

Rheumatoid Neutrophilic Dermatitis: Case Report and Review

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Rheumatoid neutrophilic dermatitis (RND) is a rare cutaneous condition that manifests in patients with rheumatoid arthritis (RA) and is characterized by erythematous papules, plaques, nodules, and urticarial lesions on joints, extensor surfaces of the extremities, and the trunk. Although the condition most commonly is reported in patients with severe seropositive RA, it also presents in patients with seronegative RA and must be differentiated from other neutrophilic dermatoses. Because the lesions have a cosmetic effect on patients, managing the underlying RA may help improve cutaneous manifestations of RND. We report a case of RND in a woman with RA and review the literature on the condition.

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Rheumatoid arthritis (RA) is a chronic synovial inflammatory disease that is characterized by polyarthritis and associated joint destruction, primarily in a symmetric distribution. Coexisting cutaneous involvement of the extra-articular surfaces also can manifest in patients with RA.¹ Common skin manifestations of RA include rheumatoid nodules, vasculitis, and pyoderma gangrenosum. Less frequently, patients with RA may develop urticaria; vitiligo; neutrophilic lobular panniculitis; and rheumatoid neutrophilic dermatitis (RND),² a rare condition that was first described by Ackerman et al³ in 1978. Most commonly, RND affects middle-aged women with severe seropositive RA and appears as erythematous

papules, plaques, nodules, and urticarial or annular lesions.⁴ However, there also have been cases of RND reported in patients with seronegative RA.^{5,6} Predominantly occurring on joints, extensor surfaces of the extremities, buttocks, and the trunk, lesions can be asymptomatic but often are mildly tender and usually are symmetrically distributed with a predilection for the dorsal hands.^{7,8} Because these lesions are cosmetically displeasing, managing the underlying RA may help improve the cutaneous manifestations of RND. We report a case of RND in a patient with RA and review the etiology, histopathology, differential diagnosis, and treatment of RND.

CASE REPORT

A 61-year-old woman presented with recurrent painful red papules on her back, lateral aspect of the arms, and posterior aspect of the legs over 35 years. According to the patient, the lesions continually appeared and resolved for several years, with periods of months without lesions. Her medical history included RA. The patient previously was treated with betamethasone dipropionate ointment and intralesional triamcinolone acetonide along with oral antibacterial regimens of ciprofloxacin and doxycycline

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with no improvement. Physical examination revealed 3- to 5-mm erythematous urticarialike papules scattered on the patient's legs (Figure 1), upper arms, and back. A punch biopsy was performed on a representative papule located on her back (Figure 2). Histologic examination revealed a neutrophilic dermatosis characterized by a nodular and diffuse collection of neutrophils and extravasated red blood cells. No interface dermatitis or leukocytoclastic vasculitis was present.

COMMENT

Rheumatoid neutrophilic dermatitis is a rare cutaneous reaction in patients with underlying RA characterized by erythematous papules, plaques, nodules, and urticarial lesions on joints, extensor surfaces of the extremities, and the trunk, commonly in a symmetric distribution. In most reported cases, patients with RND have severe RA with high titers of rheumatoid factor; however, cases also have been reported in patients with seronegative RA.^{2,5,6}



Figure 1. Three- to 5-mm erythematous urticarial papules on the leg that also were found on the upper arms and back.

Etiology

The pathogenesis of RND largely remains unknown; however, Jorizzo et al⁹ suggested that neutrophilic dermatoses are immune complex mediated. Rheumatoid neutrophilic dermatitis occurs in patients with RA, which is an immune complex-mediated inflammatory condition; therefore, because circulating immune

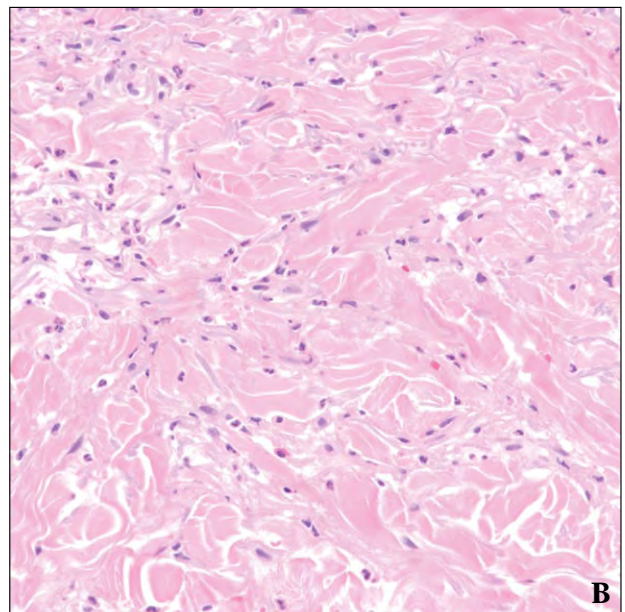
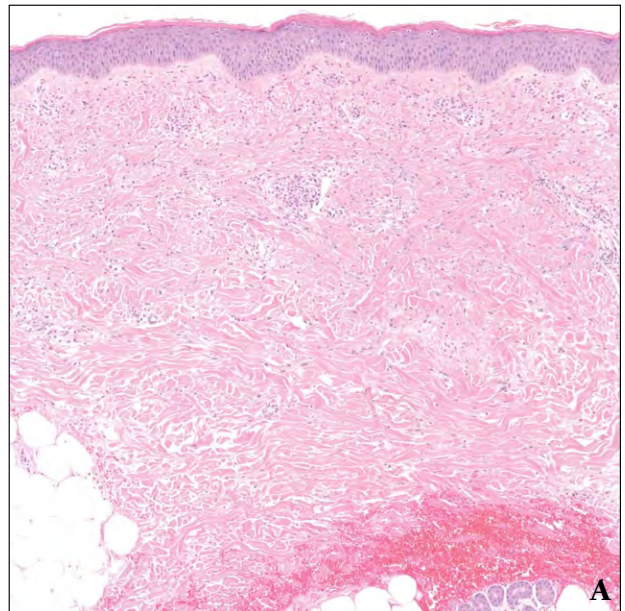


Figure 2. Histologic examination revealed a neutrophilic dermatosis characterized by a nodular and diffuse collection of neutrophils and extravasated red blood cells (A)(H&E, original magnification $\times 100$). A diffuse collection of neutrophils was present (B)(H&E, original magnification $\times 400$). No interface dermatitis or leukocytoclastic vasculitis was present.

complexes involving rheumatoid factor may precipitate the development of RND, this condition should be implicated as the potential result of a neutrophilic vascular reaction.¹⁰ Another cause of RND has been attributed to the release of the chemotactic cytokines IL-6 and IL-8, which may lead to the accumulation of neutrophils in the skin.¹¹

Histopathology

Histopathologically, RND is characterized by a dense dermal neutrophilic infiltrate without vasculitis.⁵ Microabscesses may form within the dermis because of the abundant number of neutrophils.⁴ Additionally, leukocytoclasia, basophilic collagen degeneration, and papillary dermal edema are histopathologic findings that are commonly demonstrated in RND.⁸

Differential Diagnosis

Rheumatoid neutrophilic dermatitis must be differentiated from other neutrophilic dermatoses; most challenging is its distinction from Sweet syndrome because of the similar clinical and histopathologic presentations for both conditions. Sweet syndrome is well-described as acute febrile neutrophilic dermatosis and is characterized by accompanying systemic symptoms. Leukocytosis, fever, and malaise have been reported in up to 78% of patients with Sweet syndrome.¹² In comparison, RND typically is asymptomatic. Additionally, lesions seen in Sweet syndrome typically involve the face, neck and extremities, characteristically in an asymmetric distribution.¹³ Histologically, both RND and Sweet syndrome exhibit a prevalent neutrophilic infiltrate with the presence of leukocytoclasia and absence of vasculitis.^{4,10} Furthermore, 16% of cases of Sweet syndrome reportedly are associated with infection or an immunologic disease such as RA.¹⁴ Because RND and Sweet syndrome are associated with RA, both may represent a spectrum of neutrophilic reactions that are mediated by the immune complex or other factors demonstrated in RA.⁵

Treatment

Treatment of RND often is challenging and different therapies have been reported with varying success. Controlling the underlying RA may benefit the course of RND and improve the patient's cutaneous presentation.⁴ Reported therapies include topical or systemic corticosteroids, dapsone, hydroxychloroquine sulfate, methotrexate, and cyclophosphamide.⁷ Some success has been reported with colchicine and etretinate.¹⁵ Brown et al⁵ described oral dapsone as the treatment shown to be most effective in treating RND.

Cosmetic Concerns

Although cutaneous manifestations of RND typically are asymptomatic, the presenting lesions often have a cosmetic effect on patients. Most patients present with papules, plaques, or nodules resembling urticaria with a characteristic erythematous-yellow discoloration in various anatomical distributions, usually on joints, extensor surfaces of the extremities, the trunk, and buttocks; there also is a particular predilection for the dorsal aspect of the hands. Occasionally these lesions may have a covered crust.⁸ Managing the underlying RA may help improve the cutaneous manifestations of RND.

CONCLUSION

Rheumatoid neutrophilic dermatitis is one of the possible cutaneous manifestations in patients with RA and should be considered in the differential diagnosis irregardless of rheumatoid factor status.

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