ISSN: 2581 – 3633
PubMed - National Library of Medicine - ID: 101745081

International Journal of Medical Science and Advanced Clinical Research (IJMACR)

Available Online at:www.ijmacr.com

Volume – 4, Issue – 6, November – December - 2021, Page No. : 222 – 225

Rheumatoid Neutrophilic Dermatosis: A Rare Entity

[1]Dr. Namita Bhutani, Department of Pathology, North DMC Medical College & Hindu Rao Hospital, Delhi.

[2]Dr. Kamlesh, Govind Ballabh Pant Institute of Postgraduate Medical Education & Research (GIPMER), Delhi.

[3]Dr. Akhil Nadesan, Department of Pathology, North DMC Medical College & Hindu Rao Hospital, Delhi**.**

Corresponding Author: Dr. Akhil Nadesan, Department of Pathology, North DMC Medical College & Hindu Rao Hospital, Delhi**.**

How to citation this article: Dr. Namita Bhutani, Dr. Kamlesh, Dr. Akhil Nadesan, "Rheumatoid Neutrophilic Dermatosis: A Rare Entity", IJMACR- November – December - 2021, Vol – 4, Issue - 6, P. No. 222 – 225.

Type of Publication: Case Report

Conflicts of Interest: Nil

Abstract

Rheumatoid arthritis (RA) is a chronic inflammatory arthritis disease characterised by systemic involvement with extra-articular manifestations. Rheumatoid nodules are the most frequently encountered skin manifestation. Rheumatoid neutrophilic dermatosis (RND) is the rarest cutaneous manifestation of RA. A 61-year-old female was admitted to the dermatology ward due to erythematous non-blanchable lesions and multiple bullae. Differential diagnosis of RND includes Sweet Syndrome, pyoderma gangrenosum, RND, neutrophilic panniculitis, eccrine hidradenitis,etc. The histological features on skin biopsy were consistent with the diagnosis of rheumatoid neutrophilic dermatosis. Early diagnosis of rheumatoid arthritis related skin entities allows for early management and prevention of cutaneous and systemic complications.

Keywords: RA, ESR, CRP, RND, CCP.

Introduction

Rheumatoid arthritis (RA) affects approximately 1.5 million adults in the USA and 2 million in Europe. In collaboration, the American College of Rheumatology (ACR) and European League Against Rheumatism (EULAR) in 2010 established new criteria for early diagnosis of RA, which included joint involvement, serological markers [e.g., increased serum rheumatoid factor (RF) and anti-cyclic citrullinated peptide antibodies (CCP)], symptoms duration, and acute phase reactants such as C-reactive protein (CRP) and erythrocyte sedimentation rate (ESR).

Rheumatoid arthritis is a chronic inflammatory arthritis disease characterised by systemic involvement with extra-articular manifestations. Rheumatoid nodules are the most frequently encountered skin manifestation.[1]

Other cutaneous manifestations include rheumatoid small-vessel vasculitis, pyoderma gangrenosum, skin ulcers, and Raynaud phenomenon. The incidence of extra-articular manifestations is around 40% in older

people with a high level of rheumatoid factor (RF), smoking, and early disability. Rheumatoid neutrophilic dermatosis (RND) is the rarest cutaneous manifestation of RA. In the literature, only 45 cases of RND are documented.[2]

RND presents frequently with symmetric erythematous papules or plaques, but blisters and vesicles are very uncommon clinical manifestations. The incidence of RND development is high in patients having advanced or long-duration seropositive arthritis.[3]

Here, we present a rare case of RND with atypical unilateral manifestations in an old female patient with known case of RA.

Case report

A 61-year-old female was admitted to the dermatology ward due to erythematous non-blanchable lesions and multiple bullae filled with clear fluid over her left lower leg and foot for one week (figure 1A &1B). She was a known case of advanced RA. A Doppler scan was performed, which revealed decreased blood flow. Other radiological investigations showed erosive arthritis. Morning stiffness and radial deviation of fingers were present as well. The urine examination was positive for albumin. The patient's right leg had been amputated in the past. Her previous medical history was not available. The serological examination was positive for serum rheumatoid factor, CRP (3.5 mg/dl), and ESR (45 mm/hr). The antinuclear antibody test was negative. The provisional diagnosis includes vasculitis and RND. A skin biopsy was taken for confirmation of the diagnosis. Multiple microsections examined from the skin biopsy showed hyperkeratosis and focal areas of orthokeratosis. The epidermis showed spongiosis and dermo-epidermal bullae formation was noticed. The papillary dermis showed a few dilated blood vessels surrounded by a mixed inflammatory infiltrate. Evidence of fibroid necrosis was not seen. The deeper dermis showed dense infiltration by a mixed inflammatory infiltrate, comprising predominantly of neutrophils, eosinophils, lymphocytes, and nuclear dust (figure 2A & 2B). The inflammation was also reaching into the adnexa and subcutaneous adipose tissue. Evidence of vasculitis was absent. Collagen fibers seem to be degenerating. Histological features were consistent with the diagnosis of rheumatoid neutrophilic dermatosis.

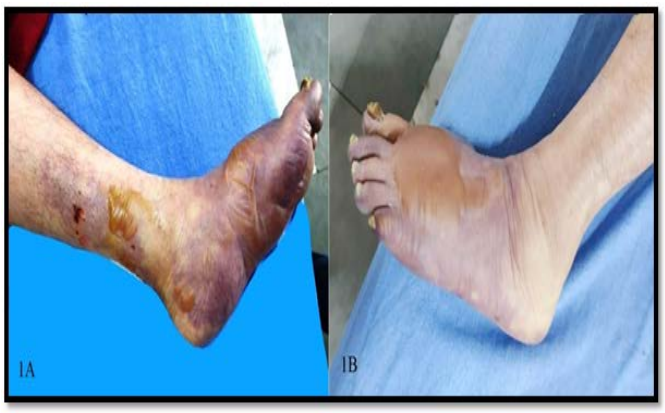

Figure1A&1B: Shows erythematous non-blanchable lesions, and multiple bullae filled-with clear fluid over left lower leg and foot.

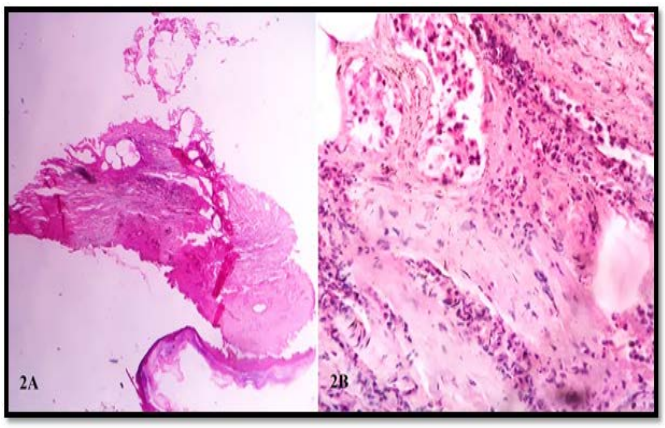

Figure2A&2B: Shows dermo-epidermal bullae (10x). Deeper dermis is infiltrated by dense mixed inflammatory cells comprising predominantly of neutrophils, eosinophils, lymphocytes, and nuclear dust

with adjacent collegen degeneration. Inflammation is extending upto the subcutaneous adipose tissue (100x).

Discussion

RA is a chronic inflammatory joint disease with autoimmune pathogenesis and systemic complications that involve not only the articular system but also the extra-articular system of the body. As the disease progresses, patients end up with systemic complications and destructive polyarthritis. Females have a higher risk of developing RA between the ages of 35-50 than males. Although the incidence of RA is higher among the American and European populations, the disease is not limited to any geographic boundaries, race, or ethnic groups. RA pathogenesis involves immune-related genes, especially HLA-DRB, which is associated with more severity and high mortality.[4]

RA classification criteria given by the ACR/EULAR collaboration include four most important elements: 1) joint involvement, 2) serology, 3) acute phase reactant, and 4) duration of symptoms. RA is a heterogeneous group of disorders, among which cutaneous lesions are the most frequent. Other extra-articular manifestations are ocular, pulmonary, cardiac, renal, neurologic, and hematologic manifestations.[1]

Cutaneous lesions may be categorised as RA-specific and RA-related in RA patients. RA-specific cutaneous lesions are seen only in RA patients among whom rheumatoid nodules are the most common. RA-related skin lesions are seen not only in RA but also in other disorders as well, such as Sweet syndrome, Raynaud's phenomenon, and pyoderma gangrenosum.[5]

Our case report is related to RND, which is a rare entity and was first documented by Ackerman in 1978. It is believed to belong to an immune-complex-mediated disorder, but pathogenesis is yet to be confirmed. Middle- aged females have a twofold higher incidence than their male counterparts.[1]

An RND patient presents with asymptomatic, symmetric, erythematous, and urticarial-like plaques and papules on the extensor aspects of the extremities, neck, and trunk. Vesicle and bullous lesions are very uncommon.[6]

In our case, the patient presented with erythematous plaque-like bullous lesions on the left lower extremity only.Lesions were absent in other parts of the body, such as the thigh, buttocks, or upper extremities. Her right leg was amputated below the knee, the medical history of which was unavailable, and she was unable to convey her past history. Gabriela Zuk et al reported similar case of RND in 65-year-old man.[3] Kumari I et al reported RND case in young female presented over the extremities.[2]

RND belongs to the neutrophilic dermatoses (ND) group characterized by inflammatory skin lesions comprised predominantly of neutrophils, no evidence of vasculitis or infection. ND includes Sweet Syndrome, pyoderma gangrenosum, RND, neutrophilic panniculitis, eccrine hidradenitis, etc.[7]

Hence, it's important to keep these entities in differential diagnosis before arriving at the final diagnosis. RND is typically an asymptomatic symmetric skin lesion and its characteristic histopathologic features are dense dermal neutrophilic infiltration with no evidence of vasculitis. Papillary dermal microabscess is frequently seen. No granular IgA deposits within the dermal papilla on direct immunofluorescence. While Sweet Syndrome is an acute febrile disorder, ND shows an abrupt onset of tender asymmetrical annular skin lesions comprised of superficial dermal edema with diffuse dermal infiltration of neutrophils.[6]

Conclusion

We report a rare entity with an atypical clinical presentation. It will add one more case to the literature, which will help clinicians as well as pathologists narrow down the final diagnosis. Early diagnosis of rheumatoid arthritis related skin entities allows for early management and prevention of cutaneous and systemic complications.

References

1. Tilstra JS, Lienesch DW. Rheumatoid nodules. Dermatologic clinics. 2015 Jul;33:361-71.

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

3. Gabriela Żuk, KamilaJaworecka, DominikSamotij, et al. Rheumatoid neutrophilic dermatitis. Reumatologia 2019;6:350-53.

4. Lora V, Cerroni L, Cota C. Skin manifestations of rheumatoid arthritis. Giornaleitaliano di dermatologia e venereologia: organoufficiale, Societaitaliana di dermatologia e sifilografia. 2018 Jan;153:243-55.

5. Cugno M, Gualtierotti R, Meroni PL, Marzano AV. Inflammatory joint disorders and neutrophilicdermatoses: a comprehensive review. Clinical reviews in allergy & immunology. 2018 Apr;54:269-81.

6. Filosa A, Filosa G. Neutrophilicdermatoses: a broad spectrum of disease. Giornaleitaliano di dermatologia e venereologia: organoufficiale, Societaitaliana di dermatologia e sifilografia. 2018 Jan;153:265-72.

7. Nischal KC, Khopkar U. An approach to the diagnosis of neutrophilicdermatoses: A histopathological perspective. Indian journal of dermatology venereology and leprology. 2007 Jul; 73:222.