

## 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 atrophic 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 (25mg/day). It should be differentiated from lichen myxedematosus, lichen amyloidosis, xanthomatosis and colloid milium.

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-Weithe 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.

implicated in the pathogenesis. The condition clears off as the baby grows older. Treatment consists of removal of crusts with 3% salicylic acid; and application of low-potency steroids and antifungal agents. Clinical differentials include tinea capitis (easy pluckability of hairs), atopic dermatitis (presence of pityriasis alba, positive family history), impetigo (honey colored crust),

and psoriasis (scaly erythematous plaque with involvement of other sites and nails).

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## Nevus of Ota

A 6-year-old girl presented with an asymptomatic dark patch over the left side of her face. Her parents first noticed a small patch of bluish discoloration, when she was two-months-old, that gradually increased in size and became darker in color. On examination, mottled blue-gray macules were seen over the left cheek extending to the left temporal area (*Fig. 1*). In addition, two small blue-black spots were seen on the sclera of her left eye. Oral mucosa, hairs, and nails were normal. Systemic examination was non-contributory. Based on the distinct color, morphology, and location of the lesion, the condition was diagnosed as nevus of Ota.

Nevus of Ota is believed to occur due to migration arrest of melanocytes on their way to the epidermis from the neural crest. It typically occurs as a persistent, speckled, blue-black, or slate-gray hyperpigmentation in the distribution of the ophthalmic and maxillary division of the trigeminal nerve. The bluish color is due to scattering of light by dermal melanin (Tyndall phenomenon). Though mostly present at birth, it can also appear later; 80% of the cases occur in females. Unilateral involvement is seen in 95% of the cases. Patchy scleral pigmentation is seen in two-third of the cases. Pigmentation may also be seen in the oral mucosa, cornea,



**FIG. 1** Mottled blue-grey macules of Nevus of Ota.

pharynx, and nasal mucosa. The condition is generally benign and persists for life. The treatment of choice for the condition is laser surgery; cosmetic cover-up may also be used.

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## Ecthyma

An 8-year-old boy presented with a painful crusted lesion over the left forearm. Cutaneous examination revealed a solitary coin-sized, indurated, ulcerated, tender plaque with central brownish adherent crust and yellowish-brown dried exudates at the margin (*Fig. 1*). There was no preceding history of any insect or arthropod bite. The Gram stain from pus obtained from underneath the crust

revealed gram-positive cocci, and culture grew both Group A  $\beta$ -hemolytic streptococci and *Staphylococcus aureus*. A diagnosis of ecthyma was made; oral cefixime and topical mupirocin ointment were prescribed along with removal of crust using diluted white vinegar soaks. Complete healing with scarring occurred within 2 weeks.

Ecthyma denotes cutaneous bacterial infection that extends deep into the dermis and heals with scarring. It usually develops over disrupted skin on extremities and rapidly develops into a vesicopustule and finally a