

in forms other than plaque psoriasis is scant. Daudén et al.⁷ reported a case of generalized pustular psoriasis that responded with excellent results following treatment with ustekinumab. In a series of 4 patients with palmoplantar pustulosis, Gerdes et al.⁸ reported good response in 1 patient, no response in 2 patients, and a partial response in the fourth. On the basis of these findings, they did not consider ustekinumab to be an appropriate treatment for regular use in this disease. Two other authors recently reported good results with ustekinumab in hyperkeratotic psoriasis in patients similar to ours.^{4,9} From a pathophysiologic standpoint, increased IL-23 expression has been observed not only in plaque psoriasis but also in palmoplantar psoriasis and hyperkeratotic hand dermatitis.¹⁰ Moreover, IL-23 stimulates production of IL-17 and IL-22, and the latter induces epidermal hyperplasia and acanthosis, key pathologic findings in psoriasis.¹¹ Thus, the inhibitory effect of ustekinumab on IL-23 could explain the improvement of palmoplantar hyperkeratosis treated with ustekinumab. However, further studies involving larger numbers of patients are needed to determine whether ustekinumab is useful in this setting.

Conflicts of interest

Dr. Almudena Nuño González, Dr. Enrique Gómez de la Fuente, and Dr. Francisco Javier Vicente Martín declare that they have no conflicts of interest.

Dr. José Luis López Estebanz has taken part in clinical trials and evaluations for Janssen, Abbott, Pfizer, and Schering-Plough.

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Acrodermatitis Enteropathica in a Breast-Fed Infant[☆]

Acrodermatitis enteropática en bebé alimentado con lactancia materna

To the Editor:

Acrodermatitis enteropathica (AE) is a rare autosomal recessively inherited disease caused by alterations in zinc absorption in the digestive tract.^{1–3} Although it was first described in 1942 by Danbolt and Closs,⁴ it was not until 1973 that Moynahan and Barnes⁵ identified its association with low plasma zinc levels. It is estimated to affect 1 in 500 000

children.¹ Clinically, it is characterized by acral and periorificial dermatitis, alopecia, and diarrhea, although this triad of symptoms only occurs in 20% of cases.³ AE usually appears days or weeks after birth in bottle-fed infants, or shortly after discontinuing breastfeeding in exclusively breastfed infants.⁶

Here we describe the case of a full-term male infant who was diagnosed with AE. The only remarkable finding in his history was neonatal indirect hyperbilirubinemia. The patient, who had been exclusively breastfed since birth, developed cutaneous lesions on the scalp, around the mouth, and in the diaper area at 3 months of age. Additionally, his parents reported that he was more irritable at night. He had received various treatments for the cutaneous lesions, including topical and systemic antibiotics, topical antifungal drugs, and topical corticosteroids, but showed no improvement.

Skin examination revealed scaly, erythematous plaques with well-demarcated edges and psoriasiform morphology,

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Fig. 1 Scaly, erythematous plaques on the face (in a periorificial distribution) (A), on the scalp (B), and in the diaper area (C).

and some honey-colored crusts covering zones of erosion. The plaques were located on the face, predominantly in the perioral, perinasal, and frontal areas (Fig. 1A), as well as on the scalp (Fig. 1B) and on the neck, buttocks, and intergluteal cleft (Fig. 1C). Additionally, several isolated plaques with similar characteristics were observed on the thighs and on the dorsal surfaces of fingers and toes.

A serum zinc test showed clearly deficient levels of zinc ($9 \mu\text{g}/\text{dL}$; normal range, $70\text{--}120 \mu\text{g}/\text{dL}$), supporting the suspected diagnosis of AE. Normal zinc levels in maternal milk ruled out an AE-like eruption. After confirming the diagnosis of AE, zinc sulfite treatment was started at a dosage of $3 \text{ mg}/\text{kg}/\text{d}$; this led to rapid resolution of the cutaneous lesions and an improvement in irritability. Currently, after 2 years of treatment, the child shows normal growth parameters for his age, has had no recurrence of the symptoms, and has normal serum zinc levels.

The first symptom is usually acral and periorificial dermatitis and only more advanced cases present the classic triad of alopecia, diarrhea, and skin lesions.³ The disease is characterized by low levels of serum zinc, caused by deficient absorption of this metal in the digestive tract.¹ It is now known that this zinc deficiency is due to a defect in the AE gene (*SLC39A4*) located on the 8q24 chromosome, which encodes Zip4, a zinc transporter in the cells of the small intestine, and primarily in the jejunum.¹

Clinically, scaly erythematous plaques with psoriasiform morphology, distributed symmetrically in acral and periorificial areas, are frequently observed. Other symptoms include vesiculobullous lesions, alopecia, onychodystrophy, onycholysis, pachonychia, stomatitis, cheilitis, blephar-

itis, photophobia, and conjunctivitis. When diagnosis is delayed, patients can develop advanced disease, characterized by increased morbidity and mortality; conditions include growth and mental retardation, irritability, slow wound healing, immunity alterations, hypogonadism, and anemia.¹⁻³

Zinc deficiency can have either a genetic or an acquired origin. The genetic form is classically known as AE and, as mentioned above, is caused by a genetically determined defect in zinc absorption. The acquired forms are clinically similar to classic AE. Three basic causes are known: (a) zinc deficiency in maternal milk,⁷ also known as lactogenic acrodermatitis; (b) a complete lack of zinc in the parenteral nutrition; and (c) the presence of other malabsorption syndromes, such as Crohn disease and celiac disease, in which absorption is compromised, or metabolic disorders associated with a deficit of other trace elements besides zinc.^{2,3}

Diagnosis is primarily clinical, but it is important to acquire data that support the diagnosis (e.g., low serum zinc levels) and to exclude other causes, such as malnutrition, zinc deficiency in maternal milk, and other metabolic alterations.³ While histopathologic findings are not specific, they can be useful for a preliminary diagnosis.

Characteristically, zinc replacement therapy at a dosage of $3 \text{ mg}/\text{kg}/\text{d}$ leads to rapid improvement and resolution of signs and symptoms, substantiating the diagnosis. It is advisable to begin treatment as soon as possible, since delaying it can have severe consequences, including death.⁸

AE usually presents after discontinuation of breastfeeding due to the fact that full-term newborns gain sufficient

zinc stores during the last 10 weeks of pregnancy. Furthermore, maternal milk normally contains sufficient zinc for infant development. However, on rare occasions, AE can also occur in full-term infants who have been exclusively breastfed,⁶ as was the case for our patient. It is important to exclude zinc deficiency in maternal milk in such cases, since this condition requires a much shorter treatment than AE, which must be treated and monitored throughout life.⁹

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