Urticaria multiforme in a 2-year-old girl

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ABSTRACT

We describe a 2-year-old girl who presented to the emergency department for a 2-day history of a rapidly spreading, pruritic, erythematous rash. There were large polycyclic annular erythematous wheals on the face, trunk, and extremities. Urticaria multiforme was diagnosed. Systemic antihistamine therapy greatly regressed the dermal lesions in 24 hours. Urticaria multiforme, a benign cutaneous hypersensitivity reaction, is commonly misdiagnosed as erythema multiforme or a serum sickness–like reaction. Our case highlights the importance of differentiating urticaria multiforme to prevent unnecessary tests and provide proper treatment.

KEYWORDS Cutaneous hypersensitivity reaction; urticaria multiforme

CASE DESCRIPTION

A 2-year-old girl presented to the emergency department on day 2 of a pruritic, erythematous rash. The rash had rapidly spread from her face to her legs and trunk and she had developed a fever, with a high of 103°F, within the last 24 hours. Two weeks previously she had been diagnosed by a rapid strep test with streptococcal pharyngitis, and 5 days prior to presentation she had completed a 10-day course of amoxicillin. Large polycyclic annular erythematous wheals, a few with dusky centers but no true targets, were present on the face, trunk, and extremities, affecting 50% total body surface area (*Figure 1a*). The palms and soles were spared, and there was no ocular or mucosal involvement. After careful history and physical examination, she was diagnosed with urticaria multiforme. She was subsequently treated with systemic antihistamine therapy. Upon evaluation 24 hours later, the rash had greatly regressed with no further sequelae, therefore confirming the diagnosis of urticaria multiforme (*Figure 1b*).

DISCUSSION

Urticaria multiforme, a distinct morphological subtype of urticaria, is a benign cutaneous hypersensitivity reaction primarily mediated by histamine release characterized by transient erythema and angioedema.^{1,2} Pruritus is an almost ubiquitous finding and systemic symptoms include a mild fever that

Figure 1. Urticaria multiforme involving the trunk and legs (a) upon presentation and (b) 24 hours later. Numerous large, polycyclic, annular, erythematous wheals, a few with dusky centers, almost completely resolved, with a few scattered lesions remaining.

usually resolves within 1 to 3 days of eruption.³ It is observed in the pediatric population, most often in those between 4 months and 4 years old. It is characterized by the presence of erythematous macules, papules, and plaques that quickly coalesce into large, blanchable, annular, polycyclic, violaceous wheals with dusky centers usually affecting the face, trunk, and extremities. In most cases there is also associated acral and facial angioedema. The rash usually persists for 2 to 12 days with full resolution and a favorable response to antihistamines.²

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The diagnosis of urticaria multiforme is largely clinical and can be obtained through a careful history and physical examination.¹ A few hallmarks of the rash are the resolution of individual lesions within 24 hours and the presence of dermatographism, a wheal-and-flare reaction after minor skin trauma. Antecedent viral illnesses, antipyretics, and antibiotics, especially amoxicillin and cephalosporins, have all been listed as culprits.

First termed “acute annular urticaria” in 1997 due to the characteristic polycyclic lesion, the name “urticaria multiforme” was coined in 2007 by Shah et al to differentiate this unique urticarial variant.³ It is commonly misdiagnosed as erythema multiforme due to the impressive annular erythematous plaques in a febrile patient or a serum sickness–like reaction due to the presence of acral angioedema. The lesions of erythema multiforme are true target lesions with skin necrosis, blistering, and significant mucous membrane involvement, all of which are not present in urticaria multiforme. Serum sickness–like reactions can be differentiated by their significant systemic symptoms, including high fevers, arthralgias, and lymphadenopathy. The fleeting nature of the

lesions, dermatographism, and favorable rapid response to antihistamines help to confirm the diagnosis of urticaria multiforme.² Due to the difference in prognoses, workup, and treatment between these three conditions, our case seeks to highlight the importance of differentiating urticaria multiforme in order to prevent unnecessary biopsies and laboratory tests as well as establish an appropriate treatment plan and provide reassurance regarding the benign nature of this condition.

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