CASE REPORT

Urticaria Multiforme

A Case Study

CE 1.0 Contact Hour

Kristi Maynard

ABSTRACT: Urticaria multiforme is a rare, hypersensitivity reaction that occurs in infants and small children. Because of its rare occurrence, many providers are unable to accurately identify the rash and associated symptoms to unnecessary laboratory and histological testing or misdiagnosis. This article highlights the case of an 11-month-old female infant with urticaria multiforme. It explores the identification, diagnosis, and current treatment recommendations.

Key words: Urticaria Multiforme, Pediatric Rash, Hypersensitivity Reaction

CASE REPORT

An 11-month-old female infant with no known medical history presented to the urgent care clinic accompanied by her mother with complaints of a full body rash. The rash began 2 days prior as small, elevated erythematous papules on the torso and back but had since progressed to disseminated, blanchable, erythematous annular lesions on the torso, back, face, bilateral upper, and lower extremities (Figure 1). The palms of the hands and soles of the feet were spared. The upper palate and buccal mucosal surfaces were intact with no evidence of rash. Edema of the face, hands, and feet was apparent. The patient's mother denied fevers, decreased appetite, or evidence of joint pain. The child had no indication of pain but had been attempting to scratch her face and neck. The patient met all developmental and growth milestones and was up-to-date on immunizations. She was currently on Day 8 of a 10-day course of amoxicillin/clavulanate 400 mg twice daily for a recurrent otitis media and has historically tolerated complete courses of amoxicillin and cefdinir without incident. The appearance of the rash was unchanged with oral Benadryl recommended by pediatric on-call.

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MANAGEMENT AND OUTCOME

The patient was treated for urticaria multiforme (UM) based on the appearance of the rash, acroedema, and recent history of otitis media with subsequent antibiotic therapy. The cause was indeterminate because of the patient's overlapping recent illness and current antibiotic use. It was recommended that the patient begin once-daily Zyrtec to control pruritic symptoms. The rash gradually faded with complete resolution of all lesions by Day 10.

DISCUSSION

UM was first described in 1997, previously known as "acute annular urticaria"; it is a rare, benign, cutaneous hypersensitivity reaction that affects infants and small children (Emer, Bernardo, Kovalerchik, & Ahmad, 2013; Samorano et al., 2017; Sempau, Martín-Sáez, Gutiérrez-Rodríguez, & Gutiérrez-Ortega, 2016) The rash is caused by a Type IV hypersensitivity reaction, particularly a misappropriated release of histamine in response to recent viral illness or medications. The hallmark of UM is a disseminated, blanchable, erythematous, annular rash with potential confluence. The rash may be associated with acroedema or edema of the face; mucosal membranes are not affected in UM.

The condition is commonly misdiagnosed as erythema multiforme (EM) or urticarial vasculitis (Shah, Honig, & Yan, 2007). EM, although similar in appearance, is a more serious condition that generally involves the palms and soles of the feet and usually presents with a distinctive central vesiculation of annular lesions. Mucosal lesions may develop and are limited to the oral cavity. Patients may also complain of joint pain and fatigue. EM is commonly associated with herpetic infection, specifically herpes simplex virus (HSV) Type 1 and possibly Type 2 (Lamoreux, Sternbach, & Hsu, 2006), but may also be preceded by viral illness or recent drug therapy. The presentation of EM in relation to HSV is thought to manifest as a result of an immune reaction against HSV antigens after a recent outbreak (Orton, Huff, Tonnesen, & Weston, 1984). EM is categorized by severity; cases are classified as EM minor or EM major depending on presenting symptoms. EM minor is a self-limiting condition

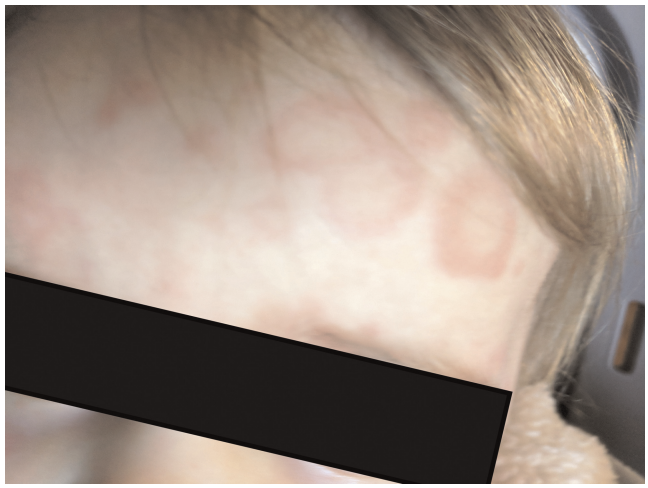

FIGURE 1. Typical presentation of urticaria multiforme.

that will typically resolve in 3–5 weeks but has the possibility of recurrence. Conversely, EM major, also known as Stevens–Johnson syndrome, has the potential to be fatal without prompt, appropriate treatment (Huff, Weston, & Tonnesen, 1983).

Urticarial vasculitis is a recurrent skin rash characterized by pruritic or painful lesions. Lesions may appear on any body surface and are characterized as red patches with central clearing. Petechiae commonly accompany the erythematous rash (Venzor, Lee, & Huston, 2002). Cutaneous manifestations may present concurrently with lymphadenopathy, photosensitivity, joint pain, or fever. Lesions last a minimum of 24 hours and may resolve spontaneously. Urticarial vasculitis is idiopathic in many cases; however, the condition has been linked to viral illnesses, inflammatory connective disorders, and immunoglobulin disorders. A skin biopsy is required for a definitive diagnosis with treatment determined by the identified underlying cause (Griffiths, Barker, Bleiker, Chalmers, & Creamer, 2016).

Accurate diagnosis is dependent on a thorough history and physical examination to differentiate from other conditions. Rarely will laboratory or histological testing be required for diagnosis unless there is a strong suspicion of a more severe pathology. Patients who present with myalgia, high fevers, mucosal involvement, or blistering rash require further evaluation to exclude the possibility of a more severe diagnosis.

In the case of UM, the rash is self-limiting and will gradually resolve within 2–12 days with possible post-inflammatory depigmentation development (Ercan et al., 2017; Madan, Sardana, & Garg, 2015). If a medication is suspected to be the cause of reaction, prompt discontinuation is warranted. Otherwise, treatment is aimed at improving patient comfort as the rash is typically pruritic. Patients may be treated with H_1 antihistamines as appropriate for age and weight to improve pruritus. Although first-generation H_1 antihistamines such as diphenhydramine are effective, they pose a higher risk for the development of anticholinergic side effects including, but not limited to, sedation. For this reason, second-generation H_1 antihistamines such as loratadine (Claritin) or cetirizine (Zyrtec) are preferential as they have little to no anticholinergic effects (Kaplan, 2002). H_2 receptor antagonists such as ranitidine (Zantac) may be considered in addition to H_1 antihistamines for increased symptom control (Harvey, Wegs, & Schocket, 1981). There is no benefit to the use of corticosteroids in this patient population (Madan et al., 2015).

Prompt and accurate recognition of UM is critical in designating an appropriate plan of care. Misdiagnosis of the condition may subject the patient to unnecessary laboratory testing, medication administration, or hospitalization. Although UM may present similarly to other, more severe conditions such as EM or urticarial vasculitis, the provider should be prepared to discern between subtle differences in presentation based on patient history and physical examination. ■

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