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Pregnancy-associated neutrophilic figurate erythema

Sir,

Neutrophilic figurate erythema is a rare inflammatory dermatosis of unknown etiology. It usually presents as annular or polycyclic erythematous lesions with histopathological feature of predominant neutrophilic infiltrate.[1] Herein, we report a case of neutrophilic figurate erythema in a pregnant woman with onset of symptoms in the third trimester and spontaneous resolution after delivery.

A 46-year-old primigravida of 32 weeks gestation presented with a two-week history of itchy skin eruptions on her lower limbs. She was otherwise healthy without fever, chills, malaise or arthralgia. There was no history of insect bite, systemic diseases or new medication usage. She had been treated with topical corticosteroid and oral antihistamine without clinical improvement. Physical examination revealed multiple erythematous and purpuric arciform plaques with peripheral scales on her lower limbs [Figure 1a]. Some of the rashes on her left calf became indurated and tender which later developed pustules [Figure 1b]. Laboratory studies revealed a normal complete blood count, electrolytes, liver and renal function and complements levels (C3 and C4). Antinuclear antibody and anti-extractable nuclear antigen antibodies (Ro/SS-A, La/SS-B, Scl-70 and Jo-1) were negative.

Histopathological examination revealed superficial perivascular and interstitial infiltration of lymphocytes and numerous neutrophils. Papillary dermal edema and erythrocyte extravasation were present without any evidence of vasculitis. Mild epidermal spongiosis and subcorneal pustules were also visible with a negative periodic acid-Schiff stain [Figures 2a and 2b]. Direct immunofluorescence was negative for immunoglobulin G, immunoglobulin A, immunoglobulin M, complement three and fibrinogen deposition. Based on the clinical and histopathological findings, neutrophilic figurate erythema was diagnosed. The patient received oral prednisolone (15 mg/day) and topical fluocinonide cream (0.05%) for two weeks and showed a significant clinical improvement. At 38 weeks of gestation, she gave birth to a healthy baby by cesarean section. The rashes gradually resolved postpartum without further treatment [Figure 3]. No recurrence was observed during one year after delivery.

Neutrophilic figurate erythema is a rarely reported benign annular erythema with a clinical presentation similar to erythema annulare centrifugum. Histopathologically, predominant neutrophilic perivascular and interstitial infiltrate in the upper dermis, lack of vasculitis, and exclusion of other specific entities are crucial for diagnosis.[2] There are no distinct triggers or direct association with major systemic disorders. We found two case reports of this entity associated with hematologic malignancies[3,4] and another two cases induced by medication.[2] Our patient appears to be the first reported case of pregnancy-associated neutrophilic figurate erythema, the pathogenesis of which is presently unclear.

Clinically, various inflammatory and infectious disorders presenting with annular erythema should be differentiated from neutrophilic figurate erythema, including subacute cutaneous lupus erythematosus, annular erythema of

How to cite this article: Kuo YW, Liau JY. Pregnancy-associated neutrophilic figurate erythema. Indian J Dermatol Venereol Leprol 2022;88:99-101.

Received: January, 2021 **Accepted:** April, 2021 **EPub Ahead of Print:** September, 2021 **Published:** December 2021

DOI: 10.25259/IJDVL_52_2021 **PMID:** 34623053

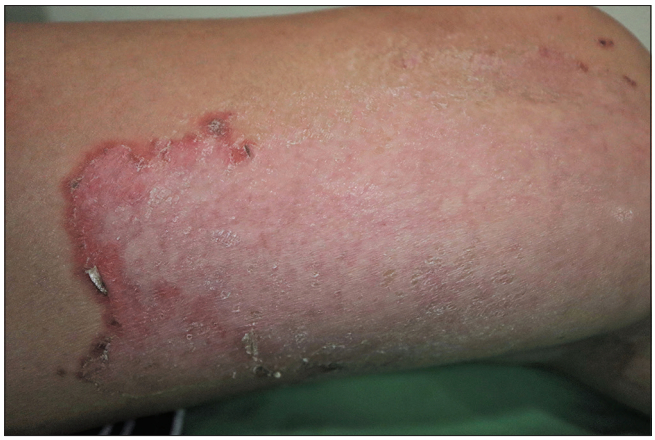

Figure 1a: Erythematous arciform plaques with peripheral scales on the left thigh

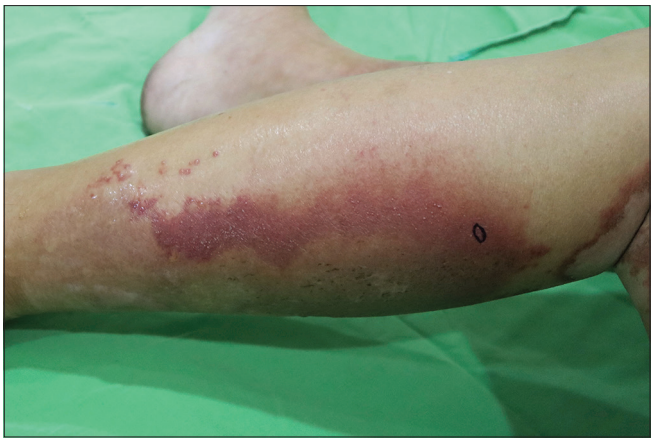

Figure 1b: Plaques with pustules on the left calf. Biopsy site is marked.

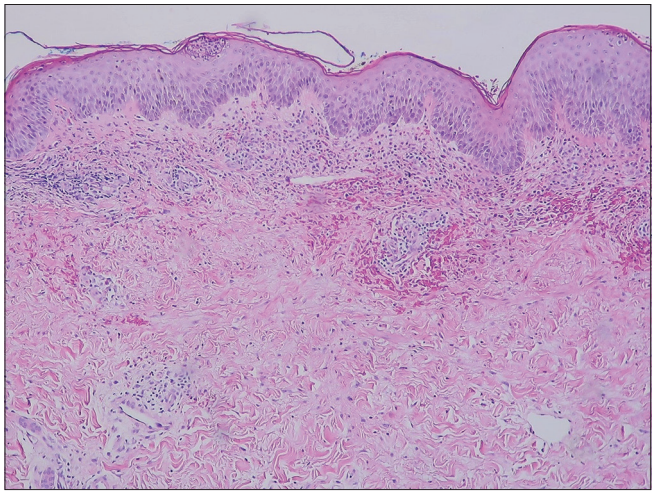

Figure 2a: Subcorneal pustule, perivascular and interstitial infiltrate in the upper dermis with erythrocyte extravasation and mild papillary dermal edema. (hematoxylin and eosin, ×100)

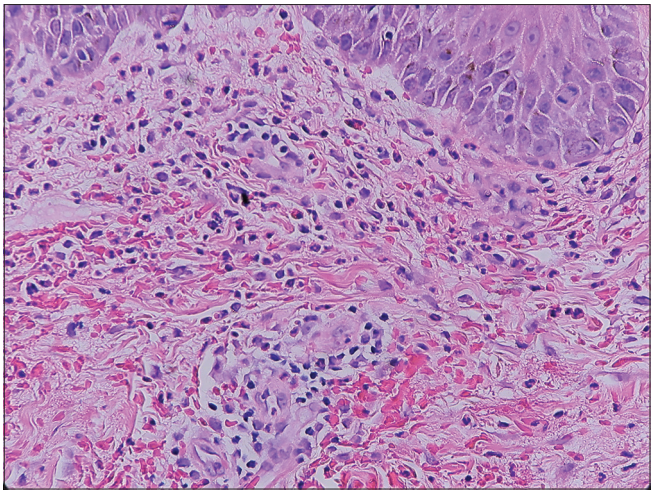

Figure 2b: The dermal infiltrate composed predominantly of neutrophils. (hematoxylin and eosin, ×400)

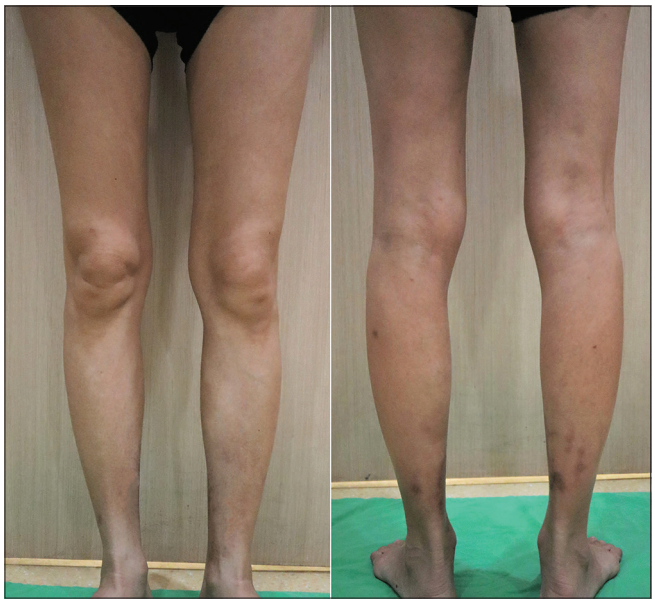

Figure 3: Complete resolution of the rash postpartum

Sjögren's syndrome, erythema annulare centrifugum, granuloma annulare, erythema chronicum migrans and tinea corporis. A correct diagnosis relies on a thorough history taking, laboratory studies and clinico-pathological correlation. Blister formation, purpuric changes and ring-shaped scales were observed in some patients in addition to annular erythema.[2] However, none of the reported cases demonstrated arciform plaques with pustules, as seen in our patient. Impetigo herpetiformis or annular pustular psoriasis may also be considered in this case. Histopathologically, no typical features of psoriasis such as parakeratosis, Munro microabscess, spongiform pustule or dilated capillaries within the dermal papillae were found in our case.

The histological findings of neutrophilic figurate erythema need to be distinguished from other neutrophilic dermatoses. In particular, Sweet syndrome is characterized by more diffuse, deeper and denser neutrophilic infiltrate and prominent papillary dermal edema. Bullous systemic lupus erythematosus is marked by dermal neutrophilic infiltrate

with leukocytoclasia, interface dermatitis and mucin deposition. Urticarial lesions of dermatitis herpetiformis or linear immunoglobulin A dermatosis usually show dermal papillary neutrophilic microabscesses.

A few cases of pregnancy-associated neutrophilic dermatoses have been documented in the literature. An alteration in the immune system during gestation might be a common factor. Pregnant women show a progressive neutrophilia due to increased levels of pro-inflammatory factors (e.g., granulocyte colony-stimulating factor and T helper-17), which may lead to neutrophil hyper-reactivity.[5]

We present a peculiar case of pregnancy-associated neutrophilic figurate erythema with a unique clinical feature of tender arciform erythematous plaques with pustules. While dealing with similar cases, extensive laboratory studies and skin biopsy are required to exclude other serious autoimmune and infectious disorders or associated malignancies. The treatment regimen for neutrophilic dermatoses may be followed to control the symptoms. Since this disease tends to run a benign course, clinicians should weigh benefits and risks before treating pregnant patients.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

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Coexistence of psoriasis and linear IgA disease: An uncommon presentation

Sir,

Psoriasis is a common immune-mediated, autoinflammatory disorder occurring in genetically predisposed individuals. It is seen in 0.4–2.8% of Indian population. Linear IgA disease is a relatively rare dermatoses with an estimated incidence of 0.2–2.3 cases per million per year.[1] It is characterized by the formation of tense vesicles or bullae, often in an annular pattern with blistering along the edge of the lesion – the so-called 'string of pearl' appearance. It is observed in two peaks of different age groups. The first peak usually occurs in six months to six years of age and is known as chronic bullous disease of childhood, while the second peak occurs at around 60 years of age. The coexistence of autoimmune bullous diseases with psoriasis is not uncommon. Most of the reported cases show association with bullous pemphigoid; however, the coexistence of psoriasis with pemphigus vulgaris, pemphigus foliaceous, linear IgA disease, cicatricial pemphigoid, epidermolysis bullosa acquisita and anti p200 pemphigoid is also known.[2] The development of psoriasis in a patient of linear IgA disease is rare in literature and presents a unique therapeutic challenge.

How to cite this article: Neema S, Bhatt S, Kashif AW, Radhakrishnan S. Co-existence of psoriasis and linear IgA disease: An uncommon presentation. Indian J Dermatol Venereol Leprol 2022;88:101-3.

Received: June, 2020 **Accepted:** June, 2021 **EPub Ahead of Print:** September, 2021 **Published:** December 2021

DOI: 10.25259/IJDVL_906_20 **PMID:** 34623057