ACRODERMATITIS ENTEROPATHICA

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Summary

Fifteen cases of Acrodermatitis Enteropathica, a rare disease, are reported from S. M. H. S. Hospital, Srinagar. A review of literature pertaining to incidence, aetiopathogenesis of this disease is briefly discussed.

The first description of this rare disease was provided by Danbolt and Closs[1] in 1942. The original two cases described by them had a symptom complex of diarrhoea, periorificial dermatitis, alopecia and apathy. The condition, though chiefly restricted to children, has been reported in adults also. Of the 65 cases reported till 1963, 3 were adults[2].

Though Acrodermatitis Enteropathica is a rare disease, cases are reported from many centres off and on. The fact that in a period of about two and a half years fifteen cases of Acrodermatitis Enteropathica were spotted in the S.M. H.S. Hospital, Srinagar, is noteworthy. The review of literature did not reveal any single collection of this size.

Case Material

Fourteen out of the 15 cases were diagnosed in the skin outpatients department. One case, a child of 1½ years of age, was referred to Skin Department from the Surgical Department for the cutaneous lesions and had been admitted there for severe trismus which probably was secondary to severe mucosal ulcerations. Table 1 shows the age and sex distribution of the 15 cases:

TABLE 1

Age		
1–2 years	...	10 cases
2—3 years	...	5 cases
Sex		
Male	...	8 cases
Female	...	7 cases

The criterion of diagnosis of this condition was the clinical picture of symmetrical vesiculo-pustular dermatitis, in upper and lower limbs and periorificial regions. 13 cases had variable degrees of diarrhoea, 10 were having glossitis and stomatitis, while 4 had conjuctivitis, blepharitis and chronic paronychia. These children had low body weight, were apathetic and irritable. Another additional diagnostic criterion was the fairly prompt therapeutic response to the oral administration of quinoline derivatives. Only one case died during the period of observation, the child with severe trismus (already referred to). This child was severely dehydrated and probably died of electrolyte imbalance. Two cases had a positive family history. Two of their siblings had suffered from similar disease. Two cases had no history of diarrhoea but had typical skin lesions and these responded to the Quinolines without any supplementary systemic or topical medication. Apart from anaemia and ascariasis infestation in some of the cases other routine laboratory investigations were non-contributory.

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Discussion

Acrodermatitis Enteropathica is a rare hereditary disease of infancy and early childhood. The mode of inheritance has been suggested as a recessive trait. Typical cutaneous lesions situated on the distal part of extremities and around the body orifices occur in association with diarrhoea, weight loss and apathy. The incidence of super added bacterial and monilial infection has been reported to be high. That a majority of these cases respond to the administration of halogenated hydroxyquinolines has been a constant observation.

The etiology and pathogenesis of this condition are obscure. The observations of Mucke, Dittmer, Gymreck et al[4] are a pointer to the basic defect being a disturbance of Tryptophan Metabolism. These workers performed tryptophan loading studies on three of their cases and showed increased urinary elimination of kynurinic and xanthiurinic acid. The response to tryptophan administration was the appearance of new skin lesions or exacerbation of old ones. Control study on one patient demonstrated the return to normal of the tryptophan loading test after the administration of oxyquinoline. These authors suggest that this disease is dependent upon a defect in the kynurinic system with the elaboration of pathogenic metabolites. The therapeutic effect of 8-hydroxyquinolines is explained by competitive inhibition of tryptophan degradation at some step prior to the formation of kynurinic acid. Cash and Berger[5] studied fatty acid metabolism in a patient of this disease and demonstrated a defective interconversion of the unsaturated fatty acids.

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