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Pediatric Acrodermatitis Enteropathica-Report of 4 cases

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Abstract: To report a serie of 4 patients of Acrodermatitis enteropathica which refers to a rare autosomal recessive disorder caused by hereditary and acquired defiency. 4 infants aged between the ages of 4 and 11 Months (a girl and 3 boys) presented around the age of 3 months an erythematous, vesicular and erosive lesions localized mainly on acral and perioral and periorificial areas. Consanguinity was found in two patient .There was no history of prematurity. All four infants were born at full term and received exclusive breastfeeding. There was no digestive or neurological clinical manifestation. The diagnosis was confirmed by the dosage of zinc which was low in our 4 patients and by the therapeutic test of oral zinc intake, a normalized the skin lesions in a few days was achieved. No clinical relapse was observed after oral form of zinc intake. This series reflects a rare inherited condition, characterized by perioficial, perioral and acral dermatitis without digestive symptoms, and should promptly recognized because fatality without appropriate treatment is probable.
Keywords: Acrodermatitis enteropathica, zinc, dermatitis.

INTRODUCTION

Acrodermatitis enteropathica (AE) is a rare autosomal recessive disorder of zinc absorption by the intestine which can be inherited or acquired. Acquired zinc deficiency can be due to insufficient intake, malabsorption, or a combination of these factors.

The classic clinical manifestations of acrodermatitis enteropathica are characterized by a triad of eczematous and erosive dermatitis, acral and perioficial symmetrical dermatitis, alopecia and diarrhea[1]. Angular stomatitis, cheilitis, conjunctivitis, paronychia can also exists[2,3], with the high predisposition to fungal and bacterial infection, which can trigger systemic severe symptoms[4,5]. The estimated prevlance for inherited cases is 1 case per 500,000 population at no race or sex predilection. The age depends on the underlying etiology of acquired or congenital AE.

CASES REPORT

4 infants aged between 4 and 11 Months (one girl and 3 boys) presented around the age of 3 months an erythematous , vesicular and erosive lesions lead to scaly and eczematoid patches localized mainly on acral, perioral, sacral and inguinal areas (figures1,2,3&4).Two boys had nail dystrophy. Consanguinity was found in two patients.

There was no history of prematurity. All these patients had exclusive breastfeeding. All four infants were born at full term. They had no diarrhea, anorexia, photophobia, as well as neither diabetes; neither inflammatory bowel disease, celiac disease nor other digestive or neurological signs were detected.

The average dose of the serum concentrations revealed: zinc=0.48mg/l (0.72-1.55); total protein=6.5 -g/dL (6.0-8.5); alkaline phosphatase=66 UI/L (124-341). Blood count, serum iron, Immunoglobulin (IgG, IgM and IgA),albumin and glycemia were all within normal ranges.

DISCUSSION

Zinc is essential to stabilize cell membranes by reducing free radicals and blocking lipid peroxidation. It is required for normal immunity, fertility and wound healing[6]. It is found in both the epidermis and dermis and aids in cell division and raises keratinocyte proliferation when applied localy to wounds. Zinc deficiency also produces a loss of epidermal Langerhans cells[7].

The gene *SLC39A4*, located in chromosome 8q24.3, was found to encode the transmembrane protein called human zinc/iron-regulated transporter-like protein (hZIP4) required for zinc absorption, which is expressed in the duodenum and jejunum. Its mutation reduces the intestinal ability to absorb dietetic zinc[8,9]. Another cause is expressed by a genetic mutation in a breastfeeding mother, the SLC30A2 gene which codifies a zinc transporter, ZnT2[10]. Which results in sequestration of zinc in the lysosomes of mammary

tissue, necessitating zinc supplementaion. This can analyse our patients totaly breastfed with low intake of zinc which is classified under the inadequate intake of zinc.

Moreover, the absence of the binding ligand in the maternal milk may contribute to zinc malabsorption during brestfeeding.

The standard analysis for the diagnosis of acrodermatitis enteropathica is plasma zinc and the dosage of alkaline phosphatase can be useful. Since it is a zinc-dependent enzyme, it responds to its replacement by increasing the low serum levels observed initially[11]. In our four cases we had low level results in both.

Some cases where associated by diarrhea, anorexia, photophobia, as well as diabetes, inflammatory bowel disease, celiac disease and other digestive or neurological signs were detected[12,13]. Our cases lacked those symptoms and diseases. Otherwise, a nail destrophy was found in some patients as our two candidates[14].

The differential diagnosis of dermatitis enteropathica requires a consideration of a variety of diseases including: Acquired zinc deficiency, atopic dermatitis, cutaneous candidiasis, leucinosis, and malabsorption syndromes secondary to cystic fibrosis or intestinal diseases or acquired immunodeficiency syndrome.

This was a real challenge which required a rapid clinical and biological investigation in our patients to confirm a diagnosis and begin treatment.

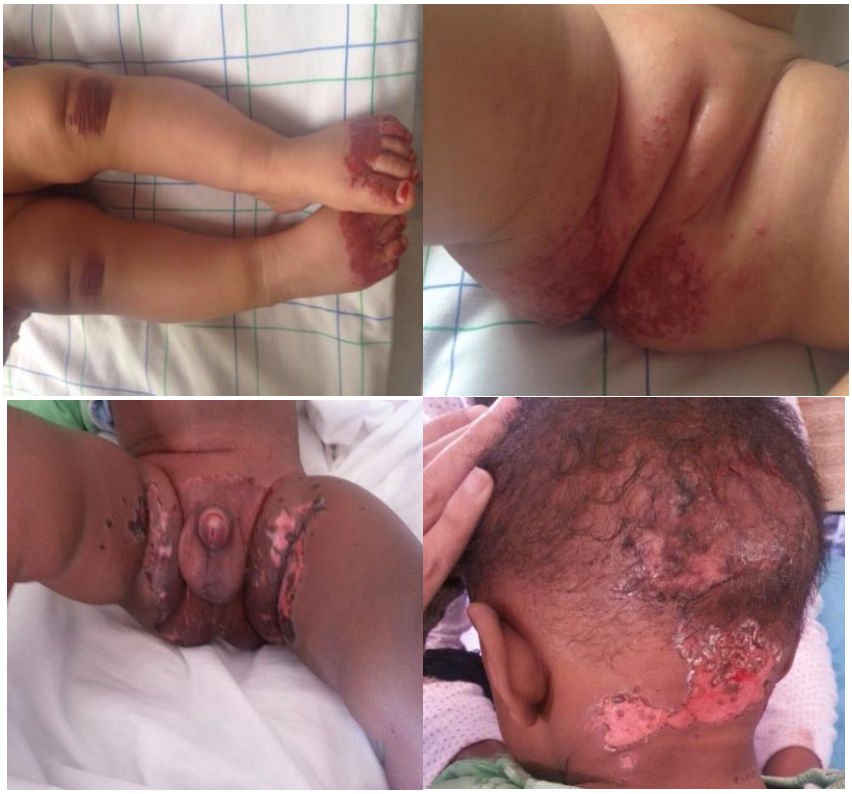

Fig-1-4: Erosive, scaly and eczematoid patches localized on acral, sacral and inguinal areas

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