Urbach-Weithe disease

A 10-year-old girl born of consanguineous marriage presented with hoarseness of voice and difficulty in protruding tongue since last five years. No other family member was affected. Cardiovascular, ophthalmological and neurological examination was normal. Hematological profile was normal, including X-ray skull. Cutaneous examination showed classical multiple beaded papules along the eyelid margins - also called as moniliform blepharosis (Fig. 1a) – with pock like scars on the bilateral elbows and verrucous, hyperkeratotic plaque on the left elbow (Fig. 1b). Oral cavity examination showed woody hard tongue showing yellow white infiltration and inability to protrude tongue beyond lip margin (Fig. 1c). Multiple yellowish papules with atropic scars were noted in bilateral axillae (Fig. 1d). On the basis of classical cutaneous findings, a diagnosis of Lipoid proteinosis was made. The patient was started on acitretin (25_mg/day). It should be differentiated from lichen myxedematosus, lichen amyloidosis, xanthomatosis and colloid miliium.

Lipoid proteinosis, also known as Hyalinosis cutis *et mucosa* or Urbach-Weithe disease, is a rare autosomal recessive disease with multisystem involvement and is caused due to mutations in the *ECM1* gene located on chromosome 1q21. There is no promising specific treatment available for Urbach–Wiethe disease although

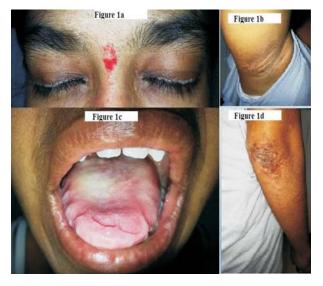


FIG. 1 (a) Multiple beaded papules along the bilateral eyelid margins; (b) Pock like scars on the elbows (c) Infiltrated tongue with inability to protrude; and (d) Yellowish papules and scars in axillae.

symptoms can be treated individually. Prognosis is good and patients usually have normal life span.

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Cradle Cap

A 3-month-old boy presented with a lesion over the scalp for a week. Examination revealed a well-defined patch with a greasy surface, and scaling on the top of the lesion. It was surmounted by thick yellow-brown crust (*Fig.* 1). Axillae, groins and other areas of the body were free from any lesions; nails and mucosae were normal. A diagnosis of cradle cap was made and ketoconazole shampoo was prescribed.

Cradle cap is a type of seborrheic dermatitis, exclusively affecting infants; more commonly within first 6 weeks of life. It usually presents as asymptomatic thick, crusty, yellow-brown patches. Fungal infection (*Malassezia furfur*) and overactive sebaceous glands are



Fig. 1 Well-defined lesion on scalp covered with crust.

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