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Wells' Syndrome

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Summary

A 60 years old female patient presented with tender annular erythematous plaques, of two weeks duration, distributed all over the body. Histopathological examination revealed flame figure appearance of perivascular eosinophilic infiltrate.

Introduction

Wells' syndrome was first described by George Wells as a recurrent granulomatous dermatitis with eosinophilia in 1971 [1]. Wells and Smith renamed it eosinophilic cellulitis in 1979 [2]. Wells' syndrome is rare; only about 80 cases have been reported internationally. It usually affects adults, but it has been known to occur in children as well with no race or sexual predilection [3,4]. The disease is often sporadic, but some familial cases have been reported. There are suggested precipitating factors including: arthropod bites, cutaneous viral infections, cutaneous parasitic infestations, myeloproliferative disorders, atopic dermatitis and hypersensitivity reactions to medications [5].

Wells' syndrome usually presents as a tender or mildly pruritic cellulitis-like eruption, occasionally, papular and nodular eruptions may be seen first [6]. The clinical picture can vary widely and may include: annular plaques, urticaria and edema [7] or vesicles and bullae. Bullous Wells' syndrome - are associated with non-Hodgkin lymphoma [8]. Systemic symptoms such as: asthma, arthralgia and fever may occur. Complete resolution of the disease with no scarring is the rule.

Case Presentation

A 60 years old female patient presented with tender annular plaques over the face, trunk and extremities (**fig. 1- 2**). The lesions progressed over two weeks to become large, indurated plaques of edema and erythema, with violaceous edges and no collar. No other lesions of the skin or mucous membrane were found on examination. Routine investigations were done including: complete blood picture, liver function tests, kidney function tests, erythrocyte sedimentation rate and blood sugar level. They were all normal with the exception for mild elevation of leucocytic count and eosinophilia in addition to elevated liver enzymes. Differential diagnosis of this condition included: Wells' disease, granuloma annulare, annular erythema and chronic eosinophilia.





Histopathological examination (**fig. 3- 5**) revealed superficial & deep perivascular infiltrate which was rich in eosinophils giving flame figures appearance. Diagnosis thus was well's syndrome.

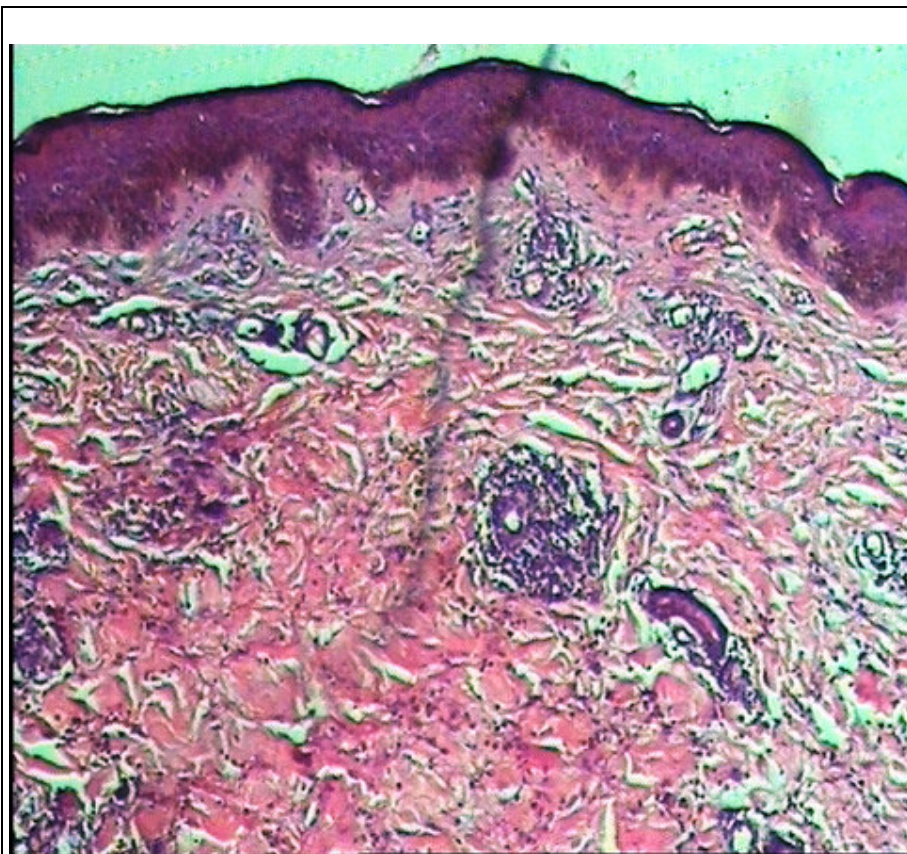


Fig 3: H&E stained sections showing superficial & deep perivascular infiltrate.

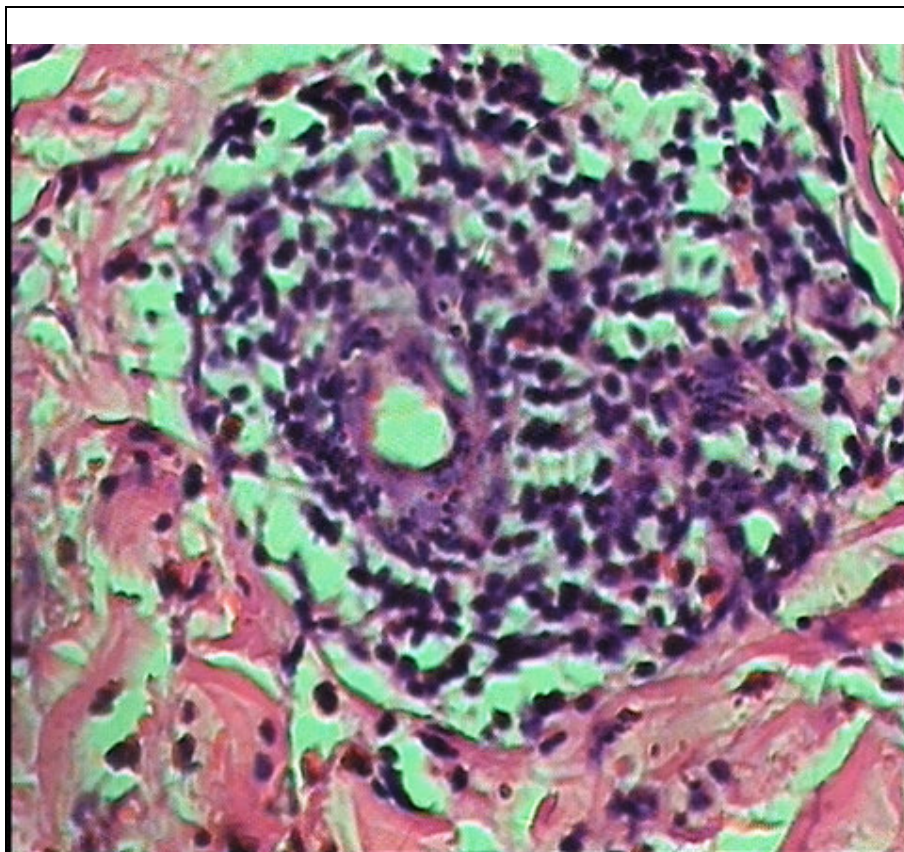


Fig 4: The perivascular infiltrate was rich in eosinophils.

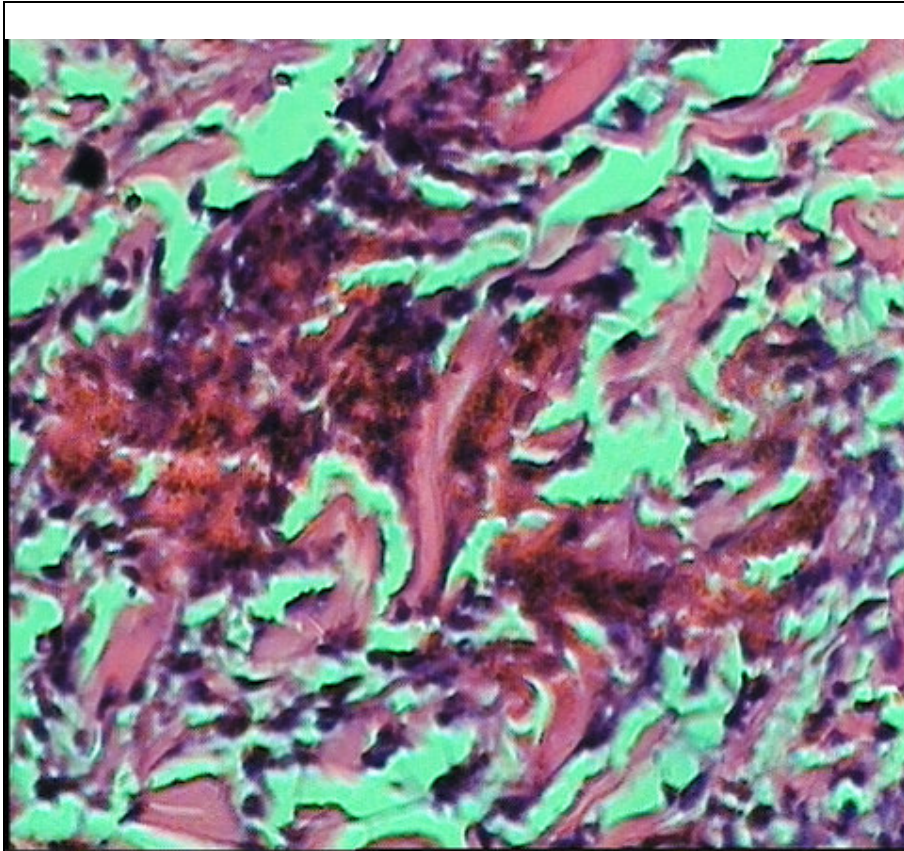


Fig 5: The flame figures characteristic of the syndrome.

Discussion

Eosinophilic cellulitis (Wells' syndrome) is an uncommon condition of unknown etiology. An abnormal or dysregulated eosinophil response appears to be implicated. The presentation usually involves a mildly pruritic or tender cellulitis-like eruption but can take the form of annular plaques as seen in our case with typical histologic features characterized by edema, flame figures, and a marked infiltrate of eosinophils in the dermis [9].

Some authors do not believe that Wells' syndrome is a distinct clinical entity, but rather consider it a histopathologic reaction pattern common to multiple disorders characterised by tissue eosinophilia [10]. Others emphasize that the diagnosis should be reserved for cases with typical features, characteristic histopathology and recurrent course.

Although the histopathologic findings of eosinophilia, histiocytes, and flame figures are characteristic of Wells' disease, they are also found in other conditions, including bullous pemphigoid, eczema, tinea infection, and insect bites [11].

Complete resolution with no scarring is typical, but scarring alopecia may occur in some cases. Although systemic steroids appear to be the only

therapeutic modality of benefit in Wells' syndrome, anti-histaminics and dapsone have been found to be useful.

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