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CASE REPORT

A Case of Acrodermatitis Enteropathica Localized on the Hands and Feet with a Normal Serum Zinc Level

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Acrodermatitis enteropathica is classified as a congenital autosomal recessive type and an acquired transient type. This disease manifests as acral and periorificial dermatitis, alopecia, intractable diarrhea, and failure to thrive. Whereas the autosomal hereditary type is caused by malabsorption of zinc in the intestine, the acquired type is caused by low nutritional support or decreased peripheral release of zinc from blood. We experienced a case of a 5-month old, breast feeding, full-term female presenting with only acral bullous dermatitis without diarrhea, periorificial dermatitis and an abnormal serum zinc level. **(Ann Dermatol 23(S1) S88~S90, 2011)**

-Keywords-
Acrodermatitis enteropathica, Palmoplantar, Zinc

INTRODUCTION

Acrodermatitis enteropathica was first described by Brandt[1] in 1936 and, was named by Danbolt[2]; it is classified as a congenital and acquired type. Characteristic eczematous, bullous cutaneous lesions on periorificial and acral sites with chronic diarrhea, paronychia, alopecia[3], and failure to thrive may be presented, and all of these clinical features are caused by zinc deficiency. In the classic type, the serum zinc level was revealed to be lower than the normal limit; however, a normal serum zinc level has been observed in approximately 30% of all cases[4]. We report on a case of acrodermatitis entreopathica localized on the hands and feet, with a normal serum zinc level, in a breast-feeding full-term baby.

CASE REPORT

The patient was a 5-month old female baby, who presented with bullous lesions on acral sites of the hands and feet. She was born by a normal spontaneous vaginal delivery at IUP 40 weeks and was breast feed. Approximately 2 weeks earlier, she had been treated with antibiotics under the impression of impetigo at a local pediatric clinic, but showed no clinical improvement. The patient did not have diarrhea, and noperiorificial cutaneous lesion was observed. There was no family history of acrodermatits enteropathica or zinc deficiency. Multiple, variable sized bullae and pustules with an erythematous base were observed on acral sites of the hands and feet (Fig. 1). Serum zinc level was as 109.2 μg/dl (normal value: 66.0~110.0 μg/dl), and serum alkaline phosphatase was 69.0 U/L (normal value: under 250.0 U/L), both within normal limits. Specimens obtained from skin biopsy on a bullous lesion on the Lt. 4th toe showed acute spongiotic dermatitis, some hyperkeratosis, upper dermal mild eosinophil infiltration, and cytoplasmic pallor in the upper layer of the epidermis (Fig. 2). After administration of zinc sulfate 6 mg/day orally for one week, the cutaneous lesions showed improvement, and, thereafter, oral zinc sulfate 6 mg/day and zinc oxide ointment were applied for 1 week. All lesions have disappeared and are now under observation.

DISCUSSION

Acrodermatitis enteropathica is a disease related to zinc

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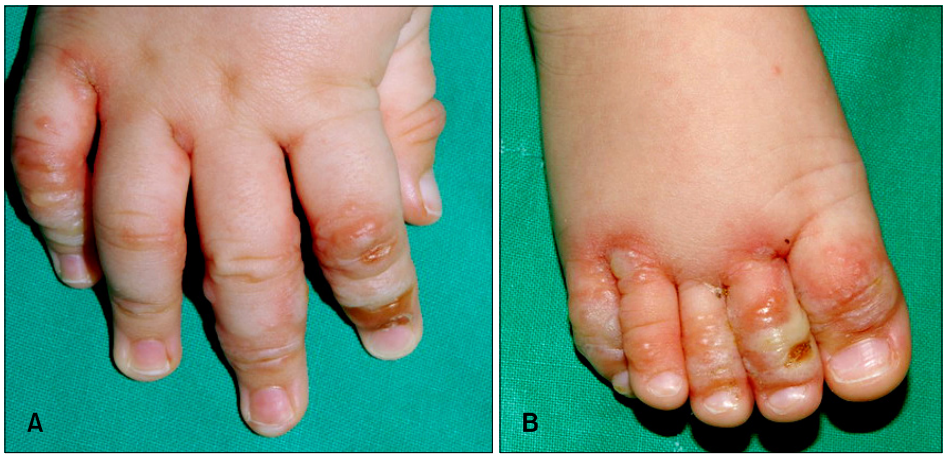

Fig. 1. Multiple tense bullae and vesicles on the distal parts of foot and hand.

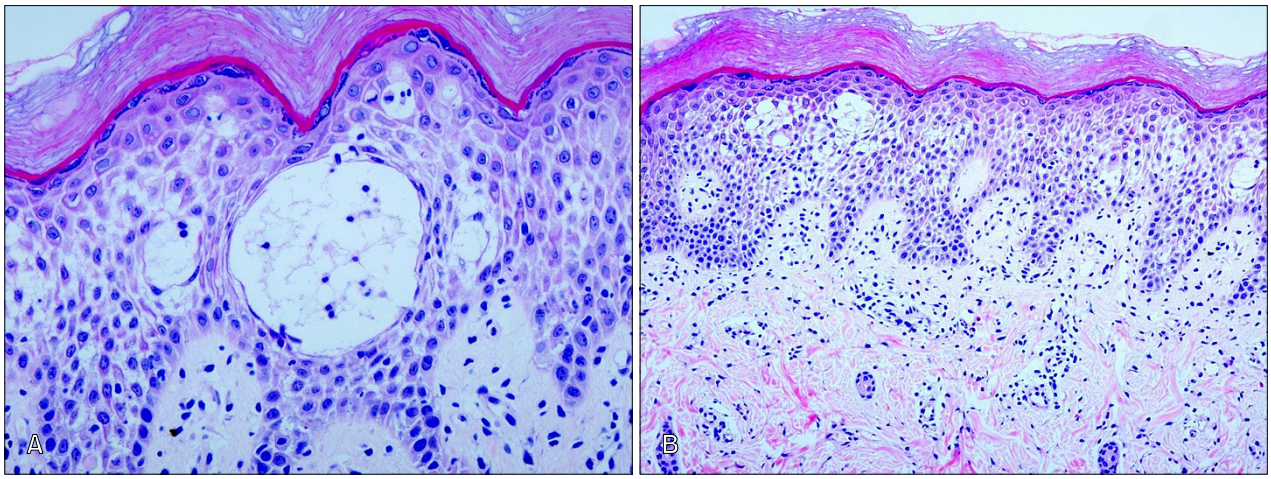

Fig. 2. (A) Acute spongiotic dermatitis and infiltration of upper dermal inflammatory cells (H&E, ×40). (B) Cytoplasmic pallor, the arrow, intraepidermal vesicle formation in the upper epidermis (H&E, ×200).

deficiency caused by an abnormality of intestinal zinc absorption, which is classified according to hereditary congenital type and inherited as an autosomal recessive trait and acquired type. Possible causes of the acquired type include inadequate intake of zinc caused by premarturity or insufficient zinc absorption. It can be further classified according to serum zinc level, with two types: classic acrodermatitis enteripathica with a low serum zinc level and a variant type that presents characteristic clinical features with a normal serum zinc level[5]. In cases diagnosed as acrodermatitis enteropathica, approximately 30% of cases reveal a normal or higher serum zinc level[6,7]. Therefore, the level of serum zinc is not reflected in the actual bioavailability of zinc. Zinc deficiency can occur when zinc is released from blood to a microenvironment where the actual site of zinc biologic action is impaired, even when serum zinc level is normal[8]. Even when the checked serum zinc level is normal, if the albumin bound form of zinc were low, actual zinc deficiency can occur[8]. Diagnosis of acrodermatitis enteropathica is made primarily by clinical manifestations and a low serum zinc level is confirmative. However, if the serum zinc level is normal, with characteristic clinical features and a rapid response to zinc supplementation, the diagnosis can be established[9]. In general, characteristic clinical features of acrodermatitis enteropathica include an eczematous, scaly, or bullous lesion on periorificial areas, such as perioral and perianal and acral sites. In this case report, no periorificial skin lesion was observed.

Presentation of bullous and pustular lesions localized only on the hands and feet is unique. It is believed that the actual zinc level in those site, hands and feet, was lower than the normal level due to a micro environmental problem, such as local abnormal release from blood or a defective transport system. This case differs from other reported cases that revealed typical clinical features,

including characteristic skin lesions and diarrhea, and a normal serum zinc level. In this case, diagnosis was established by a rapid response to the zinc supplement; and, because we were suspicious of acrodermatitits enteropathica, we did not delay treatment. In this case, only bullous and vesicular lesions on acral sites were presented. However, skin biopsy showed findings that were consistent with acrodermatitis enteropathica and the cutaneous lesions had completely disappeared 14 days after treatment with zinc. This is the first reported case where the only presenting clinical features were acral bullous skin lesions, with a normal serum zinc level. This finding indicates that serum zinc level is not an absolute value in diagnosis of acrodermatitis enteropathica. It also means that treatment with a zinc supplement should not be delayed, even if all of the typical symptoms and signs of the classic type of acrodermatitis enteropathica are not presented.

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