Case Report

DOI: 10.7860/JCDR/2021/49390.15643

Internal Medicine Section

Rheumatoid Neutrophilic Dermatitis as a Presenting Manifestation of Seronegative Rheumatoid Arthritis

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ABSTRACT

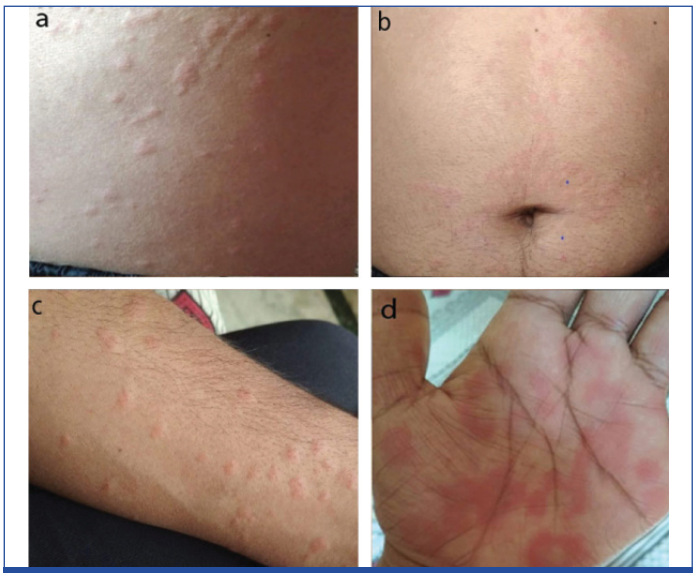
Rheumatoid Neutrophilic Dermatitis (RND) is rare type of skin manifestation in Rheumatoid Arthritis (RA) patients usually seen in long standing seropositive RA. It has very rarely been reported in seronegative RA patients. Here, a case of a young 24-year-old male with clinical features including skin rash over the body, non tender macular lesions on palms, forearms, legs, trunk and pain in joints has been reported. Clinical examination, skin biopsy, raised inflammatory markers and other investigations were suggestive of seronegative RA with neutrophilic dermatitis. The patient was successfully managed with steroids and conventional disease modifying drugs like hydroxychloroquine and methotrexate at three months follow-up period. Thus, RND may involve complications with progressive joint involvement in patients with seronegative RA.

Keywords: Inflammation, Joint pain, Rheumatic disease, Skin manifestation

CASE REPORT

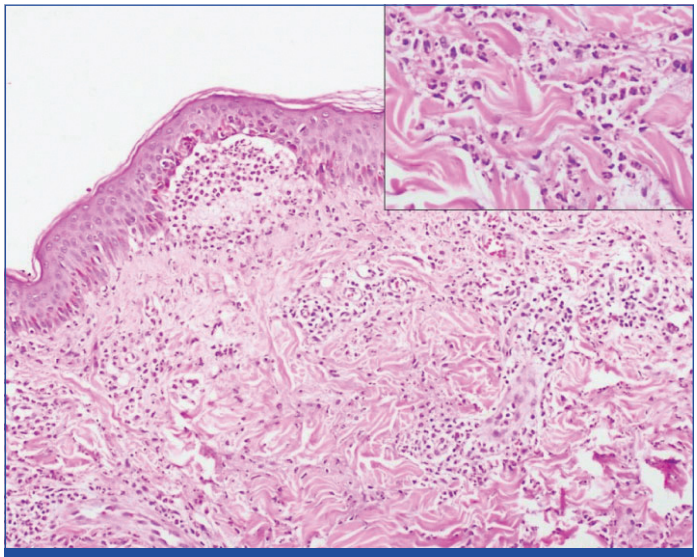
A 24-year-old male patient presented to rheumatology outpatient department with complains of urticarial skin rash all over the body, and symmetric polyarthritis involving hand and knee joints, for past one and half months duration. Skin rash initially started at palm and forearm then rapidly progressed to involve the whole body within a day. There was no history of fever, photosensitivity, alopecia, pain abdomen, loose stool or history suggestive of mononeuritis. He denied any history of drug intake prior to this illness.

On examination, patient had non tender macular lesions [Table/Fig-1] over palms, forearms, legs, and trunk with some scratch marks. Symmetric synovitis of all proximal interphalangeal, metacarpophalangeal, and wrist joints were present. Other systemic examinations were unremarkable. With the present history and physical examination, the differential diagnosis includes urticarial vasculitis, hypocomplementemic urticarial vasculitis, systemic lupus erythematosus or primary systemic small vessel vasculitis.



[Table/Fig-1]: Maculopapular skin lesions over palms, forearms, legs and trunk with some scratch marks.

On evaluation, inflammatory markers were raised {Erythrocyte Sedimentation Rate (ESR) 40 mm in first hour and C-reactive protein-30 mg/L}. Other investigations such as complete blood count, liver function test, renal function test, X-ray chest, urine examination were within normal limits and viral markers for hepatitis-B, hepatitis-C and Human Immunodeficiency Virus (HIV) were also negative. Work-up for autoimmune diseases which were considered as differential diagnosis (Rheumatoid factor, anti-citrullinated protein antibody, anti-nuclear antibody, anti-neutrophil cytoplasmic antibody, C3 and C4 Complement levels) were negative in this case. Skin biopsy was performed from active lesion over forearm which showed diffuse pandermal neutrophilic infiltration with collagen degeneration without any evidence of granuloma or vasculitis [Table/Fig-2]. Direct immunofluorescence was negative for Immunoglobulin A (IgA) or IgG deposition.



[Table/Fig-2]: Photomicrograph of skin biopsy showing pandermal neutrophilic infiltrate (H&E, X100) with leucocytoclasis and collagen degeneration (Inset:(H&E, X400).

The diagnosis of RA was based on the recommendations stated in The American College of Rheumatology (ACR) and the European League Against Rheumatism (EULAR) 2010 [1]. On the basis of history, clinical examination and investigations, patient was diagnosed as seronegative RA with neutrophilic dermatitis.

Patient was managed with tapering dose of steroids and conventional disease modifying drugs {hydroxychloroquine (300 mg once daily) and methotrexate (started with 20 mg per week and then 15 mg per week)}. At three months follow-up, patient had complete resolution of skin rashes as well as arthritis without any recurrence in symptoms.

DISCUSSION

The RA is a chronic inflammatory disease of the synovium involving multiple joints. RND, a type of neutrophilic dermatosis, is a rare skin manifestation of RA which is characterised by dense neutrophilic infiltration of dermis without vasculitis. It is commonly seen in longstanding cases of seropositive RA. However, few case reports have shown association of RND in seronegative RA patients as well [Table/Fig-3] [2-7]. These patients responded well to steroids and conventional disease modified drugs.

Author name	Age/Sex	Skin presentation	RA	Treatment	Response
Gay-Cosier F et al., 2000 [2]	52/F	Isolated erythematous papules on extensor surface of extremities	Since five years	Topical corticosteroids	Responded to treatment
Brown TS et al., 2001 [3]	67/F	Erythematous papules and nodules on extensor surface of thighs and legs	Since 25 years	Dapsone and topical corticosteroids	Discontinuation of dapsone increased the eruptions
Lazarov A et al., 2002 [4]	35/F	Erythematous tender papules on extensor surface of left arm	Since three months	Oral steroids	Disappearance of skin lesion with residual hyperpigmentation
Yamamoto T et al., 2003 [5]	58/F	Subcutaneous and intradermal nodules on elbows, fingers and head	Onset not mentioned	Topical steroid ointment	Skin lesions were alleviated
Defaria D and Kroumpouzos G, 2004 [6]	74/M	Pruritic eruptions on upper trunk and neck	RA symptoms started after appearance of skin lesions	Dapsone	Skin lesions would improve but reoccur on discontinuation of dapsone
Bevin AA et al., 2006 [7]	22/F	Firm nodules over both elbows since six years	Onset not mentioned	Prednisone	Improvement in skin lesions

[Table/Fig-3]: Various case reports of Rheumatoid Neutrophilic Dermatitis (RND) in seronegative Rheumatoid Arthritis (RA) [2-7].

The RA has extra-articular presentation in about 40% cases amongst which the cutaneous manifestations include rheumatic nodules, ulcers, Raynaud's phenomenon and pyoderma gangrenosum [8]. But skin conditions such as neutrophilic lobular panniculitis, granulomatous dermatitis and neutrophilic dermatosis [9] occur in less than 1% cases. RND, a type of neutrophilic dermatosis is a rare skin manifestation of RA which is characterised by dense neutrophilic infiltration of dermis without vasculitis. It is more common in female than male patients having long standing RA and seropositivity [10]. The exact pathogenesis is not clear but it is proposed to be due to immune complex mechanism [11]. RND usually presents as asymptomatic symmetrically distributed erythematous papules, plaques, nodules and urticaria like lesions on the extensor surfaces of the extremities, trunk, shoulders and neck. Lesions like annular, vesicular, bullous, and ulcers are less commonly observed in RND [12].

Other neutrophilic dermatosis seen in RA is Sweet's syndrome, is a close differential of RND. Sweet's syndrome usually presents with variably extensive polymorphous skin eruptions sharply demarcated with a characteristic vesiculation on their surface usually tender on palpation with leucocytosis, fever and malaise [13]. Histopathology shows dense neutrophilic infiltrate in the superficial dermis with massive oedema without vasculitis. In the present case, there was no leucocytosis or fever and even the skin lesions were not tender and did not had characteristic features of sweet's syndrome on histopathology. So, possibility of sweet's syndrome was unlikely. He was presented with urticarial skin lesions with polyarthritis and common differential diagnosis were urticarial vasculitis, hypocomplementemic urticarial vasculitis, systemic lupus erythematosus and primary systemic small vessel vasculitis, but this patient had interestingly turned out to be a case of neutrophilic dermatitis with seronegative RA.

Only six case reports [Table/Fig-3] are there on PubMed search with seronegative RA presenting with RND. The search was done using "RND" and "seronegative" terms without any restrictions and six reports were found to be seronegative. Since it is very rare presentation of RA, it may be challenging to identity the correct diagnosis. Hence, it is important to investigate the patient thoroughly especially with histopathology before starting any treatment. Accurate diagnosis aids in deciding future course of management and also avoids any future complications. Majority of these cases were in females with long history of RA with only one case where skin manifestations appeared first followed by arthralgia [6]. Most of the cases responded to topical steroids, dapsone, prednisone with few cases of flare up of skin lesions on discontinuing treatment [3,6].

CONCLUSION

The RND in a patient with seronegative RA is rarely found. These cases propose that patients with RA can present with RND despite seropositive or negative. Also, there is a need for histopathological evaluation to rule out several skin conditions with similar presentations therein determining the treatment modality.

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PLAGIARISM CHECKING METHODS: [Jain H et al.]
- Plagiarism X-checker: Mar 12, 2021
- Manual Googling: Jul 09, 2021
- iThenticate Software: Oct 25, 2021 (8%)

ETYMOLOGY: Author Origin

AUTHOR DECLARATION:
- Financial or Other Competing Interests: None
- Was informed consent obtained from the subjects involved in the study? Yes
- For any images presented appropriate consent has been obtained from the subjects. Yes

Date of Submission: **Mar 10, 2021**
Date of Peer Review: **Apr 27, 2021**
Date of Acceptance: **Jul 10, 2021**
Date of Publishing: **Nov 01, 2021**