KINDLER SYNDROME; CLINICOPATHOLOGICAL AND ULTRASTRUCTURAL STUDY OF ORAL LESIONS

Mohamed S. El-Zarka,* Zeinab Darwish** and Manal El-Nouaem**

ABSTRACT

Kindler syndrome is a rare, inherited mucocutaneous disease characterized by acral bullae formation in infancy and early childhood and generalized progressive poikiloderma with marked cutaneous atrophy. A number of oral features have been described, such as gingival swelling and mucosal atrophy and erosions. Three cases (one male and 2 females) were included in the present study. These cases were already diagnosed as having kindler syndrome. Clinical examination of the oral and paraoral tissues was done. In addition, routine histopathological, immunohistochemical and electron microscopical studies were applied to the gingival biopsies of 2 patients (the male and one of the 2 females). The results revealed abnormality in the basement membrane and in the staining for monoclonal anti keratin antibody. These alterations are likely to play a role in the pathology of this disease and the techniques done on the present study may be useful for identifying kindler syndrome.

* Prof. of Oral Pathology Dept, Faculty of Dentistry. University of Alexandria.

** Ass. Prof. of Oral Pathology Dept, Faculty of Dentistry. University of Alexandria.