Some lesions exhibited raised borders with central clearing and mild desquamation. In the right axilla, coalescing plaques with a geographic pattern and smooth surface were noted. There were no signs of ulceration, purulence, or mucosal involvement (Figures 1–5).

A skin biopsy was performed. Histopathologic analysis revealed a dense dermal infiltrate composed predominantly of mature neutrophils, with areas of nuclear debris (karyorrhexis) and no evidence of vasculitis or fibrinoid necrosis. These findings were consistent with a diagnosis of rheumatoid neutrophilic dermatosis (Figure 6).

Given the patient's known diagnosis of rheumatoid arthritis and the compatible clinical and histological features, treatment was focused on optimizing the management of the underlying rheumatologic condition. Low-dose systemic corticosteroids were initiated, with gradual improvement of the cutaneous lesions over the following weeks.

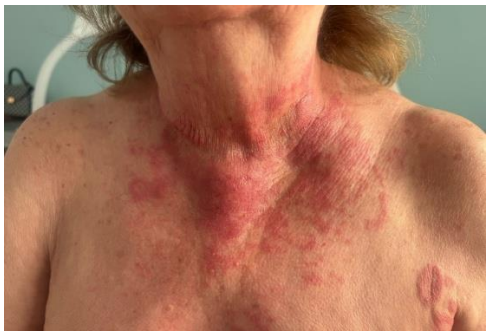

Figure 1: Erythematous, infiltrated plaques on the anterior chest, some with annular and circinate shapes, slightly raised borders and central clearing

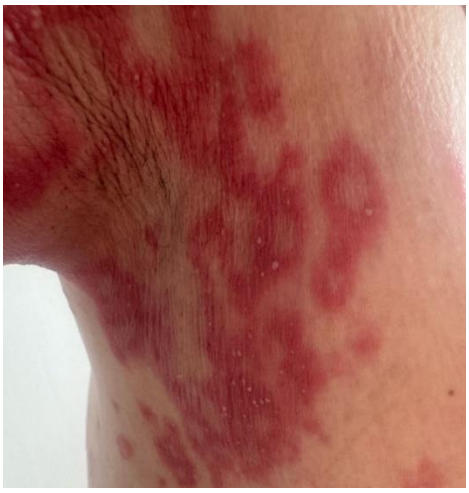

Figure 2: Grouped erythematous plaques in the right axillary region, with geographic configuration and smooth surface

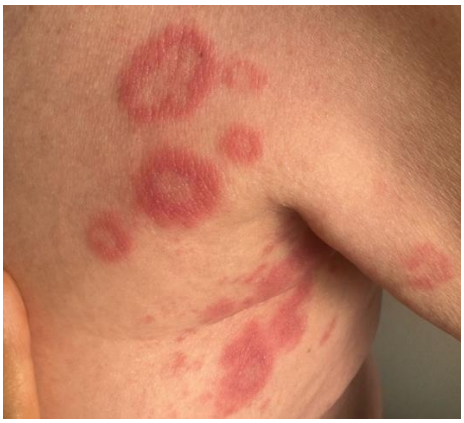

Figure 3: Erythematous, annular plaques with well-defined active borders and pale centers in the left inframammary region

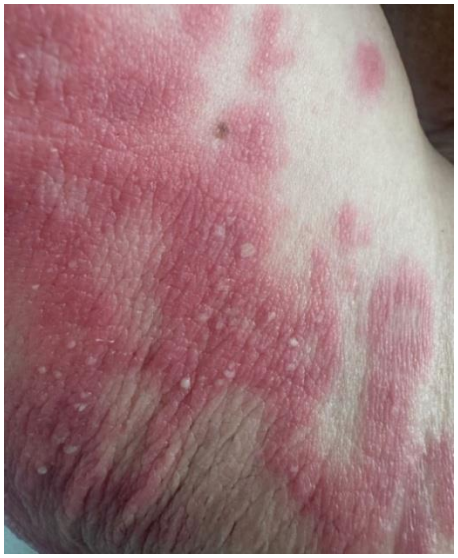

Figure 4: Close-up view showing a rough surface with mild desquamation and small pustules on an erythematous base

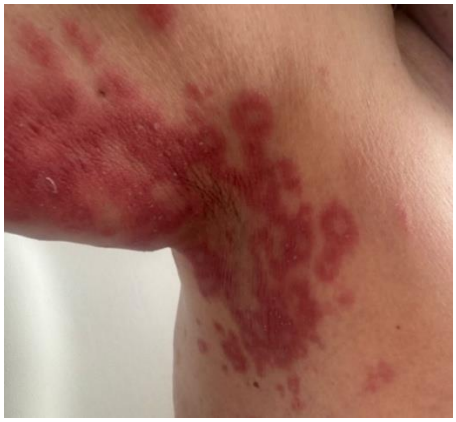

Figure 5: Coalescing erythematous plaques in the right lateral thoracic and axillary area, with a smooth and infiltrated appearance

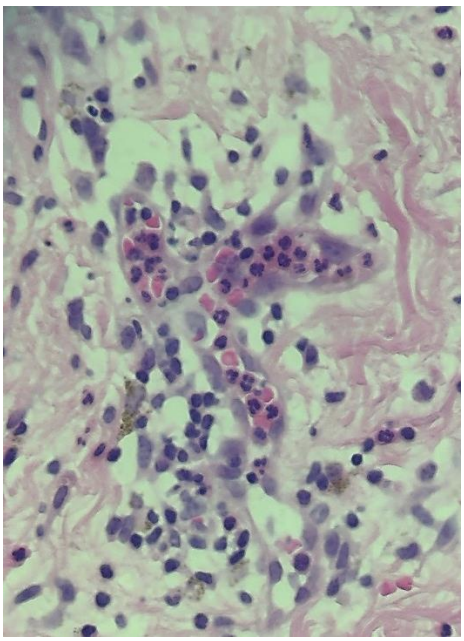

Figure 6: Histopathological image (H&E, 40x) showing a dense dermal infiltrate composed predominantly of mature neutrophils without evidence of frank vasculitis. Fragmented nuclei (karyorrhexis) are also observed, a feature consistent with rheumatoid neutrophilic dermatosis.

DISCUSSION

Rheumatoid neutrophilic dermatosis (RND) is an uncommon cutaneous manifestation of rheumatoid arthritis (RA), first described by Ackerman in 1978. Fewer than 100 cases have been reported in the literature, reflecting its rarity and the diagnostic challenge it presents for dermatologists and rheumatologists alike [1]. Due to its clinical similarity to other neutrophilic dermatoses and cutaneous vasculitis, RND is often misdiagnosed.

Clinically, RND typically manifests as asymptomatic or mildly pruritic erythematous papules, nodules, or plaques, commonly affecting extensor surfaces of the limbs. However, atypical presentations—such as lesions on the trunk or intertriginous areas, as observed in our patient—have also been described [4]. Walling and Sontheimer noted that most patients with RND have severe or long-standing RA, although a few reports describe cases appearing early in the disease course [4].

Histopathologically, RND is characterized by a dense neutrophilic infiltrate in the dermis, without evidence of leukocytoclastic vasculitis [2,5,6]. This is a key finding that helps differentiate RND from rheumatoid vasculitis. In their series of six cases, Tan *et al.,* emphasized the importance of obtaining deep biopsies, as the inflammatory infiltrate may extend to the deep dermis or subcutaneous tissue [6]. Some cases may also exhibit band-like neutrophilic infiltrates and focal necrosis, but without the fibrinoid necrosis characteristic of true vasculitis.

The pathogenesis of RND remains unclear. However, it is believed to fall within the spectrum of neutrophilic dermatoses, such as Sweet's syndrome and pyoderma gangrenosum, all of which involve dysregulated activation of the innate immune system. Marzano *et al.,* highlighted the shared pathogenic features of these conditions, including the overexpression of pro-inflammatory cytokines such as IL-1, IL-6, and TNF-α—cytokines also implicated in RA [3]. This suggests that RND may represent a cutaneous manifestation of systemic inflammatory activity.

Treatment primarily focuses on controlling the underlying RA. Most patients respond well to adjustment of disease-modifying antirheumatic drugs (DMARDs) or the introduction of corticosteroids. Weedon and others reported favorable responses with topical or systemic corticosteroids, especially in symptomatic or extensive cases [2]. For persistent or recurrent lesions, escalation to biological therapy may be considered [4,10]

Requena and Sánchez Yus emphasized that one of the most common diagnostic pitfalls in RND is misidentification as an infectious or vasculitic process, which can lead to unnecessary or overly aggressive treatments [8]. Momen and Ghosh described a case in which histologic examination helped avoid misdiagnosis and the inappropriate use of cyclophosphamide [7].

This case contributes to the body of literature by highlighting an unusual presentation of RND and reinforcing the importance of including it in the differential diagnosis of cutaneous eruptions in patients with RA. Prompt biopsy and interdisciplinary evaluation are essential for accurate diagnosis, appropriate management, and prevention of unnecessary interventions.

CONCLUSIONS

Rheumatoid neutrophilic dermatosis is a rare but important cutaneous manifestation of rheumatoid arthritis, often associated with severe or long-standing disease. Its clinical presentation can mimic other neutrophilic dermatoses or vasculitic processes, which underscores the need for a high index of suspicion and histopathologic confirmation.

This case reinforces the importance of considering RND in the differential diagnosis of inflammatory skin lesions in patients with rheumatoid arthritis. Early recognition and appropriate treatment based on accurate diagnosis can prevent unnecessary interventions and imp

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