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Well's Syndrome – A Case Report

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Abstract

Well's syndrome is an uncommon condition of unknown etiology, clinically characterized by pruritic or tender cellulitis like eruption and histologically by flame figures in the dermis. We report a case of Well's syndrome in an 18-month-old boy who presented with urticaria-like eruptions.

Keywords: Eosinophilic cellulitis, flame figures, Well's syndrome

Introduction

An 18-month-old boy was brought by his parents with erythematous, oedematous plaques associated with mild scaling all over the body [Figures 1 and 2].

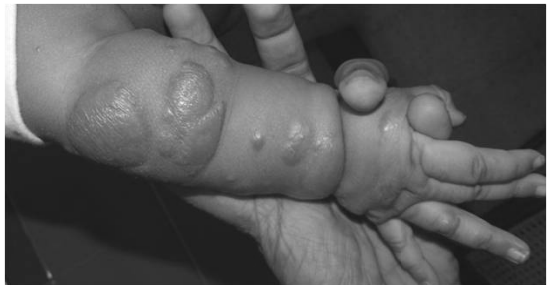

Figure 1: Erythematous, edematous plaques on the forearm of an 18-month-old boy

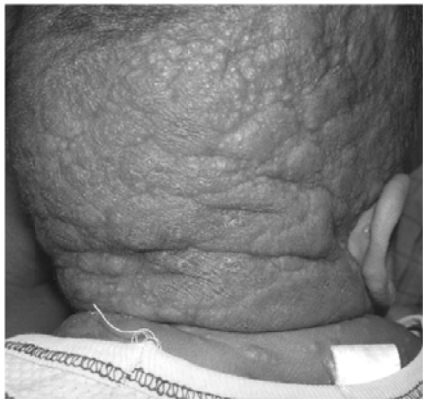

Figure 2: *Urticaria-like plaques associated with mild scaling seen over the scalp*

The skin lesions that started soon after birth were pruritic but not tender. The differential diagnoses considered were urticaria and atopic dermatitis. The child was started on antibiotics and antihistamines with noimprovement. Complete blood count was done, which revealed leucocytosis with eosinophilia. Urine examination and other routine investigations were normal.

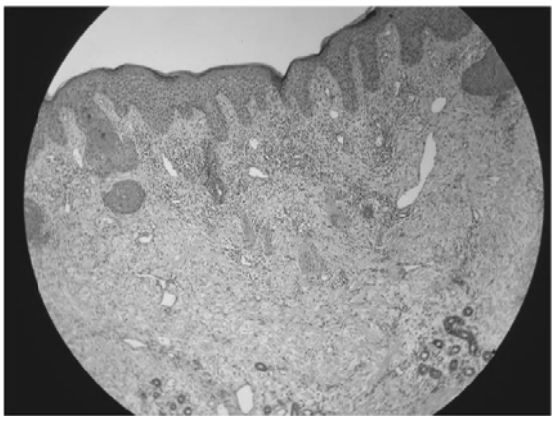

Figure 3: *Histology showing an acanthotic epidermis and dense eosinophilic infiltrate with "flame figures" in the upper dermis (low power view)*

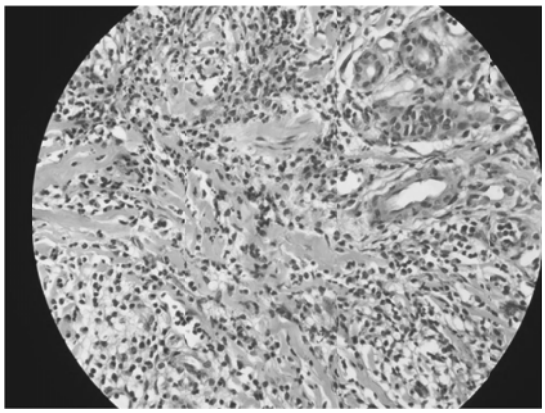

Figure 4: *"Flame figures" are seen in the dermis; they represent intense eosinophilic degranulation of major basic protein coating collagen bundles in the dermis (high power view, magnification ×40)*

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Histology showed an acanthotic epidermis and dense eosinophilic infiltrate with "flame figures" in the upper dermis [Fig. 3 and 4]. On the basis of lesional morphology, peripheral eosinophilia, and cutaneous histopathological features, a diagnosis of Well's syndrome was made, and the patient was started on a short course of systemic steroids to which he responded well.

Diagnosis

WELL'S SYNDROME

Pathogenesis

Well's syndrome is usually considered as a differential diagnosis for any atypical cellulitis or bacterial cellulitis not responding to a course of antibiotics. This condition is rare in children and classically presents with erythematous, oedematous plaques. Other clinical variants that have been documented include urticaria like granuloma annulare, papulovesicular, bullous, nodular, and fixed drug eruption-like lesions.[2]

Description/ Clinical Picture

Well's syndrome[1] or eosinophilic cellulitis is a rare inflammatory disorder of unknown etiology, clinically characterized by two phases. The first phase begins as erythematous, centrifugally spreading plaques, usually following a prodrome of burning sensation and pain. They later evolved into the second phase in 1–3 weeks, characterized by induration, which resolves with morphea like skin atrophy or hyperpigmentation, by 4–8 weeks. Systemic involvement is rare, but fever, arthralgia,[3] lymphadenopathy, and anterior uveitis[4] have been reported. The exact etiology is unknown, but evidence suggests a hypersensitivity response to various triggering factors like insect bites, drugs, infection, inflammatory disorders, thiomersal containing vaccines, malignancy, or idiopathic. Congenital[5,6] and familial cases[3] have also been reported in the literature. Histopathological findings involve an acute stage, subacute stage, and resolving stage.

The acute stage is characterized by dermal edema and an eosinophilic infiltrate. "Flame figures" are seen in the subacute stage; they represent intense eosinophilic degranulation of major basic protein coating collagen bundles in the dermis. In the resolving stage, phagocytic histiocytes palisade around the flame figures, and a granuloma may form in an attempt to eliminate the flame figures. These flame figures are not specific to eosinophilic cellulitis and occur in other conditions, such as bullous pemphigoid, tinea pedis, or insect bite reactions. The peripheral eosinophilia correlates with increased serum interleukin-5 (IL-5) and eosinophilic cationic proteins seen in the majority of patients. IL-5 is responsible for mobilizing eosinophils from the bone marrow, degranulation, and ultimately tissue destruction.[7]

Management

Patients respond well to a course of oral steroids, Prednisolone, at a dosage of 2 mg/kg/day, gradually tapered over 2 to 3 weeks.[7] In recalcitrant cases, a combination of prednisolone, dapsone, and antihistamines,[8] minocycline,[9] PUVA therapy[10] and cyclosporine has been found to be effective.

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