

Rheumatoid neutrophilic dermatitis:

A report of two cases

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ABSTRACT

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Rheumatoid neutrophilic dermatitis (RND) has been recognized as a rare cutaneous manifestation of rheumatoid arthritis. The exact etiology remained unknown, but the possibility of immune complex activation, cell adhesion and migration and chemotactic cytokines has been proposed. The clinical manifestations are variable including erythematous or yellowish papules, plaques, nodules, urticarial-like, crusted, ulcerated or vesiculobullae. We describe two cases of rheumatoid neutrophilic dermatitis that were quite different in their clinical manifestations.

The conditions were good controlled by colchicine in both patients.

Key words: rheumatoid neutrophilic dermatitis, rheumatoid arthritis, neutrophilic dermatitis

บทคัดย่อ:

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หน่วยโรคผิวหนัง, ภาควิชาอายุรศาสตร์, คณะแพทยศาสตร์โรงพยาบาลรามาธิบดี, มหาวิทยาลัยมหิดล

Rheumatoid neutrophilic dermatitis เป็นลักษณะทางผิวหนังที่พบได้ไม่บ่อยในผู้ป่วยโรคข้ออักเสบ รูมาตอยด์ สำหรับสาเหตุยังไม่ทราบแน่ชัด คาดว่าอาจเกี่ยวข้องกับการกระตุ้นของอิมมูโนคอมเพลกซ์, ไซโตไคน์ต่างๆที่หลั่งออกมามีผลให้นิวโตรฟิล มายังบริเวณผิวหนังมากขึ้น ลักษณะทางคลินิกค่อนข้างหลากหลาย เช่น ผื่นสีแดงหรือสีเหลืองที่เป็นตุ่มนูนเล็ก/ใหญ่, ผื่นหนา, ผื่นคล้ายลมพิษ, ผื่นตกสะเก็ด, แผลถลอกลึก และ ตุ่มน้ำพอง รายงานผู้ป่วย Rheumatoid neutrophilic dermatitis 2 ราย ที่มีลักษณะทางคลินิกที่ต่างกัน ได้รับการรักษาด้วยโคลชิซินทั้ง 2 ราย พบว่าได้ผลดี

คำสำคัญ : ข้ออักเสบรูมาตอยด์, ผื่นอักเสบจากนิวโตรฟิล

Introduction

Rheumatoid neutrophilic dermatitis (RND) is a rare cutaneous reaction in patients with rheumatoid arthritis. It was first described by Ackerman in 1978.¹ The clinical presentations can vary from erythematous papules to plaques or nodules or urticarial lesions with symmetrical distribution over the extensor surfaces of extremities, trunk and back. Less commonly, annular, vesicular, ulcerative or crusted lesions occur.

Rheumatoid neutrophilic dermatitis lesions may be asymptomatic, pruritus or tender. It generally occurs in severe disabling rheumatoid arthritis with positive rheumatoid factor. The exact etiology of this clinical entity is still unknown. We describe two

cases of rheumatoid neutrophilic dermatitis that were quite different in their clinical manifestations.

Case 1

A 32-year-old woman, a known case of rheumatoid arthritis with severe articular involvement, presented with a complaint of intensely itchy papules on legs. These lesions then developed into ulcerated papules, ulcerated lesions. Her current medications were prednisolone 20 mg/day, azathioprine 50 mg/day, methotrexate 7.5 mg/week, hydroxychloroquine 400 mg/day and indocid 100 mg/day. Clinical manifestation revealed symmetrical multiple erythematous papules, crusted papules and ulcers at both lower extremities (Figure 1). She had active arthritis of the both wrist joints at the time of presentation.



Figure 1 Symmetrical multiple erythematous papules, crusted papules and ulcers at both lower extremities

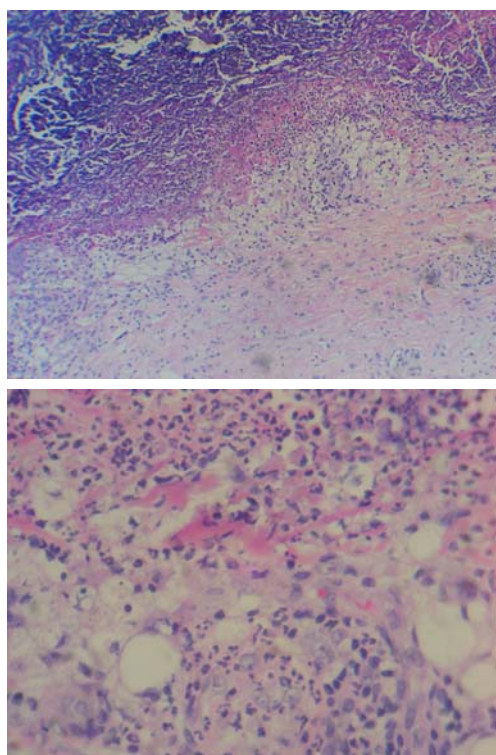


Figure 2.1 , 2.2 Biopsy specimen showed superficial ulceration with dense neutrophilic infiltration in the entire dermis. No leucocytoclasia or evidence of vasculitis was seen

Biopsy specimen showed superficial ulceration with dense neutrophilic infiltration in the entire dermis. No leucocytoclasia or evidence of vasculitis was seen (Figure 2.1, 2.2). The bacterial culture was positive for *Staphylococcus epidermidis*. Laboratory findings showed a positive rheumatoid factor 49.9 IU/cc (0 - 15) and negative antinuclear antibody. The white blood cell count was $11.7 \times 10^3 / \mu\text{l}$ with mild elevated erythrocyte sedimentation rate. The patient was treated with two weeks of oral cloxacillin 2 g/day which led to mild improvement of the lesions. But after replacing the treatment with the combination of topical steroid and colchicines 1.2 mg/day, the condition was better controlled.

Case 2

A 63-year-old woman with history of rheumatoid arthritis for 7 years was well-controlled with sulindac 200 mg/day, chloroquine 250 mg/day, methotrexate 12.5 mg/week. She developed pruritic, mild tender erythematous plaque at left elbow (Figure 3). She had no arthralgia or active arthritis at the time of presentation. Clinical examination revealed indurated erythematous plaques on left elbow and few erythematous papules at right elbow. Biopsy specimen showed diffuse neutrophilic infiltrate in the dermis without any evidence of vasculitis. The patient received colchicine 1.2 – 1.8 mg/day resulting in gradual resolution of the lesions.



Figure 3 Mild tender erythematous plaque at left elbow.

Discussion

Rheumatoid arthritis is a systemic inflammatory autoimmune disease that is characterized by a chronic, symmetric polyarthritis and a significant extraarticular manifestation. The cutaneous manifestations appearing in rheumatoid arthritis included rheumatoid nodules, rheumatoid vasculitis, leg ulcers and pyoderma gangrenosum. Less commonly, palisaded neutrophilic granulomatous dermatitis and interstitial granulomatous dermatitis.

Rheumatoid neutrophilic dermatitis was first described as a rare cutaneous manifestation that occurs in patients with severe rheumatoid arthritis by Ackerman in 1978.¹ Fewer than 20 patients with RND have been previously reported thus far. RND usually occurs in patient with severe seropositive rheumatoid arthritis. RND can also less commonly occur in seronegative patients.²⁻⁴

The lesion of RND are variable, but most commonly presented with erythematous or yellowish papules, plaques, nodules, urticarial-like, crusted, ulcerated or rarely vesicubullae.² The lesions are usually symmetrically located on extensor surfaces of the extremities and trunk.

In addition, RND can be found in the juvenile form of rheumatoid arthritis.⁵ The resolution occurs spontaneously or with improvement of the underlying RA. The exact etiology of RND remained unknown, but the possibility of immune complex activation, cell adhesion and migration and cytokine release, especially IL-8, has been proposed.⁶

Histologically, RND is characterized by a dense dermal neutrophilic infiltrate without vasculitis. In some cases, leucocytoclasia or basophilic degeneration of collagen may be seen. Spongiotic intraepidermal blisters, subepidermal bullae with neutrophils, and/or neutrophilic papillary microabscess have also been reported.⁷ Negative results of direct immunofluorescence to IgG, IgA, IgM, C3 and fibrinogen have been found in RND lesions.⁸

Another neutrophilic dermatosis that may be confused clinically and histologically with RND is Sweet's syndrome. It is characterized by tender erythematous plaques and nodules on face, trunk, extremities, fever, arthralgia and leucocytosis. Histologic findings include a dense dermal neutrophilic infiltrate, prominent leucocytoclasia,

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and the absence of vasculitis but tend to have significant papillary edema leading to the formation of subepidermal blisters.

Another neutrophilic dermatosis associated with rheumatoid arthritis that must be differentiated from RND is erythema elevatum diutinum (EED). It presents with chronic erythematous to brownish infiltrated plaques and nodules on the extensor of extremities. Histologically, in contrast to RND, it has a dense perivascular infiltrate predominantly neutrophils and nuclear dust and vasculitis should usually be seen.⁹

Regarding the treatment of RND, like the others of neutrophilic dermatoses, dapsone has been found to be the most responsive therapy. Colchicine, hydroxychloroquine, cyclophosphamide, topical or systemic corticosteroids had been trialed with some success. Some authors believe that spontaneous remission will occur with improvement of rheumatoid arthritis.²

We conclude that RND can occur in any cases of rheumatoid arthritis with all levels of severity and

should be considered in any faces of cutaneous lesions such as erythematous / yellowish papules, plaques, nodules, urticarial-like, crusted, ulcerated or even vesiculobullae.

References

1. Ackerman AB. Histologic diagnosis of inflammatory skin diseases: A method by pattern analysis. Philadelphia, Pa: Lea & Febiger 1978; :449-51.
2. Timothy SB, Paul KF, Jyoti BB , Jeffry PC. Rheumatoid neutrophilic dermatitis in a woman with seronegative rheumatoid arthritis. J Am Acad Dermatol 2001; 596-600.
3. Yamamoto T, Ohkubo H, Nishioka K. Rheumatoid neutrophilic dermatitis, rheumatoid papules, and rheumatoid nodules in a patient with seronegative rheumatoid arthritis. J Am Acad Dermatol 2003; 48: 634-5.
4. Defaria D, Kroumpouzou G. Rheumatoid neutrophilic dermatitis as presenting sign of seronegative arthritis. Acta Derm Venereol 2004; 84: 236-7.
5. Hussein M, Qasem A, Mohamed A. Rheumatoid neutrophilic dermatitis. Int J Dermatol 1996; 35: 207-8.
6. Yamamoto T, Ohkubo H, Nishioka K. Rheumatoid neutrophilic dermatitis. Int J Dermatol 1994; 33: 798-800.
7. Lowe L, Kornfeld B, Clayman J, Loren E. Rheumatoid neutrophilic dermatitis. J Cutan Pathol 1992; 19:48-53.
8. Sanchez JL, Cruz A. Rheumatoid neutrophilic dermatitis. J Am Acad Dermatol 1990; 5: 922-5.
9. Mashek HA , Pham CT , Helm TN , Klaus M. Rheumatoid neutrophilic dermatitis. Arch Dermatol 1997;133:757-60.