

## INTRODUCTION

Lipoid proteinosis is a rare recessively inherited multisystem disorder which primarily affects the skin, oral cavity and larynx. About 200 cases have been described with an increased incidence in South Africa and Sweden. The characteristic abnormality of the disease is the deposition of an amorphous hyaline material in the skin and mucous membrane (Harper et al., 1983a).

Aetiology or pathogenesis is still unknown. It is considered as a congenital and familial mucopolysaccharidosis (Moynahan, 1966 and Moschella, 1975). Histochemical studies identified the nature of the hyaline material as a carbohydrate-protein complex and also has a lipid component (Harper et al., 1983b).

As regards the pathogenesis, Harper et al (1985) suggested that lipoid proteinosis results from primary perturbation of collagen metabolism.

Lipoid proteinosis is of two types (Bleehen, 1979), the Urbach-Wiethe type- the common presenting feature is hoarseness from infancy due to infiltration of larynx. The second form is the light sensitive form which is more correctly designated as erythropoietic protoporphyria.

The disease occurs equally in both sexes (Findlay, 1960).

As regards treatment, there is no known satisfactory therapy for lipoid proteinosis (Caro, 1982 and Haneke et al., 1984).