A breast-fed 4-month-old infant was well until 10 weeks of age, when a rash developed on his right cheek. The rash did not respond to topical steroids or systemic antibiotics, and it spread to his entire face and scalp, as well as to his extremities and patchy areas of the trunk and groin. Decreased weight gain was noted in the last month. There was no history of diarrhea.

On physical examination, the infant appeared well and interactive. Multiple, erythematous, thin, scaly papules and plaques were present on his face, predominantly in the perioral and periorbital areas (Figure 1). Similar lesions were present on the scalp, trunk, and extremities. His genital and gluteal regions revealed markedly erythematous, scaly, eroded plaques (Figure 2).

Laboratory tests performed the results of which were normal included a complete blood cell count; sweat chloride, antinuclear, anti-Ro, and anti-La antibodies; and serum electrolyte levels. The alkaline phosphatase level was low normal, 52 U/L (reference range, 47-191 U/L). Additional laboratory information: the serum zinc level was decreased to 4.0 µmol/L; reference range, 7.7 to 24.5 µmol/L. The maternal serum zinc level was 14.8 µmol/L.
Denouement and Discussion

Acrodermatitis Enteropathica–like Rash in a Breast-fed, Full-term Infant With Zinc Deficiency

Figure 1. Slightly scaly, thin, erythematous papules and plaques on the face, predominantly in a periorbital and perioral distribution.

Figure 2. Eroded, erythematous plaques of the genital and gluteal regions.

Acrodermatitis enteropathica (AE), a disorder in which gastrointestinal absorption of zinc is defective, is inherited in an autosomal recessive fashion. The name was coined by Danbolt and Closs in 1942 to describe the acrally predominant rash present in some patients with diarrhea. The association of AE with zinc deficiency was not discovered until 1974.

Zinc deficiency associated with a similar rash may be the result of a dietary deficiency, malabsorption of zinc, or an abnormal loss of zinc. Zinc-deficient infants typically have diarrhea and are irritable, apathetic, and fail to thrive. Premature infants seem to be at an increased risk for zinc deficiency and may exhibit a negative zinc balance until the third month of life, even under conditions of adequate zinc intake. Factors contributing to zinc deficiency in preterm infants include high zinc requirements and insufficient body stores, possibly related to high urinary excretion of zinc, losses from bony resorption, and immature absorption mechanisms.

Breast-feeding typically protects against zinc deficiency. Breast-fed infants who develop AE frequently do not show symptoms until weaning. One speculation regarding the mechanism of this protective effect is the presence of a zinc-binding ligand, unique to human milk, that increases zinc bioavailability. Symptomatic zinc deficiency has been reported in several breast-fed infants, both premature and full term, suggesting that the protective effect of human milk, once assumed, requires redefinition. It has been suggested that low levels of zinc in the breast milk of mothers of affected infants, despite normal maternal serum zinc levels, are responsible for zinc deficiency in infants, a theory that has been demonstrated to be true in many cases. Theories regarding the cause of low levels of zinc in breast milk include defective mammary zinc secretion or abnormal zinc uptake from plasma by the mammary gland.

Clinical Manifestations

The clinical manifestations of zinc deficiency are similar regardless of the cause. Erythematous, scaly, thin papules and coalescing plaques are distributed predominantly in acral locations, on the face and extremities, and, most prominently, around the oral, orbital, genital, and anal orifices. The rash may also involve the trunk. The skin lesions are often erosive, especially in the periorificial locations. Occasionally, bullous or pustular lesions are present. Alopecia is a common feature. Diarrhea, paronychia, stomatitis, apathy, irritability, and failure to thrive occur with varying frequency.

Abnormal Laboratory Test Results

Affected infants have low serum zinc levels. In breast-fed infants who do not have inherited AE, levels of zinc in maternal breast milk are low. Levels of serum alkaline phosphatase, a zinc-dependent enzyme, are usually decreased.

Treatment

Zinc sulfate, 5 to 10 mg/kg per day by mouth, rapidly reverses the cutaneous lesions. Improvement is noted within days and clearance occurs within 2 to 3 weeks. Stool patterns quickly return to normal after treatment. Parents report that their infants have higher energy levels and appetites shortly after therapy is started. Zinc supplementation in infants who do not have inherited AE is usually unnecessary after weaning.

Differential Diagnosis

The cutaneous findings of zinc deficiency must be differentiated from other nutritional disorders such as essential fatty acid, carboxylase, and biotin deficiencies, isoleucine deficiency seen in some aminoacidurias under treatment, and the AE-like rash associated with cystic fibrosis. The rashes of psoriasis and severe seborrheic dermatitis may have some similar features.

Accepted for publication July 22, 1997.

Reprints: Anthony J. Mancini, MD, Division of Dermatology, 107, Children's Memorial Hospital, 2300 Children's Plaza, Chicago, IL 60614.

References


©1998 American Medical Association. All rights reserved.