

## Rheumatoid Neutrophilic Dermatitis: The Distinct Entity with Florid Presentation

Sir,

Rheumatoid neutrophilic dermatosis (RND) is a very rare cutaneous manifestation of severe rheumatoid arthritis (RA).<sup>[1]</sup> It usually presents as chronic, asymptomatic, symmetric erythematous and urticaria-like plaques and papules. The annular, vesicular, ulcerative or necrotic lesions are less frequently reported in a single patient. The diagnosis is confirmed on histology which shows dense dermal neutrophils with leukocytoclasia but no vasculitis, a feature described for sweet syndrome.<sup>[2]</sup>

A 21-year-old female presented with asymptomatic, red raised, and hyperpigmented lesions over extremities since 5 months. She had severe morning stiffness and debilitating joint pain and swelling over all the joints since 3 years. She was not on any medication since 6 months. On examination, multiple erythematous papules, pustules, and plaques were seen over both feet, legs extending to thighs and buttocks [Figure 1a and b]. Many of them were arranged in annular fashion with central hyperpigmentation and peripheral vesicles. Few lesions over the thighs were ulcerated and had necrotic eschar. Erythematous pseudovesicular papules and pustules were seen over bilateral forearms [Figure 1c]. Petechiae, pustules, and scaling were seen over bilateral soles [Figure 1d]. There was radial deviation of fingers. On routine investigation, hemogram showed microcytic anemia (hemoglobin: 10.4g/dl, MCV: 78fl)



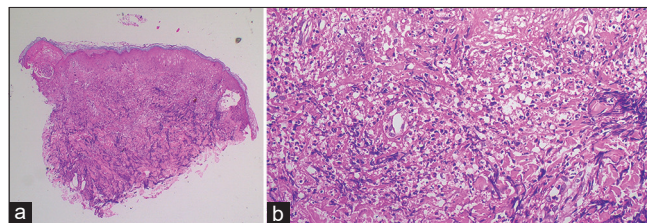
**Figure 1:** (a) Multiple erythematous papules and plaques present over bilateral lower legs. Few lesions arranged in annular fashion. (b) Similar lesions, some with necrosis extending upto the buttocks. (c) Erythematous and pseudovesicular papules present over the forearm. (d) Petechiae, pustules, and scaling over the sole

and raised total leukocyte count (13300 cells/cc<sup>3</sup>). Erythrocyte sedimentation rate was raised (45 mm/h). RA factor (titer 1: 340) and anticitrullinated protein antibody were positive. Antinuclear antibody was negative. X-ray of bilateral knee joints showed erosive arthritis. Other investigations were within normal range. Vasculitis, rheumatoid neutrophilic dermatosis, and sweet syndrome were kept as the provisional diagnosis. Punch biopsies were performed from papules on forearm and leg. Both the biopsies showed intraepidermal spongiosis, pustule, and papillary neutrophilic microabscesses. Dense diffuse infiltrate largely of neutrophils with their nuclear dust along with a few lymphocytes and plasma cells involved the whole of the dermis extending to the subcutis. Fibrinoid degeneration of collagen was seen [Figure 2a and b]. There was no evidence of vasculitis.

She was started on oral prednisolone 20 mg daily, oral methotrexate 10 mg/week, and topical corticosteroid (clobetasol propionate). Her symptoms improved and cutaneous lesions resolved over 3 weeks with no recurrence later on.

Extraarticular manifestation of RA (ExRA) occurs in 40% of RA patients mainly in older age with high titer of rheumatoid factor, early disability, and smoking.<sup>[1]</sup> Cutaneous ExRA includes rheumatoid nodules (most common), skin ulcers, Raynaud phenomenon, vasculitis, and pyoderma gangrenosum. Palisading granulomatous dermatitis, neutrophilic lobular panniculitis, sweet syndrome, and RND (<1%) are less common. Even coexistence of different cutaneous manifestations has been reported.<sup>[3]</sup> RND was first described by Ackerman in 1978 as neutrophilic dermatosis without vasculitis.<sup>[4]</sup>

Around 45 cases of RND are known so far. Mostly, the cases have presented as symmetric pseudovesicular and urticarial papules and plaques. Tense blisters, nodules, and chronic recurrent annular neutrophilic dermatosis comprise few unusual cases of RND.<sup>[5]</sup> Tender acral vesiculobullous



**Figure 2:** (a) Histopathology on scanner view from papule on forearm showing pustule, spongiosis, and dense diffuse dermal infiltration extending to subcutis (H and E 4×). (b) Higher magnification with abundance of neutrophils, nuclear dusts, and collagen degeneration (H and E 40×)

and purpuric lesions were reported by Soza and Griffin, but there was little evidence to differentiate it from sweet syndrome.<sup>[6]</sup> Along with the widespread presence of annular, vesicular, and ulcerated lesions, our patient had plantar pustular lesions mimicking pustular psoriasis, a feature which has been rarely described in RND so far.<sup>[5]</sup> The lesions over trauma prone areas in this case, supported the case reported by Zhang *et al.* on Koebner phenomenon in RND.<sup>[7]</sup>

It is extremely difficult to differentiate between sweet syndrome and RND. Many authors have previously tried to fit it same as sweet syndrome in RA.<sup>[8]</sup> In our case, the following features favor RND: 1) Occurrence of cutaneous lesions with worsening of RA and its resolution when RA was treated. 2) Nontender, persistent, and symmetric lesions, more over lower extremities. 3) The presence of plasma cells and lymphocytes admixed with predominant neutrophils spanning the whole dermis upto subcutis.

Neutrophilic dermatosis without vasculitis is also seen in pyoderma gangrenosum but the clinical presentation is distinct. This case is being reported to make us more cognizant of its widespread occurrence in young female having severe RA, because of the rarity of its occurrence with palmoplantar pustulosis and its clinical and histopathological resemblance to sweet syndrome.

### Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

### Financial support and sponsorship

Nil.

### Conflicts of interest

There are no conflicts of interest.

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<b>DOI:</b> 10.4103/idoj.IDOJ_161_19	

**How to cite this article:** Kumari I, Dongre A, Mohanty S. Rheumatoid neutrophilic dermatosis: The distinct entity with florid presentation. *Indian Dermatol Online J* 2020;11:114-5.

**Received:** April, 2019. **Revised:** April, 2019.

**Accepted:** June, 2019. **Published:** January, 2020.

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