Acrodermatitis Enteropathica in a Full-term Breast-fed Infant

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Abstract. Acrodermatitis enteropathica which is a manifestation of zinc deficiency, is rarely seen in breast-fed infants. A child with typical acral rash, zinc deficiency and low breast milk zinc levels is described. The importance of awareness of this entity in breast-fed infants along with differential diagnosis is being emphasized. [Indian J Pediatr 2002; 69 (7) : 631-633]

Key words : Breast feeding; Zinc deficiency, Acrodermatitis enteropathica

Zinc deficiency, hereditary or acquired is an important nutritional disorder of children, predominantly affecting the skin, hair, nails and the gastrointestinal system.1 The term 'acrodermatitis enteropathica' (AE) is used for all patients with acral dermatitis due to zinc deficiency.2 The hereditary form is an autosomal recessive disorder of zinc malabsorption seen exclusively in infants not receiving breast milk; either due to bottle feeding or soon after weaning from the breast milk. Increased zinc bioavailability of breast milk due to the presence of a zinc binding ligand in it confers protection of breast fed infants against its deficiency.1,2 However, acquired zinc deficiency may rarely be observed in premature or full term breast-fed infants.3,4 We present a full-term exclusively breast-fed infant with AE and discuss the importance of early recognition of this rare cause of zinc deficiency along with the differential diagnosis of anogenital rashes in infants and children.

CASE REPORT

A 5-month-old female infant, born normally at full term to non-consanguineous parents presented with a one-month history of a rash over face, legs and buttocks. The rash had not responded to topical corticosteroids, antifungal preparations, or systemic antibiotics prescribed elsewhere. The child was exclusively breast fed since birth and had not been weaned. General health and development of the child was preserved except for poor weight gain in the last 2 months. No history of diarrhea or a similar problem in the family was obtained. The child had an elder brother aged 4 years, who often used to share the maternal breast milk. Examination revealed an irritable child weighing 4.5 kg with a head circumference of 42 cm. The general physical and systemic examination was unremarkable. On cutaneous examination large, erythematosus, well-defined, psoriasiform, dry and scaly plaques were present over the buttocks, perigenital area, and legs (Fig 1). Lesions showed no tendency towards central clearing. Similar lesions were observed over the dorsum of fingers and toes, and peri-orbital region along with angular cheilitis and scaling around the nasal orifices (Fig 2). Scalp, hair, nails, mucosae and joints were normal.

Fig. 1. Well defined psoriasiform annular plaques over buttocks and legs.

A clinical diagnosis of acrodermatitis enteropathica was considered. The child's serum zinc levels were decreased to 47 microgram/dl (normal: 70-120 microgram/dl) and the maternal breast milk zinc levels were also low; 8 microgram/L (normal at 20 weeks gestation; 11-12 micromol/L). Serum alkaline phosphatase
Fig. 2. Angular cheilitis and scaly plaques around nasal and oral orifice.

was reduced to 98 IU/L (normal: 108-306 IU/L). A microscopic examination of potassium hydroxide (KOH) preparation of the skin scrapings was negative. Treatment with oral zinc sulphate 5 mg/kg per day was started and weaning was encouraged. The skin lesions resolved over the next two weeks and the child also became more playful and cheerful.

DISCUSSION

Although recognized by Brandt in 1936, the term acrodermatitis enteropathica was coined by Danbolt and Closs in 1942 to describe the acrally predominant rash present in some patients with diarrhoea. Prior to recognition of zinc deficiency as the etiologic factor of AE by Moynahan in 1971, this disorder was usually fatal in infancy or childhood.

AE is characterized clinically by a triad of dermatitis, diarrhoea and alopecia, and the complete triad is seen in only 20% of patients. The characteristic distribution of the dermatitis in patients with AE over face, hands, feet and anogenital area is recognized as a pathognomonic cutaneous marker for zinc deficiency. The cutaneous lesions are psoriasiform or annular, erythematous, scaly and crusted plaques. As the disease progresses these plaques may become vesicobullous, pustular and erosive. Other features, which occur with varying frequency, 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 of zinc deficiency. The ocular manifestations include photophobia, blepharitis, conjunctivitis and corneal dystrophy. Histopathology of skin lesions is non-specific and reveals parakeratosis, psoriasiform epidermal hyperkeratosis, large pale keratinocytes, few dyskeratotic cells and absence of granular layer in the epidermis.

Presence of psoriasiform or dermatitic lesions over the face, perioral, perigenital and perianal areas in an acral distribution in a child with diarrhoea is quite suggestive of AE. However, it needs to be differentiated from other causes of anogenital rash as such as diaper dermatitis, candidiasis, infantile seborrheic dermatitis, infantile psoriasis, dermatophyte infections, Langerhan's cell histiocytosis and Leiner's disease. Diaper dermatitis usually presents with confluent erythema over convex surfaces of buttocks, genitalia and lower abdomen with sparing of flexures, while in candidiasis lesions have a sharply margined erythematosus border studded with satellite pustules. The rash of seborrheic dermatitis involves the face, scalp, and napkin area comprising of well-defined erythema, papulo-vesicles and small adherent scales. Infantile psoriasis has confluent dry erythema with sharp scalloped margins. Dermatophyte infections characterized by erythematous annular plaques and central clearing are rare in very young children; however, they can be definitely excluded by KOH examination. Langerhans' cell histiocytosis presents in infancy with clusters of small translucent yellowish papules, ulceration and petechiae. Scalp, especially the retroauricular area is almost always concurrently affected. In Leiner's disease severe diarrhoea, wasting, dystrophy and recurrent infections accompany the skin rash. Certain other disorders such as essential fatty acid, carboxylase and aminoacid deficiencies, methylmalonic aciduria, aminoacidopathies and organic acidemias may also have similar cutaneous features and need to be excluded especially if the lesions persist despite zinc supplementation or relapse or recur frequently.

A clinical diagnosis of AE supported by decreased serum zinc levels is usually confirmatory (normal serum zinc level - 70 to 110 micrograms/L). However, the serum levels of zinc may not be totally reliable especially following an intercurrent infection, injury, burns, or any other stressful stimuli, which result in rapid redistribution of zinc. In breast fed infants with acquired zinc deficiency, maternal breast milk zinc levels are also reported to be low, while maternal serum zinc levels may be normal. This non-hereditary form of the disease, as observed in this child, develops due to low breast milk zinc levels secondary to a defective mammary zinc secretion or an abnormal uptake of plasma zinc by mammary gland despite normal maternal serum zinc levels.

Treatment with zinc supplementation, in a dose of 220 mg thrice daily in adults and 3-5 mg/kg/day in children is recommended. There is rapid improvement of diarrhoea within 24 hours and the skin lesions within 1 to 2 weeks. In the hereditary form zinc needs to be supplemented throughout life, while it is usually not necessary after weaning in the acquired form. In the present case sharing of breast milk by the elder sibling may have contributed to depletion of zinc levels in breast milk. Prompt recognition of this disorder and initiation of zinc supplementation and weaning rapidly reverses the