

Pseudo Acrodermatitis Enteropathica in Bamako: Study of 19 Cases

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Abstract

Original Research Article

Zinc is a co-factor in more than 1000 enzymatic reactions. Zinc deficiency causes cutaneous, digestive and neurological manifestations. Several mechanisms can lead to zinc deficiency. The genetic cause is acrodermatitis enteropathica. Other causes are pseudo enteropathic acrodermatitis. We conducted a 12-month study at the dermatology hospital in Bamako on the epidemiological aspects of pseudo enteropathic acrodermatitis. A total of 19 cases were recruited out of 17212 consultations. The clinical and epidemiological characteristics were consistent with those described in the literature. Quickly improvement was seen in all of our cases under zinc supplementation.

Keywords: Pseudo acrodermatitis, zinc, Bamako.

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INTRODUCTION

Zinc is a cofactor in over 1,000 enzymatic reactions [1]. The skin is the third richest tissue in zinc (Zn) in the body.

It is essential for a number of metabolic processes. Several mechanisms can lead to zinc deficiency.

Zinc deficiency is a major health problem worldwide. It affects 17% of the world's population [2]. The causes of this deficiency can be of several origins: a deficiency in intake, an absorption disorder, a genetic cause, or excessive loss [2]. Severe zinc deficiency results in symptoms such as pustular dermatitis, alopecia, weight loss, diarrhea, infections secondary to immune dysfunction, hypogonadism and ulcer healing problems [3].

Among the various etiologies of zinc deficiency, the most serious is Acrodermatitis enteropathica (AE). This is an autosomal recessive genodermatosis characterized by mutation of the *scl394a* gene. It results in zinc deficiency due to impaired jejunal absorption.

Pseudo enteropathic acrodermatitis includes other etiologies, such as intake deficiency, excessive loss and absorption disorders. In practice, the clinical manifestations of all these forms are similar.

Several factors may be associated with the onset of pseudoacrodermatitis enteropathica. Prematurity, which increases nutrient requirements, intake deficiency or excessive losses [2].

The disease may be confused with kwashiorkor or simple impetigo.

African authors have studied pseudo enteropathic acrodermatitis [4].

We do not know the possible role of nutritional factors on the evolution of the disease in children.

A better understanding of the profile of children with zinc deficiency will help to improve screening and management.

The aim of our work is to describe the epidemiological and clinical aspects of pseudoacrodermatitis enteropathica at the dermatological hospital in Bamako.

II. PATIENT AND METHOD

We conducted a twelve-month study of pseudo enteropathic acrodermatitis at the dermatology hospital in Bamako between October 2018 and September 2019.

We included all children presenting with periorificial erosive dermatitis with low zincemia and/or collapsed alkaline phosphatase.

Parental assent was obtained prior to each inclusion.

The variables used were:

Socio-Demographic Data: Age, sex, duration of disease progression.

Clinical Data: Topography of lesions, diarrhoea, nutritional status, eating habits. Alkaline phosphatase and/or zincemia assays.

Data were analyzed manually.

III. RESULTS

In one year, out of 17212 children with dermatological consultations, we recruited 19 cases of pseudo AE, representing a hospital frequency of 0.11%. Females accounted for 52.63% (10/19). The mean age was 8 months, with extremes of 4 months and 19 months. Cases under one year of age represented 84% of the sample (16/19). Prematurity was reported in 63% (12/19) of our cases. The mean duration of evolution was 2 days, with extremes of 1 month and 15 days. Consanguinity between the parents was found in 57% of cases (11/19). A familial case was found in 26% (5/19). Diarrhea was reported in 75% (15/19).

Mixed breastfeeding was found in 52% (10/19), exclusive breast milk in 47.36% (9/19). Protein-energy malnutrition was found in 63% of cases (12/19).

Lesions were located on the buttocks in 74% (14/19), neck (13/19), perioral region (9/19), groin folds in 42% (8/19).

Alkaline phosphatase was measured in 11 cases (i.e.). They were collapsed in 30% of cases. Zincemia was measured in 3 cases, two of which had collapsed.

Improvement was reported in all cases following zinc supplementation after two weeks.



Figure 1: Erosions, crusts



Figure 2: Erosions on cheeks



Figure 3: Before Treatment



Figure 4: After treatment

III. DISCUSSION

The hospital frequency of 0.11% may be underestimated due to the strictly hospital-based recruitment method. Some authors estimate the proportion of people with acquired zinc deficiency at 17% [5].

Several authors report a frequency of the disease during the weaning period [4, 6]. Prematurity was frequently associated with the disease, as reported by authors [2, 7]. Children born prematurely have increased nutrient requirements.

The role of nutritional factors remains debated in our cases. In fact, all of them were moderately malnourished, and 25% were severely malnourished. Lack of intake has indeed been described as a factor that can induce zinc deficiency.

We found a female predominance, as described by several authors [Ouali, Kury] [6, 7].

We did not carry out genetic testing, but certain factors, such as the consanguinity between parents described in over half the cases, and the presence of familial cases, may suggest enteropathic acrodermatitis. Indeed, the presence of familial cases is usually associated with enteropathic acrodermatitis, as described by certain authors [8].

In our series, diarrhea was present in 79% of cases. Diarrhea and erosive skin lesions are the most common signs reported in zinc deficiency – [8].

Diarrhea is frequently associated with nutritional disorders.

Skin lesions were found on the neck, buttocks and perioral regions. This is the preferred site usually described in the literature for micronutrient deficiencies [6, 9].

Zincemia was performed in 3 of our cases, where it had collapsed. Zincemia is an important diagnostic parameter, and authors agree on a significant titre when it is less than 50% of the normal value [3]. However, several authors report collapsed zinc levels in patients with no clinical lesions [3] or suggestive clinical lesions without zinc deficiency [9].

In the absence of zincemia, alkaline phosphatase assay represents another indirect parameter for diagnostic orientation. In our series, alkaline phosphatase levels were found in 52% of our cases, half of whom had collapsed.

The authors report collapsed alkaline phosphatase levels in some patients with zinc deficiency [7, 10].

The use of alkaline phosphatase as a diagnostic tool remains controversial.

Our cases received zinc supplementation, with clinical improvement of skin lesions and diarrhea within a week. The literature reports the efficacy of zinc in the management of diarrhoea.

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CONCLUSION

The profile of cases of pseudo AE is similar to that reported in the literature.

Further work is needed to differentiate pseudo enteropathic acrodermatitis from enteropathic acrodermatitis, including genetic mutation testing.

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