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Eosinophilic Panniculitis in Wells Syndrome: A Unique Association

Sir,

Wells syndrome (WS) is a rare inflammatory dermatoses first described by Wells in 1971.[1] It is characterized by acute, recurrent, and inflammatory urticarial and cellulitis-like indurated plaques with diffuse tissue eosinophilia, marked edema, and fibrinoid "flame figures."[2] Eosinophilic panniculitis (EP) is a rare type of panniculitis with prominent eosinophilic infiltration of subcutaneous fat.[3] Both are rare entities by themselves; their association is even rarer.

A middle-aged female presented with multiple painful indurated reddish lesions on the abdomen of 2-week duration. There were no systemic symptoms. Past history revealed four similar episodes over a span of 6 months which healed with mild hyperpigmentation. There were no history of arthropod bite, drug intake, and trauma before the onset of symptoms. Dermatological examination revealed multiple solid erythematous deeply indurated infiltrated plaques of varying sizes, with the largest being 6 cm × 4 cm over the right upper quadrant of the abdomen [Figure 1]. Tenderness and the local rise of temperature were present. There was no regional or generalized lymphadenopathy. Laboratory investigations, including hemoglobin and total leukocyte

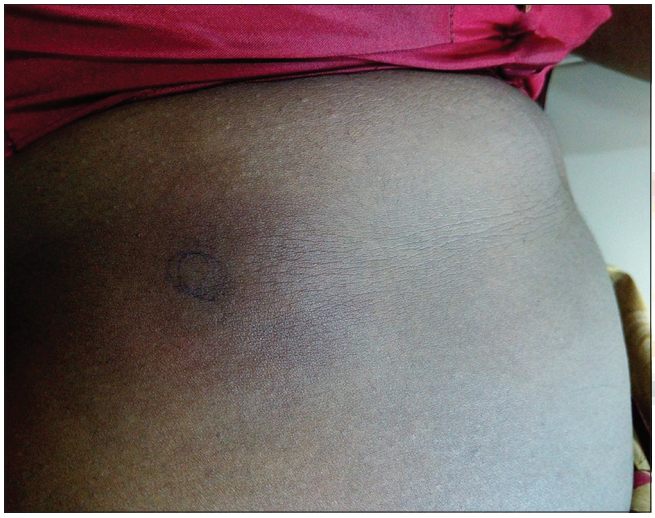

Figure 1: Erythematous indurated infiltrated plaque of size 6 cm × 4 cm over the right upper quadrant of the abdomen

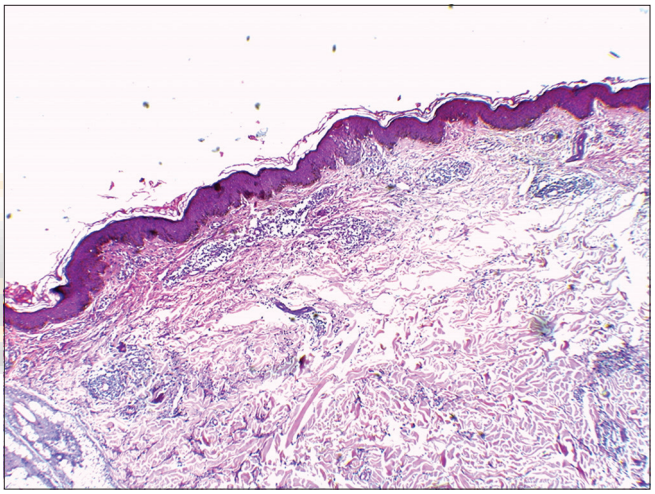

Figure 2: H and E (scanner view): Epidermis – no changes seen

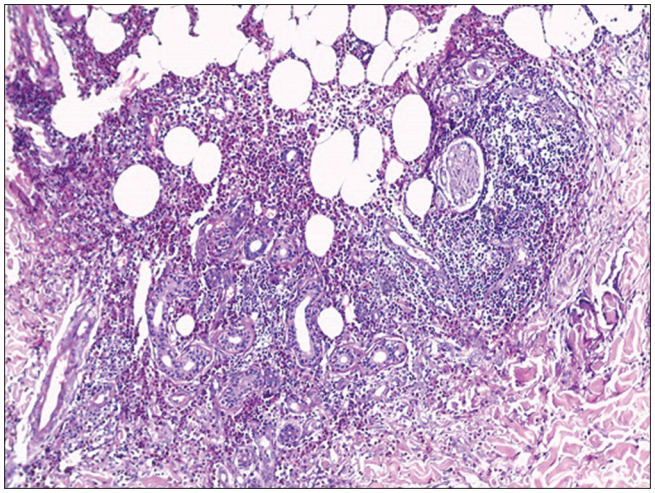

Figure 3: Dermis showing dense inflammatory infiltrate of eosinophil and lymphocytes in perivascular and periadnexal regions (H and E, ×10)

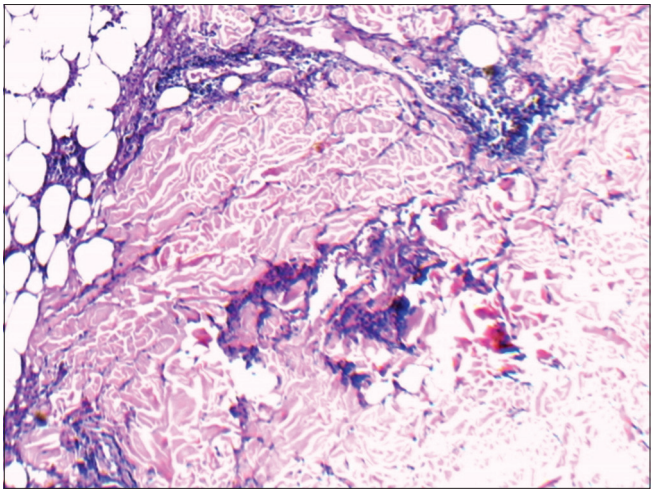

Figure 4: Aggregates of eosinophilic granules seen among dermal collagen fibers (flame figures) (H and E, ×40)

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Table 1: Eosinophilic cellulitis versus eosinophilic panniculitis - a detailed analysis

Features	Wells syndrome (eosinophilic cellulitis)	Eosinophilic panniculitis
First described	George Wells - 1971	Burket and Burket - 1985
Pathogenesis	Increased CD 3 and CD 4 cells causing increased serum and tissue levels of interleukin-5 Eosinophil major basic protein	Increased levels of interleukin-4 and 5 causing altered immune response
Sex predisposition	No predilection	Females: males: 3:1
Clinical features	Edematous infiltrated plaques Often with blister formation followed by slate-gray morphea-like induration	Solitary or multiple nodules, pustules, and plaques
Histopathological characteristics	Early lesions: Dermal edema massive eosinophilic infiltration. Subepidermal blisters containing eosinophils	Lobules and septa infiltrated with eosinophils
	After 1 week: scattered histiocytes and characteristic "flame figures" surrounded by a palisade of histiocytes and multinucleate giant cells	Fat necrosis may be seen
	Panniculitis may or may not be present	No vasculitis
Associations/triggers	Myeloproliferative disorders, infections/infestations (including dermatophytes, viruses, and Toxocara canis, molluscum contagiosum), insect bites or stings, and drugs, HIV infection, eosinophilic fasciitis, Churg-Strauss syndrome, bronchogenic carcinoma, ulcerative colitis, and hypereosinophilic syndrome	Arthropod bite, trauma, Erythema nodosum, immune complex, and leukocytoclastic vasculitis, parasitic infestation, malignant lymphoma, atopy, HIV infection, narcotic dependency with injection granulomas Kimuras disease, hypersensitivity to calcium, heparin, refractory anemia, chronic recurrent parotitis, drug reactions, eosinophilic cellulitis, and immunotherapy with aqueous lyophilized bee venom
Treatment and prognosis	Improves dramatically after administration of systemic corticosteroids Recurrences+	Treatment of the underlying infection/condition Prednisolone and dapsone are tried Recurrences+

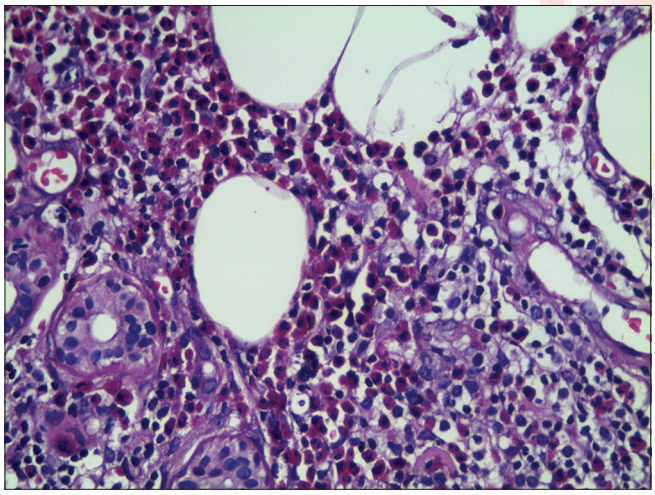

Figure 5: Subcutaneous fat showing dense eosinophilic infiltrate suggestive of eosinophilic panniculitis (H and E, ×40)

counts, absolute eosinophil count, peripheral smear, stool microscopy for parasites, liver and renal parameters, chest X-ray, and serology, were noncontributory. A differential diagnosis of WS and panniculitis was thought and biopsy was done. On histopathology, the epidermal changes were unremarkable [Figure 2]; dermis showed dense inflammatory infiltrate of eosinophil and lymphocytes in perivascular and periadnexal regions [Figure 3]. Aggregates of eosinophilic granules were noted among dermal collagen fibers (flame figures) [Figure 4]. Subcutaneous fat showed dense eosinophilic infiltrate involving both lobules and septa suggestive of EP [Figure 5]. Special stains and polarizing microscopic examination were negative for fungi and other microorganisms.

Based on the clinical presentation of recurrent cellulitis-like infiltrated plaques and the histopathology of dermal eosinophilic infiltrate, flame figures, and lobular and septal eosinophilic infiltrate, a diagnosis of WS with EP was made.

WS was first described by Wells in 1971 as recurrent granulomatous dermatitis with eosinophilia.[1] The term eosinophilic cellulitis was coined later in 1979 when they described eight patients with acute cutaneous infiltrated swellings having a characteristic histology of focal phagocytosis of eosinophilic material in dermis.[2] It is clinically characterized by urticarial or cellulitis-like indurated erythematous plaques. Peripheral blood eosinophilia is seen in 50% of patients whose levels fluctuate during the disease course.[4]

The histology varies according to the stage of the disease. The early phase is characterized by dermal edema and infiltration of dermis with eosinophil followed by subacute stage of "flame figures," and phagocytic histiocytes.[5] The final phase shows fewer eosinophils, histiocytes, and giant cells between collagen bundles. Panniculitis may be seen in WS; however, the co-occurrence of EP is rare. EP was first described by Burket and Burket in 1985 when they observed the same pathologic alterations of eosinophilic cellulitis in the subcutis. The lobules and septa are infiltrated with eosinophils and other inflammatory cells.[6] It may be sporadic or seen along with WS, immune complex vasculitis, atopy, erythema nodosum, psychiatric illness, malignancies, thyroid disease,

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glomerulonephritis, and sarcoidosis. The pathogenesis of both WS and EP is due to dysregulated tissue eosinophilia caused by an increase in CD3+ and CD4+ T lymphocytes. The increased T lymphocytes release interleukin 4 and 5 recruiting eosinophil into the dermis and subcutis. Eosinophil degranulates in the dermis and subcutis causing edema and inflammation.[7]

A comparative analysis between WS and EP is given [Table 1].[7] The treatment of WS includes topical steroids, antihistamines, griseofulvin, phototherapy, systemic steroids, cyclosporine, and dapsone.[5] The treatment of EP is aimed at the underlying or associated clinical condition. The disease in our patient was sporadic and self-limiting.

WS and EP are distinct entities, and their association is rare. As they are associated with many systemic conditions, a thorough workup and evaluation are warranted.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given her consent for her images and other clinical information to be reported in the journal. The patient understands that name and initial will not be published and due efforts will be made to conceal identity, but anonymity cannot be guaranteed.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

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Quick Response Code:

Website: www.ijdpdd.com

DOI: 10.4103/ijdpdd.ijdpdd_38_18

How to cite this article: Gopalakrishnan V, Chandrasekharan J, Prakash AE, Mathew PS, Krishnanand BR. Eosinophilic panniculitis in wells syndrome: A unique association. Indian J Dermatopathol Diagn Dermatol 2018;5:138-40.

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