

Picture stories

Acrodermatitis enteropathica in a 5 month old infant

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Introduction

Acrodermatitis enteropathica (AE) usually appears within the first few postnatal weeks in artificially fed babies or shortly after cessation of breast feeding in breast fed babies¹. It can mimic various conditions including fungal infection¹.

Case report

A 5-month-old infant born at term to non-consanguineous parents with a birth weight of 2.65kg (between 10th to 90th centile) presented with multiple skin lesions since the fourth month of age. The skin lesions appeared initially in the thigh and perianal region followed by lesions on the face He was exclusively breastfed. His weight was 4.8kg (<3rd centile) and length was 62 cm (between 3rd to 50th centile). He had attained developmental milestones age appropriately. There was no family history of similar condition. There was no past history of diarrhoea or recurrent infections.

On examination, skin lesions were erythematous, excoriated and scaly, with periorificial distribution in the perianal and perioral regions. The lesions were also present in the groin, nape of neck and thigh with a localised area of alopecia over the scalp (Figures 1-4).

There were no features of mucosal inflammation or nail changes. A clinical diagnosis of AE was made. Differential diagnoses considered were atopic dermatitis and seborrheic dermatitis. Plasma zinc level was 21.3 µg/dl (Normal range 70-115 µg/dl). The child was started on oral zinc therapy 2 mg/kg daily and showed remarkable recovery within three weeks (Figures 5-7).

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Discussion

Zinc is a cofactor for over 200 enzymes which play a major role in protein metabolism, immune function and carbon dioxide transfer². AE is caused by an autosomal recessive mutation of SLC39A4 gene on chromosome 8q24.3, which determines a congenital partial or total deficiency of the zinc transporter protein zinc-ligand binding protein 4 (ZIP 4)³. This leads to impaired absorption of zinc in the small intestines which causes reduced elemental zinc levels in the blood. Drugs like penicillin, diuretics, valproate, antimetabolites and iron can also produce zinc deficiency⁴. The worldwide incidence of AE is 1 in 500,000 births⁵.

Recurrent skin infections by *Candida* or *Staphylococcus aureus* may occur^{1,6}. Classical triad of skin manifestations, alopecia and diarrhoea occur only in 20% of cases⁷ There may be glossitis, stomatitis, angular cheilitis, blepharitis, conjunctivitis or sparse hair^{5,7}. The condition is often accompanied by severe diarrhoea which can lead to dehydration and electrolyte imbalance⁵. Our patient did not have diarrhoea. Children may exhibit growth delay, hypogonadism and anaemia in severe cases². Kharfi M, *et al*⁸ reported a case of a 4 month old infant who was exclusively breast fed with features of AE, whose mother had low breast milk zinc levels of 46 µg/100 ml. Our patient was an exclusively breast fed infant, so that low zinc levels in breastmilk may have been the reason for the presentation. Analysis of maternal zinc concentration may be helpful. The differential diagnoses to be considered are cutaneous candidiasis, atopic dermatitis and seborrheic dermatitis.

AE has a dramatic response to zinc therapy^{1,5}. Recommended dose of oral zinc is 1-2 mg /kg daily⁶. The various compositions like zinc acetate, zinc gluconate and zinc sulphate can be used for treatment⁶. Children affected severely may be treated with intravenous zinc chloride⁹. Children recover symptomatically prior to normalization of serum zinc levels, diarrhoea ceases in 24 hours, skin lesions start improving in 24 hours, severe skin infections being cured in a week⁵. Following treatment with zinc supplementation, survival rate is 100%⁶. AE requires lifelong treatment with monitoring of serum zinc levels every 6 months¹.



Figure 1: Erythematous skin lesion in genital area



Figure 2: Scaling lesions in nape of neck



Figure 3: Erythematous lesions in buttock



Figure 4: Perioral dermatitis

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Figure 5: Erythematous and scaling lesions in nape of neck and back



Figure 6: Healing perioral dermatitis

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Figure 7: Healing lesions in genital area

Early recognition and treatment of AE is important as complete recovery occurs with zinc supplementation and failure to diagnose can lead to complications such as severe skin lesions and growth disturbances.

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