Acral papular rash in a 2-year-old boy

Was this rash linked to the patient's recent viral infection?

Fang-Yih Liaw, MD; Ching-Fu Huang, MD; Li-Wei Wu, MD; Chien-Ping Chiang, MD, PhD
Department of Family and Community Health (Drs. Liaw and Wu) and Department of Dermatology (Drs. Huang and Chiang), Tri-Service General Hospital, Taipei, Taiwan

cppchiang@gmail.com

DEPARTMENT EDITOR
Richard P. Usatine, MD
University of Texas Health Science Center at San Antonio

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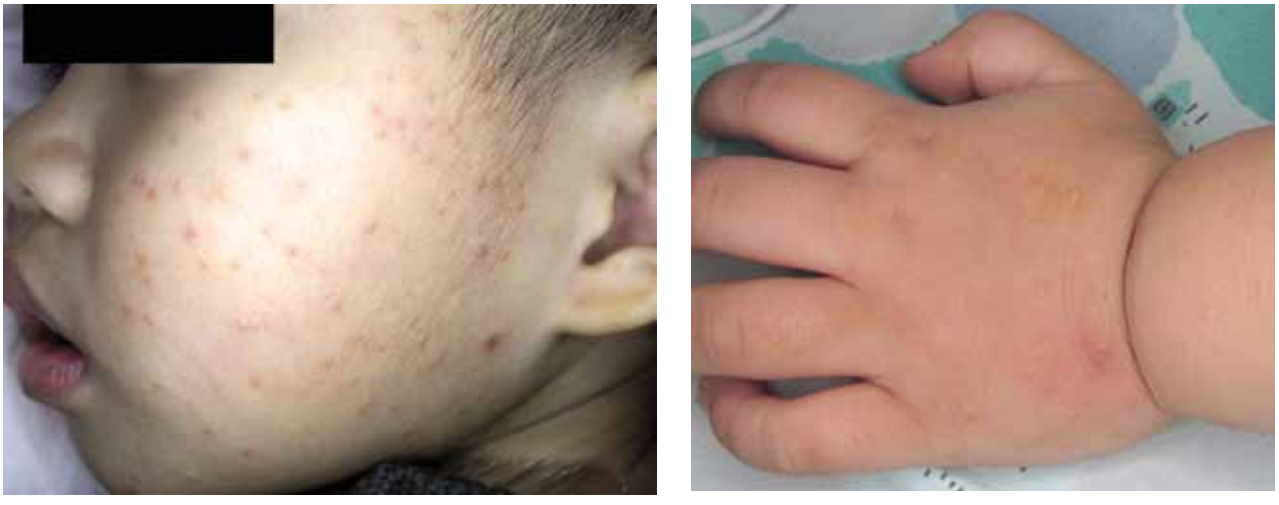
A 2-YEAR-OLD BOY was referred to our outpatient department in the spring with a mild pruritic rash that had appeared on his face, arms, and legs over the previous 2 weeks. His family said that the boy had developed an enteroviral infection the month before. Furthermore, he had a 6-month history of acute myeloid leukemia.

On examination, the child was afebrile, with numerous monomorphous flesh-colored to erythematous papules on his face and on the extensor sites of his limbs (**FIGURE**). However, his trunk, palms, and soles were spared.

- WHAT IS YOUR DIAGNOSIS?
- HOW WOULD YOU TREAT THIS PATIENT?

FIGURE
Flesh-colored to erythematous papules with a monomorphous appearance



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Lesions are usually asymptomatic, but may sometimes be accompanied by a low-grade fever, diarrhea, or malaise.

Diagnosis: Gianotti-Crosti syndrome

Gianotti-Crosti syndrome (GCS) is a pediatric disease whose incidence and prevalence are unknown. Children who have GCS may be given a diagnosis of "nonspecific viral exanthem" or "viral rash" and, as a result, the condition may be underdiagnosed.[1]

GCS—also known as papular acrodermatitis of childhood—affects children between the ages of 6 months and 12 years.[2] The pathogenesis of GCS is unclear, but is believed to involve a cutaneous reaction pattern related to viral and bacterial infections or to vaccination.[3] It is associated with the hepatitis B virus, Epstein-Barr virus, enteroviruses, parainfluenza viruses, and other viral infections. The eruption has also occurred following vaccination (hepatitis A, others).[4]

GCS is usually diagnosed clinically. Physical examination typically shows discrete, monomorphous, flesh-colored or erythematous flat-topped papulovesicles distributed symmetrically on the cheeks and on the extensor surfaces of the extremities and the buttocks. The trunk, palms, and soles are usually spared. This distribution pattern is responsible for the name "papular acrodermatitis of childhood." The lesions are usually asymptomatic, but may be accompanied by a low-grade fever, diarrhea, or malaise.[5]

Avoid confusing GCS with these 4 conditions

The differential diagnosis for GCS includes miliaria rubra, papular urticaria, lichen nitidus, and molluscum contagiosum.

Miliaria rubra ("prickly heat") is caused when keratinous plugs occlude the sweat glands. Retrograde pressure may cause rupture of the sweat duct and leakage of sweat into the surrounding tissue, thereby inducing inflammation. Most cases of miliaria rubra occur in hot and humid conditions. However, infants may develop such eruptions in winter if they are dressed too warmly indoors.[4]

Miliaria rubra manifests as superficial, erythematous, minute papulovesicles with nonfollicular distribution. The lesions, which cause a prickly sensation, are typically localized in flexural regions such as the neck, groin, and axilla, and may be confused with candidiasis or folliculitis.

Cooling by regulation of environmental temperatures and removing excessive clothing can dramatically reduce miliaria rubra. Antipyretics can relieve the symptoms in febrile patients. Topical agents are not recommended because they may exacerbate the skin eruptions.[4]

Papular urticaria predominantly affects children and is caused by allergic hypersensitivity to insect bites.[6] The skin lesions are intensely pruritic and are initially characterized by multiple small erythematous wheals and later progress to pruritic brownish papules.[3] Some lesions may have a central punctum. The patient's age and a history of symmetrically distributed lesions, hypersensitivity, and exposure to animals or insects can help diagnose papular urticaria.[6] Lesions are typically observed on exposed areas, can persist for days or weeks, and usually occur in the summer.[7]

Management of papular urticaria includes the 3 Ps:

- **Protection**. Children should wear protective clothing for outdoor play and use insect repellent.
- **Pruritus control**. Topical high-potency steroids and antihistamines may help with individual lesions, but may be ineffective when the inflammatory process extends to the dermis and the fat.
- **Patience**. Although there is a chance that papular urticaria will be persistent and recurrent, it typically improves with time.[6]

Lichen nitidus is rare and when it does occur, it typically develops in children.[8] Some lesions subside spontaneously, but others may persist for as long as several years.[8]

Lichen nitidus is clinically manifested as asymptomatic, discrete, flesh-colored, shiny, pinpoint-to-pinhead-sized papules; these papules are sharply demarcated and have fine scales. The most commonly affected sites are the genitalia, chest, abdomen, and upper extremities.[8-10] The isomorphic condition called Koebner phenomenon is observed in most cases.[3]

Lichen nitidus is diagnosed on the basis of clinical presentation. Biopsy is indicated only when atypical morphology and distribu-

tion are observed. Histologically, the pathognomonic features of lichen nitidus include focal granulomas containing lymphohistiocytic cells in the papillary dermis.

Lichen nitidus usually regresses spontaneously and most patients do not require intervention; treatment is required only when the patient complains of pruritus or cosmetically undesirable effects.[5] Topical glucocorticoid treatment may provide good results in such cases.[8]

Molluscum contagiosum is caused by poxvirus infection and generally affects young children. It is asymptomatic and presents with small pearly white or pink round/oval papules that may have umbilication. The papules are often 2 to 5 mm in diameter, but can be as large as 3 cm (a giant molluscum). Most papules appear in intertriginous sites such as the groin, axilla, and popliteal fossa.

Transmission occurs by direct mucous membrane or skin contact, leading to autoinoculation.[11] Patients should not share towels or bath water and must avoid swimming in public pools to reduce the risk of spreading the infection.[12]

The individual lesions usually persist for 6 to 9 months but may last for years. Most lesions resolve spontaneously and heal without scarring. Active treatment is used for cosmetic and epidemiologic reasons and includes curettage, cryotherapy, cantharidin, and topical imiquimod.[11] There is no consensus about the dosage and duration in current therapy modalities, but some experts suggest liquid cantharidin (0.7%-0.9%).[3]

Management of GCS? Let it run its course

Most patients with GCS do not need treatment because it is a self-limited benign disease. Although the course is variable and the skin lesions may persist for up to 60 days, the lesions will heal without scarring.[2]

Postinflammation hyperpigmentation or hypopigmentation is rarely seen. In patients who have severe pruritus, topical antipruritic lotions or oral histamines can provide relief.[5] Medium-potency topical steroids may have some benefits, but patients should be closely monitored because there have been reports of exacerbations of lesions with steroid use.[2]

A good outcome. In the case of our patient, the lesions resolved one week after applying 0.1% mometasone furoate cream once a day. JFP

CORRESPONDENCE
Chien-Ping Chiang, MD, Department of Dermatology, Tri-Service General Hospital, No. 325, Sec. 2, Chenggong Road, Neihu District, Taipei City 114, Taiwan R.O.C.; cppchiang@gmail.com

Skin lesions may persist for 5 days to 12 months, although they will heal without scarring.

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