Eosinophilic Panniculitis in Wells Syndrome: A Unique Association

Sir,

Wells syndrome (WS) is a rare inflammatory dermatoses first described by Wells in 1971. It is characterized by acute, recurrent, and inflammatory urticarial and cellulitis-like indurated plaques with diffuse tissue eosinophilia, marked edema, and fibrinoid “flame figures.” Eosinophilic panniculitis (EP) is a rare type of panniculitis with prominent eosinophilic infiltration of subcutaneous fat. 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...
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,
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.

**REFERENCES**

1. Wells GC. Recurrent granulomatous dermatitis with eosinophilia. Trans St Johns Hosp Dermatol Soc 1971;57:46-56.
2. Wells GC, Smith NP. Eosinophilic cellulitis. Br J Dermatol 1979;100:101-9.
3. Luzar B, Calonje E. Inflammatory diseases of the subcutaneous fat. In: Calonje E, Brenn T, Lazar A, Mckee PH, editors. Mckee’s Pathology of the Skin. 4th ed. Amsterdam: Elsevier Publishing; 2012. p. 326-61.
4. Caputo R, Marzano AV, Vezzoli P, Lunardon L. Wells syndrome in adults and children: A report of 19 cases. Arch Dermatol 2006;142:1157-61.
5. Moossavi M, Mehregan DR. Wells’ syndrome: A clinical and histopathologic review of seven cases. Int J Dermatol 2003;42:62-7.
6. Burket JM, Burket BJ. Eosinophilic panniculitis. J Am Acad Dermatol 1985;12:161-4.
7. Jain S, Jain P, Jakhar P, Shivkumar VB. Eosinophilic panniculitis in a female child: An unusual presentation. Indian Dermatol Online J 2015;6:34-6.

This is an open access journal, and articles are distributed under the terms of the Creative Commons Attribution-NonCommercial-ShareAlike 4.0 License, which allows others to remix, tweak, and build upon the work non-commercially, as long as appropriate credit is given and the new creations are licensed under the identical terms.

**Access this article online**

Quick Response Code:  
Website:  
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.

© 2018 Indian Journal of Dermatopathology and Diagnostic Dermatology | Published by Wolters Kluwer - Medknow