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CONTINUING MEDICAL EDUCATION

ACRODERMATITIS ENTEROPATHICA

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Zinc is an essential mineral that has been found to be present in at least 100 metalloenzymes. Zinc deficiency, whether a result of an acquired or inherited abnormality is associated with characteristic cutaneous findings. The inherited variety is known as acrodermatitis enteropathica (AE). Denbolt and Class[1] gave the condition the name AE. However it was Moyanahan[2] in 1974 who pointed out that this is a zinc deficiency disorder. AE has been reported from many parts of the world including India.[3-10] Acrodermatitis enteropathica is more common in infancy. Classic presentation is of diarrhoea, skin rash with alopecia, extreme irritability and depression. Onset is approximately 1 to 2 weeks after weaning. Infants with AE have an erythematous, scaly, psoriasiform and sometimes vesiculopustular eruption located periorificially (i.e. around the mouth, eyes and genital areas). The finger flexural creases and the palms show characteristic flat greyish bullous lesions surrounded by red brown erythema. Skin lesions also appear on extremities. Refractory diaper dermatitis may be the presenting complaint. Secondary staphylococcal and candidal superinfections are very common, and do not respond to topical therapy unless the zinc deficiency is corrected. Paronychia and nail dystrophy can take place. Hair becomes dry and complete alopecia may develop. Diarrhoea starts early in the disease; however it is not a constant feature as reported by Naik et al.[10] Ocular involvement in the form of corneal changes consisting of superficial punctate lesions, nebulous subepithelial opacities and linear epithelial erosions have been reported.[11] AE has also been reported from both bottle and breast-fed infants.[7,12] If left untreated, severe failure to thrive and death my ensue.

Sharma et al,[7] classified hypozincemia in infants into three categories:

Type 1 : Classical AE, an autosomal reces-

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sive disorder with inherited defect in the absorption of zinc from gut. Clinical manifestation appearing on weaning and child will need zinc for a very long time.

Type 2 : AE-like clinical picture in a breast-fed infant. Case reported by Sharma et al,[7] had no diarrhoea, and the hairs, nails and mucosa were not involved. Similar cases have been reported.[6,12] Ahmed et al,[13] felt that there is a deficiency of zinc-binding ligand in the breast milk. This type is also called as 'hypozincemia of infancy'.

Type 3: Preterm infant put on prolonged parenteral alimentation without zinc supplementation.

Low levels of plasma zinc are pathognomonic of AE. Care must be taken while collecting blood samples. Samples should be collected in special acid-washed glass bulbs or plastic tubes, otherwise exogenous zinc present on ordinary glassware will contaminate the specimen. Mack et al,[14] reported a patient of AE with normal serum zinc levels in whom the diagnosis was confirmed by plasma phospholipid fatty acid and a small bowel biopsy. Other associated laboratory findings are low serum alkaline phosphatase, hypogammaglobulinemia, abnormal cell-mediated immunity or anergy.[15]

The actual metabolic error in AE has not yet been defined, but it is clearly a problem with intestinal absorption and / or transport of zinc. Picolinic acid, a tryptophan metabolite, enhances the intestinal absorption of zinc which is deficient in AE.[16] Prasad at al, [17] have reported adolescents from the Middle-east with zinc deficiency which develops due to concurrent ingestion of high amounts of the zinc binding ligand, phytate present in high quantity in that area. Sandstorm et al,[18] suggested that the primary lesion in AE is in cellular defect in zinc metabolism rather than an impairment of zinc absorption.

The zinc derived from plant origin is less available for utilization and absorption due to high phytate and fibre content, than the zinc derived from animal product.[4]

The specific cutaneous manifestations of zinc deficiency may be in part due to zinc interaction with vitamin A metabolism. The interaction of zinc and vitamin A may be mediated via the enzyme retinol alcohol dehydrogenase, which converts photoactively inert retinol to the active retinaldehyde or by impairing synthesis of retinol binding protein.[19]

Hydroxyquinoline was found empirically to provide successful therapy in AE.[3] It most probably enhanced intestinal zinc absorption. Zinc supplementation has a rapid and dramatic effect that reverses all cutaneous, gastro-intestinal and neurological manifestations of the disease. Michaelsson,[20] was the first to use zinc in AE. Zinc can be administered as a sulfate (22.5mg of elemental zinc / 100mg).[8] The zinc sulfate salt may sometimes act as an irritant to the GI tract and may precipitate bloody diarrhoea.[7] Bhargava et al,[5]

reported use of 'Jasad Bhasm' (which contains zinc) useful in two cases of AE. Zinc in a dose of about 1-2mg kg/day is useful to cure all clinical manifestations within 1-2 weeks. Zinc may need to be supplemented for life-time. However, it may show improvement with age.[21]

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