

Rheumatoid Neutrophilic Dermatitis: Rare Cutaneous Manifestation of Rheumatoid Arthritis in a Patient with Palindromic Rheumatism

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Rheumatoid arthritis (RA) is associated with a variety of cutaneous disorders including rheumatoid nodules, vasculitis, leg ulcers, and pyoderma gangrenosum¹. Rheumatoid neutrophilic dermatitis (RND), first described by Ackerman in 1978, is a rare cutaneous manifestation of RA². It is characterized by well demarcated erythematous to violaceous, edematous papules and plaques, usually occurring symmetrically over joints and extensor surfaces. Histology reveals a dense neutrophilic infiltration that involves the entire dermis. Although initially believed to occur only in patients with severe seropositive RA, recent reports describe occurrence in patients with seronegative RA. We describe a case of RND in a patient with seronegative, palindromic-type rheumatism and associated asymmetrical pauciarticular arthritis.

A 44-year-old woman presented with a 2 year history of recurrent, painful, erythematous skin lesions affecting the dorsal surface of the hands. The lesions initially involved the skin overlying the metacarpophalangeal (MCP) joints of the left hand, and then spread to a similar site on the right hand. Ten years before development of skin lesions, she had experienced swelling of the left wrist, left 5th proximal interphalangeal (PIP) joint, and right sternoclavicular joint. Additionally, over the past 15 years she had experienced palindromic episodes of joint inflammation, characterized by joint pain and swelling, generally lasting for 3 days and followed by complete resolution. Initially, the episodes occurred about twice a month, but more recently were almost continuous. She was systemically ill and reported fatigue, poor appetite, and mild weight loss, but denied fever. Her only medications were occasional nonsteroidal antiinflammatory agents (NSAID) and acetaminophen. She reported no personal or family history of any other autoimmune or dermatological disease.

Physical examination showed two 3-cm, well marginated, erythematous plaques with raised firm borders over the 5th MCP region of each hand (Figure 1). The

plaques were indurated, slightly tender, and minimally blanching. No other dermatological abnormalities were identified. On musculoskeletal examination there was limited movement of the left wrist, with associated active synovitis, flexion deformity of the left 5th PIP joint, and swelling of the right sternoclavicular joint.

Laboratory findings were microcytic anemia (hemoglobin 90 g/l and mean corpuscular volume 73.3 fl) with a normal white blood cell count and platelet count. C-reactive protein was elevated at 42 mg/l and erythrocyte sedimentation rate was normal. Tests for rheumatoid factor (RF), antinuclear antibody, antineutrophilic cytoplasmic antibody, and complement C3 and C4 were negative or normal. Blood biochemistry was within normal limits. A skin biopsy specimen showed a neutrophilic infiltrate without vasculitis, with prominent papillary dermal edema (Figure 2).

A topical corticosteroid was started, with little improvement; however, a 2 week course of oral prednisone (50 mg/day) proved effective in clearing the skin lesions, which resolved with residual hyperpigmentation of the area. In view of ongoing joint disease and systemic symptoms, treatment with hydroxychloroquine (400 mg/day) was started, with no demonstrable effect after 6 months. Methotrexate (15 mg/week) was recently initiated; however, her joint symptoms have remained active and she continues to periodically experience new skin lesions that promptly resolved with prednisone (50 mg/day) therapy.

Since Ackerman first described RND as a rare disease that occurs only in patients with severe "malignant" RA, only a few cases of this entity have been reported². Until recently, all reported cases of RND have occurred in patients with severe disabling RA, usually associated with a positive RF in high titer³⁻⁹. Two recent reports described RND in patients with seronegative RA^{10,11}. We describe the first case of RND in a patient with seronegative asymmetrical pauciarticular arthritis and palindromic rheumatism.

The skin findings of RND are variable, but most



Figure 1. Well margined erythematous plaques with raised firm borders over the dorsal surface of each hand.

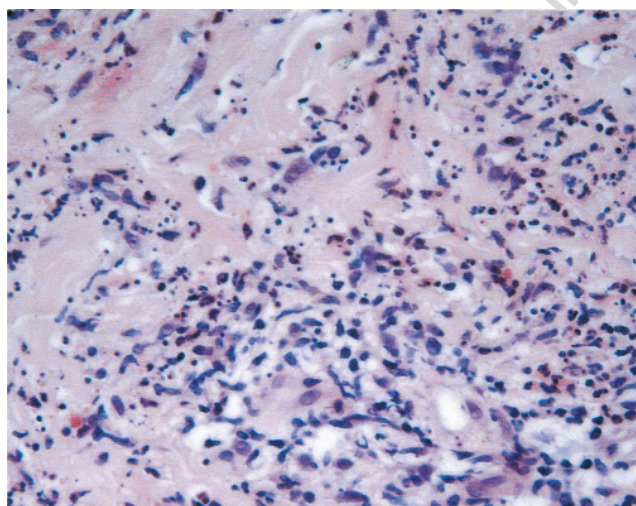


Figure 2. Dermal infiltrate of mainly neutrophils without evidence of vasculitis (hematoxylin-eosin, original magnification $\times 400$).

commonly present as well circumscribed erythematous papules, plaques, and nodules, which are occasionally covered by a crust³⁻¹¹. Rarely, annular, vesicular, and pustular lesions have been reported^{4,7-9}. The lesions tend to occur over the extensor surfaces of the extremities, the

trunk, and buttocks, and are often distributed symmetrically. They have a particular predilection for the dorsal surface of the hands, as in our patient. The lesions may be asymptomatic, but are frequently mildly tender. The histologic findings are characterized by the presence of a dense dermal infiltrate consisting mostly of neutrophils. The neutrophils may indeed be so numerous as to form micro-abscesses within the dermis^{4,6-8}. Vasculitis is not a feature of RND and is characteristically absent. Other common findings include leukocytoclasia^{4,5,7,8,10}, dermal papillary edema, and basophilic degeneration of collagen^{7,8,11}.

RND must be differentiated from other neutrophilic dermatoses, some of which may be associated with RA¹². These include Sweet's syndrome, erythema elevatum diutinum (EED), pyoderma gangrenosum, neutrophilic urticaria, urticarial vasculitis, bowel-associated dermatosis-arthritis syndrome, and Behçet's disease^{6,7,9}. RND may most commonly be confused with Sweet's syndrome and EED in the clinical setting, and the early stages of pyoderma gangrenosum histologically. Sweet's syndrome is characterized by recurrent painful edematous plaques, fever, peripheral leukocytosis, and a diffuse dermal neutrophilic infiltrate¹³. Similarly to RND, EED presents as erythematous plaques, papules, and nodules over the extensor

surfaces of the extremities, with a striking predilection for the skin overlying joints. Histological findings in EED are a dense neutrophilic dermal infiltrate and a widespread leukocytoclastic vasculitis, not seen in RND. In contrast, pyoderma gangrenosum is distinguished histologically by a perivascular neutrophilic infiltrate leading to necrosis and eventual ulceration and abscess formation.

Although the etiology of RND is largely unknown, Jorizzo, *et al*¹⁴ have suggested that the neutrophilic dermatoses are immune complex-mediated and have proposed the all-encompassing term "neutrophilic vascular reactions." The release of chemotactic cytokines such as interleukin 6 (IL-6) and IL-8 has also been implicated in the development of RND⁶. IL-8, in particular, acts as a neutrophil chemotactic factor, which may play a role in neutrophil accumulation in the skin.

RND is a rare cutaneous manifestation of RA, which probably occurs in a wider spectrum of patients with RA than previously described. The diagnosis should be considered in a patient with RA or palindromic rheumatism presenting with erythematous skin lesion, particularly localized over extensor surfaces of the joints.

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