ACRODERMATITIS ENTEROPATHICA
Case report

R. P. C. NAIK * AND M. MEERA BALIGA †

Abstract

A case of acrodermatitis enteropathica (AE) without significant diarrhoea is reported. The response to oral zinc sulphate therapy in the case has been excellent.

Key words: Acrodermatitis enteropathica, Zinc sulphate

Acrodermatitis enteropathica (AE) is a rare but characteristic disease of infancy and childhood transmitted by an autosomal recessive trait. Perioriﬁcal dermatitis starting usually in infancy, diarrhoea and alopecia are the criteria for diagnosis of this condition.

In the case reported here the patient had no significant gastrointestinal disturbances but the skin lesions were typical.

Case Report

An eleven month old male infant was brought to the skin O.P.D. of the K.M.C. Hospital, Manipal with skin lesions and alopecia. The skin lesions started as vesicles on the gluteal region when the infant was $1\frac{1}{2}$ months old. The eruption appeared to respond partially to topical antibacterial therapy. Within a month, however, fresh lesions appeared around the mouth, ears and perianal region. There was crusting and oozing from the lesions and healing occurred usually with hyperpigmentation. Vesicular lesions appeared also on the scalp and there was progressive loss of scalp hair. The boy had received several antibiotics, antihistaminics and topical therapy included, among others, scabicidal preparations. Despite these the skin lesions showed no response.

There was a history of frequent attacks of fever and occasional attacks of diarrhoea. The infant was solely breast fed till 4 months.

The parents were not consanguinous.

Physical examination revealed no abnormalities in cardio vascular system, respiratory system and CNS. The infant was undernourished (4.7 kgs) and showed vesicular, pustular and crusted lesions covering almost the entire body. An area on the abdomen was spared. There was complete absence of scalp and body hair (fig. 1). Right eye showed a corneal ulcer.

Besides a mild degree of anaemia and leucocytosis, no other abnormality was evident on routine investigations.

* Reader and Head,
Department of Skin and S.T.D.
† Associate Professor,
Department of Paediatrics
Kasturba Medical College & Hospital,
Manipal.

Received for publication on 18—4—1980

Fig. 1 Shows alopecia and extensive skin involvement

Staph. aureus was grown from pustular lesions.

The baby was put on erythromycin orally and potassium permanganate compresses were given to skin lesions. Neosporin ointment was applied to the eye. Even though fever was controlled and some of the skin lesions dried up, fresh vesicular lesions continued to appear and did not respond to application of Kenacomb (Nystatin, triaminolone and gramicidin). Diiodohydroxyquin (200 mg/day) was administered orally at this stage for about 10 days, but did not prove effective. Then 0.2% aqueous solution of zinc sulphate was given orally (5 mg/Kg body wt./day in divided doses). There was a dramatic response in 5 days. All the skin lesions disappeared and the baby became alert and active. A maintenance dose of 10 mgs of zinc sulphate per day was advised. The child gets one or two vesicles once in two or three months, which heal rapidly. Very occasionally the child has vomited after ingestion of zinc sulphate. Fig 2 shows the child 3 months after starting oral zinc sulphate. The corneal ulcer healed but left behind an opacity.

Discussion

The striking skin changes including alopecia and diarrhoea or steatorrhoea are virtually pathognomonic of AE. Absence of bowel disturbances is unusual and is seen in a minority of these cases[1,2]. There was no significant diarrhoea in our case. Involvement of eye is known to occur in AE. The corneal ulcer in this case perhaps was due to zinc deficiency.

It is now known that zinc deficiency is the sole cause of this lethal disease. The infants with the autosomal recessive gene do not develop symptoms as

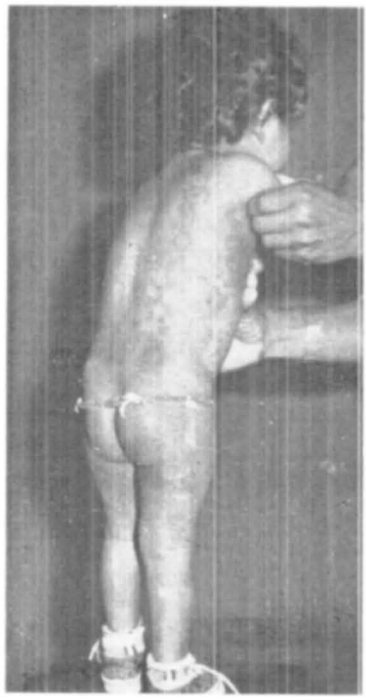

Fig. 2 Shows complete regrowth of hair and healing of skin lesions with oral Zinc Sulphate therapy.

long as they are solely breast fed[3]. Cow's milk lacks a small molecular weight zinc ligand present in human milk and it is suggested that this may be the reason for production of full syndrome of AE when cow's milk is substituted for breast milk[3]. Our case was a clear contrast to this view in that alopecia and dermatitis were severe several weeks before weaning.

Diiodohydroxyquin was the wonder drug till it yielded its place to safe oral zinc sulphate therapy. Diiodohydroxyquin, though effective, has to be given in large doses. There appears to be a threshold and below this level it may not prove beneficial[4]. This may be the reason for the poor response to diiodohydroxyquin in our case. With the advent of oral zinc sulphate therapy patients with AE can be expected to live a normal life. The requirement of zinc increases with age upto puberty and higher doses may be required during infections and pregnancy.

References

1. Shafi M and Ahmed Shah SM : Acrodermatitis enteropathica, Indian J Dermatol Venereol, 1973; 39 : 33-34.

2. Moschella SL : Diseases of nutrition and metabolism, Dermatology, Ed Moschella SL, Pillsbury DM and Hurley HH, WB Saunders Company, Philadelphia, 1975, p 1241.

3. Moynahan E and Grupper C : Acrodermatitis enteropathica and other Zinc-deficiency disorders, Dermatology in General Medicine, Chief Ed Fitzpatrick TB, McGraw-Hill Book Company, New York, 1979 p 1371.

4. Grupper C and Masters R : Acrodermatitis enteropathica, Dermatology in General Medicine, Chief Ed Fitzpatrick TB, McGraw-Hill Book Company, New York, 1971, p 1648.

Please renew your membership and subscription to avoid break in the series of the journal.

Please note that new members will get journals only subsequent to the date of receipt of information in the editorial office regarding their membership from the branch secretaries.

—Managing Editor