ACRODERMATITIS ENTEROPATHICA – A CASE REPORT

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ABSTRACT

Acrodermatitis enteropathica is a rare, inherited disorder, transmitted as an autosomal recessive trait. The clinical syndrome is characterized by a phenotypic triad of acral dermatitis, alopecia and diarrhoea. The acral and periorificial distribution of the rash is recognized as a virtually pathognomonic cutaneous marker for zinc deficiency. It may be hereditary or acquired in gastrointestinal disorders or in patients on prolonged total parenteral nutrition. Therapy with zinc gives an excellent clinical response and reduces mortality.

Key words: *acrodermatitis enteropathica, zinc, treatment, case reports.*

INTRODUCTION

Acrodermatitis enteropathica was recognized in 1936, by a Swedish dermatologist Thore Brandt[1]. Zinc absorption in young patients with this entity is low, about 2-3% compared with 27 – 65% in normal adults. The cause of this specific zinc malabsorption is not known. The decreased absorption can be overcome by an oral zinc load but also improves with age[2].

We report here a case of acrodermatitis enteropathica who showed a dramatic clinical improvement to oral zinc therapy.

CASE REPORT

A 3 year old, male, child, born normally at full term to 2nd degree consanguinous parents presented with a generalized, pruritic, eczematous eruption. Birth history was normal. Skin was apparently normal till 3 months of age.

At the time of presentation, cutaneous examination revealed an eczematous eruption on the extremities, front of the trunk and periorificial areas (Fig 1). The back of the trunk was relatively spared. There were erythematous, scaly, crusted plaques on the central part of the face (Fig 2). The palms and soles showed exfoliation (Fig3). A few white plaques were seen on the tongue which were suggestive of candidiasis. There was redness and fissuring at the angles of the mouth. All the finger and toe nails showed dystrophy, Beau's lines and were associated with various degrees of paronychial inflammation(Fig 4). Scalp hair was sparse, thin and light brown in colour.

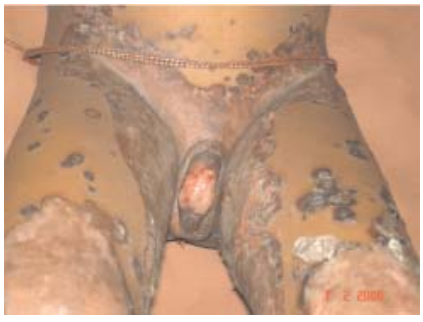

Fig 1. Before treatment.
Crusted lesions on genitalia, thighs

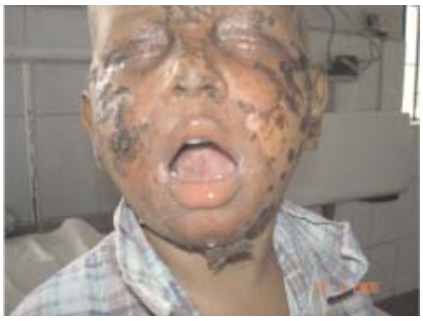

Fig 2. Before treatment.
Erosions and crusts on face

Fig 3. Before treatment.
Widespread erosions on soles, gluteal areas

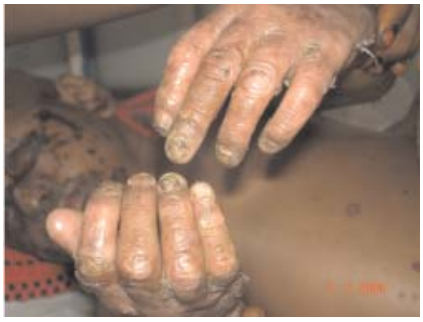

Fig 4. Before treatment.
Extensive nail involvement

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Milestones were normal. He was fully immunized for his age. The child had been exclusively breast fed since birth. He developed intolerance (diarrhoea) to cow's milk when weaned at 3 months of age and was started on infant formula feeds. The first sibling, a male child had died of similar complaints at 6 months of age.

Examination revealed an afebrile, alert but irritable child weighing 12kg. He had a height of 82cm, weight of 12kg, chest, arm and head circumference of 52cm, 15cm and 49cm respectively. Systemic examination was unremarkable. He had average intelligence and linguistic skills. A complete haemogram, liver and renal function tests were normal. Serum albumin was 3.9gm % and serum globulin 4.2gm % with a reversal of the albumin: globulin ratio. Chest X-ray was normal. Ultrasonography of abdomen revealed a polycystic left kidney. Histopathological examination revealed mild intracellular oedema in the upper part of epidermis, parakeratosis, dimunition of granular layer and mild psoriasiform hyperplasia(Fig 7). Urine and stool examination was normal. Alkaline phosphatase enzyme was 76 IU/L Plasma zinc level was low 52ug/dL (60 – 120ug/dL).

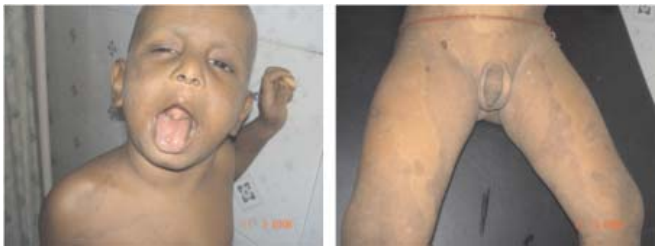

Fig 5. One month after treatment

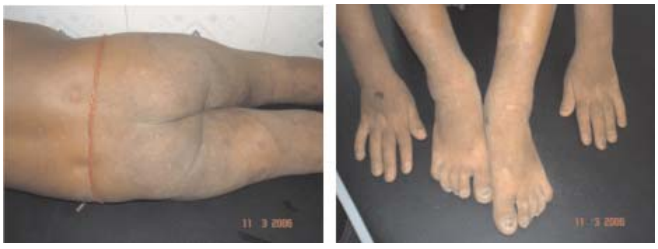

Fig 6. One month after treatment

On the basis of clinical and biochemical findings, we arrived at a diagnosis of acrodermatitis enteropathica. The patient was treated with oral zinc sulphate supplements 2mg/kg/day, topical corticosteroids, warm saline compresses. Oral fluconazole 50mg daily was given for 1 week. There was an overall dramatic response to treatment in 7 days. The child became active and playful by the second day . The skin lesions started healing by the 5th day and completely healed in one month (Fig 5,6)

Discussion:

Acrodermatitis enteropathica is a rare, inherited disorder, transmitted as an autosomal recessive trait. The clinical syndrome is characterized by a phenotypic triad of acral dermatitis, alopecia and diarrhoea. The acral and

Fig 7. Histopathology of skin showing parakeratosis, granular layer dimunition, psoriasiform hyperplasia (H&E, low power).

periorificial distribution of the rash is recognized as a virtually pathognomonic cutaneous marker for zinc deficiency. The cutaneous lesions are psoriasiform or annular, erythematous, scaly, crusted plaques. As the disease progresses these plaques become vesicobullous, pustular and erosive. Other features include stomatitis, apathy, irritability, growth retardation, failure to thrive and delayed wound healing. Delayed puberty and hypogonadism in developing males are some of the long term effects. Classic hereditary acrodermatitis enteropathica begins within days to weeks after birth in infants bottle fed with bovine milk or soon after weaning from the breast in older infants. Acquired zinc deficiency usually occurs in patients receiving prolonged total parenteral nutrition for inflammatory bowel disease and chronic diarrhoea. It may occur in alcoholic liver cirrhosis, pancreatitis, cancer chemotherapy. Acrodermatitis enteropathica needs to be differentiated from other dermatological causes of anogenital rash such as diaper dermatitis, candidiasis, seborrheic dermatitis, psoriasis. Certain other disorders such as essential fatty acid, carboxylase and amino acid deficiencies may have similar cutaneous features and need to be excluded especially if the lesions persist despite zinc supplementation or relapse frequently. Riboflavin deficiency closely mimics this condition but in addition presents with a seborrheic dermatitis like picture, corneal vascularisation and interstitial keratitis. Histopathology of acrodermatitis enteropathica is very non specific and not an absolute criterion to diagnose this condition.

Zinc has been recognised as a constituent of the human body. As it forms an integral part of carbonic anhydrase and many other highly purified enzymes, its importance in protein and carbohydrate metabolism is understandable. Zinc is present in unmilled cereals, beans, cheese, whole wheat bread, animal protein especially lean red meat, pork. Most

of the diets provide 10 – 15 ug of zinc per day (150 – 250umol). High content of phytate and fibre in unleavened bread (chapatti), cereals and vegetables binds the zinc and produces a highly insoluble Zn – calcium – phytate complex that prevents absorption. Leavening of bread with a phytate – splitting enzyme of yeast decreases its phytate content by 15 – 25% and increases zinc absorption. Fats tend to dilute zinc from the total diet. A preadolescent child should receive about 10mg of zinc in the diet per day. The requirement for adolescent male and female are 15mg and 12mg /day respectively[3].

Zinc sulphate for acrodermatitis enteropathica was first introduced in 1973.[4] Oral zinc given in a dose of about 2mg/kg/day was found to cure all clinical manifestations related to zinc deficiency within 1 or 2 weeks. In most instances dietary supplementation with two to three times the RDA in doses of 30 to 55mg of elemental zinc daily is adequate to restore normal zinc status. Dosage is based on the amount of elemental zinc present in the preparation. As observed in a study[5] treatment with zinc sulphate 1mg/kg/day helped clear the symptoms in 5 days. The serum zinc levels which were as low as 6ug% increased to 75ug% after 3 days of zinc therapy. In yet another case report[6], zinc sulphate was used in a dosage of 5mg/kg/day . There was rapid improvement of diarrhoea within 24 hours and the skin lesions within 1 to 2 weeks.

This case emphasizes the need for early diagnosis and prompt treatment required to reverse the condition, reduce mortality and prevent the long term consequences of zinc deficiency.

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