

## Case report

# Acrodermatitis enteropathica in a full-term breast-fed infant: case report and literature review

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A male full-term infant, who had been exclusively breast-fed since birth, at 2 months of age developed an erythematous, scaling eruption involving the face (in a periorificial distribution, i.e. mouth, nose, ears, and eyes), hands, and feet, which did not respond to treatment with topical corticosteroids and oral antimicrobials. He was first seen at our institution at 5 months of age (Figs 1 and 2). He had been irritable for the last 2 weeks, but had no diarrhea, alopecia, or anogenital lesions. A clinical diagnosis of acrodermatitis enteropathica was confirmed with a serum zinc level of 41.2 µg/dL (normal, 70–120 µg/dL). His mother had low-normal serum (70.5 µg/dL; normal, 70–120 µg/dL) and normal milk (0.43 µg/mL; normal, 0.2–0.72 µg/mL) zinc concentrations. Within 7 days of starting therapy with zinc sulfate, 10 mg/kg/day, all cutaneous lesions had resolved (Fig. 3).

## Discussion

Zinc is the most important trace element in humans because of its multiple functions (catalytic, structural, and regulatory).<sup>1</sup> Zinc deficiency may be acquired or congenital, as in acrodermatitis enteropathica (AE). AE is a rare autosomal recessive disease leading to severe zinc deficiency, caused by the impaired absorption of zinc in the gastrointestinal tract (duodenum and jejunum).<sup>1,2</sup> The AE gene, *SLC39A4*, localized

in the chromosomal region 8q24.3, encodes the Zip4 zinc transporter.<sup>1,3</sup> The disease is characterized by a triad of acral dermatitis, alopecia, and diarrhea. AE occurs worldwide with an estimated incidence of 1 per 500,000 children, without a predilection for sex or race.<sup>1</sup> Although the first symptoms usually develop within days after birth in bottle-fed infants, or after weaning from breast milk in older infants, it has become apparent that human milk may not always protect against the development of clinical zinc deficiency in premature



**Figure 1** Infant at 5 months of age showing an erythematous, scaling eruption involving the face (periorificial distribution, i.e. mouth, nose, and eyes) and hands



**Figure 2** Infant with dermatophytosis-like lesions



**Figure 3** Infant at 6 months of age after starting oral zinc supplementation. The lesions have resolved

and full-term infants.<sup>2,4,5</sup> The clinical manifestations are erythematous, scaly, erosive, and crusty plaques over acral and periorificial sites. Cutaneous and other manifestations respond dramatically to 1–2 mg/kg/day of elemental zinc given orally.<sup>6</sup> We report one case of AE in a full-term breast-fed infant.

AE is rare in breast-fed infants; it is usually reported in premature babies, because they have insufficient body stores of zinc as well as high zinc requirements. Maternal–fetal zinc transfer occurs predominantly during the last 10 weeks of pregnancy.<sup>1</sup> Full-term infants have adequate stores of zinc gained *in utero* and thus are more likely to maintain a positive zinc balance when breast-fed.<sup>2</sup>

Transient symptomatic zinc deficiency (TSZD) is a self-limited disease mainly observed in breast-fed premature and full-term babies.<sup>5</sup> A low zinc level in maternal milk is an important cofactor. This defect seems to result from a rare abnormality of zinc uptake from plasma by the mammary gland, probably caused by deficiency or malfunction of a zinc binding ligand.<sup>5</sup> A deficiency of zinc secretion of the mammary gland must be suspected when symptoms begin before weaning

in both premature and full-term infants. In these cases, the diagnosis is reached by demonstrating low zinc levels in the mother's milk.<sup>7,8</sup> It should be stressed that a physiologic reduction in the amount of zinc secreted by the mammary gland is normally observed during lactation.<sup>5,8</sup>

Different treatments for TSZD have been applied. It has been reported that 1 mg/kg/day of zinc is sufficient, and that treatment can be stopped shortly after alimentary diversification, without recurrence.<sup>5</sup>

It is important to differentiate AE from TSZD, because AE patients experience a recurrence of symptoms after discontinuation of oral zinc therapy.<sup>3</sup>

Our patient, a breast-fed male infant with acral dermatitis and dermatophytosis-like lesions, had been treated previously with topical drugs. He had no diarrhea or alopecia. His mother presented low–normal serum and normal milk zinc levels. Within 7 days of the start of therapy all cutaneous lesions had resolved. A diagnosis of AE was made and zinc supplementation was given, and will be continued lifelong.

## References

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