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Case Report

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Acquired Acrodermatitis Enteropathica

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Abstract

Introduction: Acrodermatitis enteropathica (AE) is a rare disorder characterized by impaired zinc absorption, leading to severe zinc deficiency. The condition primarily manifests as an autosomal recessive disorder due to mutations in the SLC39A4 gene. However, an acquired form of AE may occur in exclusively breastfed infants due to insufficient maternal dietary zinc levels.

Case Presentation: We report a case of a 6-month-old infant, born at term to consanguineous parents, presenting with erythematous and papulovesicular lesions in the perineum with areas of scaling, stomatitis, and severe paronychia involving all nails. Sparse, brittle hair was also noted. The infant, exclusively breastfed since birth, developed these symptoms at 3 months of age, with progressive worsening. There were no gastrointestinal, nor neurological symptoms. Serum zinc levels were significantly reduced confirming zinc deficiency. Oral zinc supplementation (10 mg/kg/day) resulted in rapid improvement within three weeks.

Discussion: Acrodermatitis enteropathica, in its acquired form, is linked to inadequate maternal zinc intake during lactation. The absence of systemic symptoms in this case suggests localized manifestations of zinc deficiency, consistent with milder forms of the condition. Early recognition of AE based on clinical presentation, combined with serum zinc assessment, is critical to prevent progression to systemic involvement, including immune dysfunction and growth retardation.

Conclusion: Early diagnosis and intervention with oral zinc supplementation are essential to ensure favorable outcomes in acquired acrodermatitis enteropathica. Moreover, maternal nutritional optimization during pregnancy and lactation is pivotal for preventing zinc deficiency-related conditions in infants.

Keywords: Acrodermatitis Enteropathica, Zinc Deficiency, Pediatric Dermatology.

1. Introduction

Acrodermatitis enteropathica (AE) is a rare disorder characterized by severe zinc deficiency. While the condition is most commonly inherited as an autosomal recessive disorder, an acquired form may also arise in exclusively breastfed infants whose mothers have insufficient dietary zinc levels. This is particularly critical during the first months of life when breast milk is the primary source of nutrition, as zinc plays a vital role in numerous biological processes, especially in the development of the skin, nails, and hair [1,2].

Here, we describe the case of a 6-month-old infant who developed classic features of acrodermatitis enteropathica and responded to zinc supplementation.

2. Case Presentation

A 6-month-old female infant, born at term to a first-degree consanguineous couple, presented with an erythematous, erosive rash affecting the perineum, perioral area, and acral regions. The infant had been exclusively breastfed since birth, without any introduction of formula or supplementary nutrition. The rash

had been developing since the age of 3 months, characterized by intermittent flare-ups, but there were no associated gastrointestinal symptoms such as diarrhea or vomiting, nor neurological symptoms like irritability or seizures. There was also no significant family history of similar dermatological conditions or known inherited metabolic disorders.

On clinical examination, the infant exhibited erythematous papulovesicular lesions, which had become erosive in the perioral region (**Figure 1a**), acral areas (**Figure 1b**) and perineum (**Figure 1c**). The lesions were well-demarcated and erythematous, with some areas showing slight scaling. The infant also demonstrated stomatitis and severe paronychia involving all 20 nails (**Figure 1d**). In addition, the infant had sparse, fine, brittle hair (**Figure 1e**). The clinical presentation raised suspicion for acrodermatitis enteropathica, especially considering the absence of any other systemic signs or symptoms that could explain the skin manifestations.

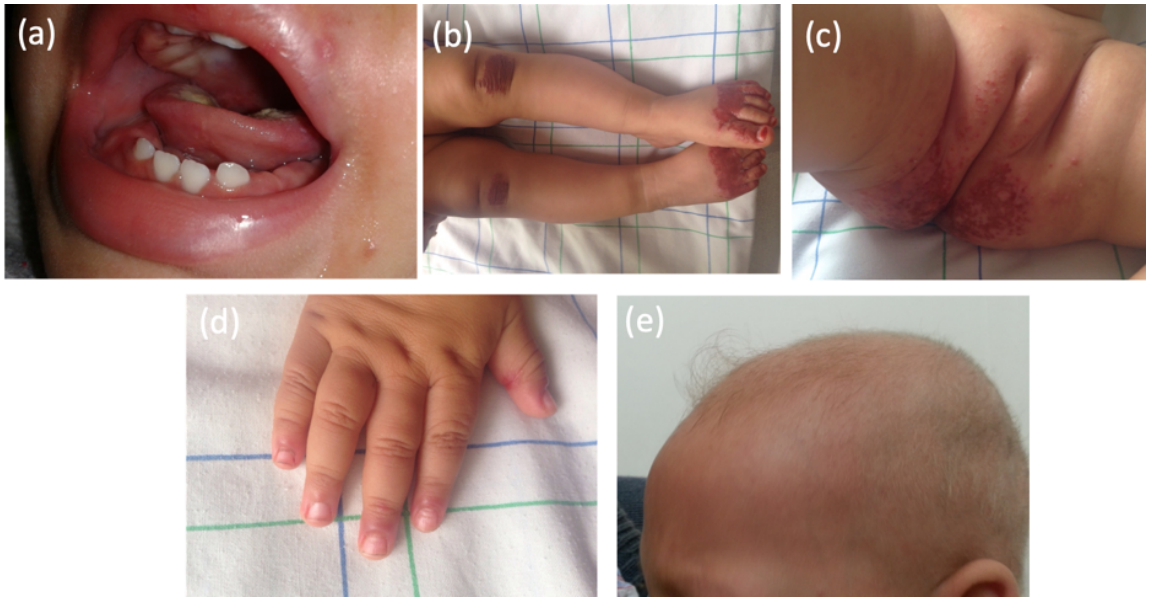

Figure 1: Erythematous Papulovesicular Lesions (a): Perioral Region. (b): Limbs. (c): Perineal Region. (d): Paronychia. (e): Sparse Brittle Hair

Laboratory tests revealed a significantly low serum zinc level of 0.48 mg/L (normal range: 0.72–1.55 mg/L), confirming the diagnosis of zinc deficiency. Serum tests for maternal zinc levels and zinc content in the breast milk were not performed in this case due to the clinical focus on the infant's presentation. A complete blood count was within normal limits, ruling out other systemic causes of the lesions.

Following the diagnosis of acrodermatitis enteropathica, oral zinc supplementation was initiated at a dose of 10 mg/kg/day. Within three weeks of treatment, there was a marked improvement in the skin lesions, with the lesions desquamating and healing (**Figure 2**). The child showed no further progression of symptoms, and overall clinical condition was stabilized.

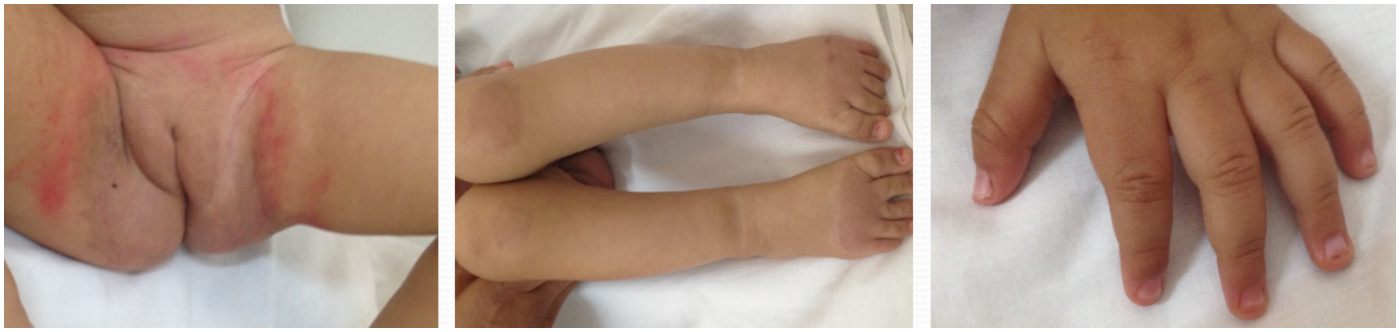

Figure 2: Resolution of Skin Lesions After 3 Weeks of Treatment

3. Discussion

Acrodermatitis enteropathica (AE) is a rare disorder caused by zinc deficiency, which can result from genetic mutations or inadequate dietary intake. While AE is typically inherited as an autosomal recessive condition due to mutations in the SLC39A4 gene resulting in the loss of the zinc transporter, it can also occur in exclusively breastfed infants when maternal zinc intake is insufficient [3,4]. This case highlights the acquired form of AE, emphasizing the critical role of maternal nutrition in preventing zinc deficiency in infants. The classic presentation of acquired AE in our patient, characterized by skin lesions, nail and hair changes, underscores the importance of recognizing zinc deficiency in this context.

The clinical diagnosis of acquired AE was confirmed in our patient through serum zinc testing, which revealed a significantly low level. This is consistent with the literature, where serum zinc levels are typically found to be low in cases of AE [5]. Moreover, although the maternal zinc levels were not evaluated, the infant's rapid recovery upon zinc supplementation strongly suggests that the deficiency originated from inadequate maternal dietary zinc, a hypothesis supported by the absence of other signs suggesting an alternative cause of acquired AE, such as celiac disease or any other absorption disorder [2,6].

Also, the absence of gastrointestinal or neurological symptoms in our patient aligns with the common presentation of acquired AE, which is frequently limited to dermatological involvement. Gastrointestinal symptoms, such as diarrhea and vomiting, and neurological signs, including irritability and developmental delay, are often seen in severe, untreated zinc deficiency [7]. The lack of systemic involvement in our patient further supports the hypothesis that the deficiency was not profound but was sufficient to manifest with skin, nail, and hair changes. This also highlights the necessity of early intervention in preventing the progression to more severe, multisystemic manifestations, which can include immune dysfunction and growth retardation if left untreated [8].

The treatment of AE relies on zinc supplementation, usually with zinc sulfate at 10–20 mg/kg/day. In this case, the infant responded promptly to treatment, with significant improvement in skin lesions, nails, and hair within 3 weeks, demonstrating the efficacy of zinc in restoring epithelial health [9,10].

Our case emphasizes the need for early recognition of AE in infants with unexplained dermatological symptoms, especially those who are exclusively breastfed. AE should be considered in the differential diagnosis of erythematous, erosive lesions in such infants. Additionally, it also underscores the importance of maternal nutritional screening, particularly in populations at risk of zinc deficiency. Preventive strategies, including maternal zinc supplementation during pregnancy and lactation, could help mitigate the risk of AE and other zinc deficiency-related disorders, ensuring better outcomes for both mothers and infants.

4. Conclusion

Acquired acrodermatitis enteropathica is a rare but treatable disorder of zinc deficiency that primarily affects exclusively breastfed infants. Early diagnosis through clinical and laboratory evaluation and prompt treatment with zinc supplementation is essential to ensure a favorable outcome. Clinicians should be aware of the role of maternal nutrition in the prevention of acrodermatitis enteropathica.

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