

0196-0644/\$-see front matter

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<http://dx.doi.org/10.1016/j.annemergmed.2013.01.020>



Figure 1. Erythematous crusted vesicles on left cheek.



Figure 3. Scaly symmetric erythematous vesiculobullous buttocks lesions. Used with permission of Corrie E. Chumpitazi, Baylor College of Medicine, Houston, TX.



Figure 2. Erythematous plaque on left foot.

[Ann Emerg Med. 2013;62:303.]

A previously healthy 12-month-old girl presented to the emergency department (ED) with a 10-week history of rash that started as discrete areas of erythematous papules in her oropharynx and lips. This had progressed to symmetric vesiculobullous lesions on her cheek, hands, feet, knees, and diaper area during the past week (Figures 1 to 3). She had low-grade fever and loose stools with flares of the rash, but no other symptoms.

For the diagnosis and teaching points, see page 318.

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DIAGNOSIS:

Acrodermatitis enteropathica. Acrodermatitis enteropathica is a rare disorder of poor zinc absorption linked to an autosomal recessive mutation of zinc transport protein SLC39A4 on chromosome 8q24.3.^{1,2} It classically presents as symmetric periorificial, intertriginous, and acral lesions; diarrhea; and alopecia. Severe forms may lead to failure to thrive, impaired immune function, increased secondary infections, and even death within the first few years of life if untreated. Skin findings mimic atopic dermatitis, with scaly, erythematous, papular plaques, but vesiculobullous lesions have been described.³ Other findings include irritability, anorexia, photophobia, glossitis, and nail changes. Treatment with oral zinc 1 to 2 mg/kg per day normally results in improvement within a few days and resolution of symptoms within a few weeks. Supplementation is lifelong.

The zinc level result was 9 (normal 60 to 120 $\mu\text{g/dL}$), and a skin biopsy revealed intraepidermal vesicles with necrotic keratinocytes without immunofluorescence staining, confirming acrodermatitis enteropathica. This outbreak coincided with the weaning from breast milk. She promptly began receiving oral zinc sulfate and after 2 weeks was noted to have dramatic improvement, with only a mild rash on her left foot.

From the Section of Emergency Medicine, Department of Pediatrics, Baylor College of Medicine, Houston, TX.

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