Urbach-Weithe disease

A 10-year-old girl born of consanguinous 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 infiltration and inability to protude 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 protcinosis was made. The patient was started on acitretin (25mg/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 symptoms can be treated individually. Prognosis is good and patients usually have normal life span.

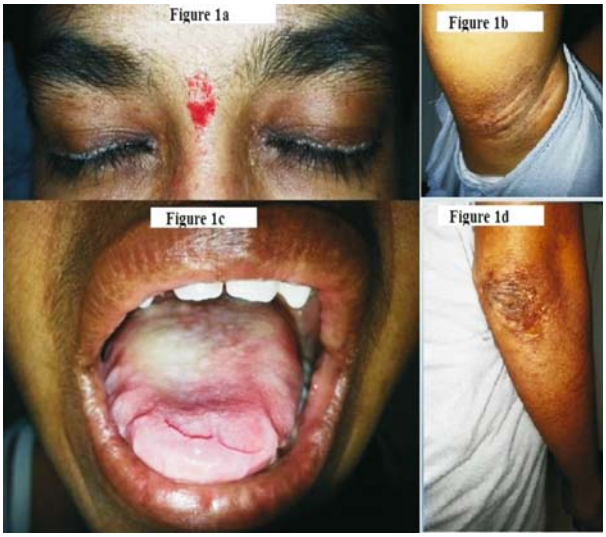

FIG. 1 *(a) Multiple beaded papules along the bilateral eyelid margins; (b) Pock like scars on the elbows (c) Infiltrated tongue with inability to protrude; and (d) Yellowish papules and scars in axillae.*

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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

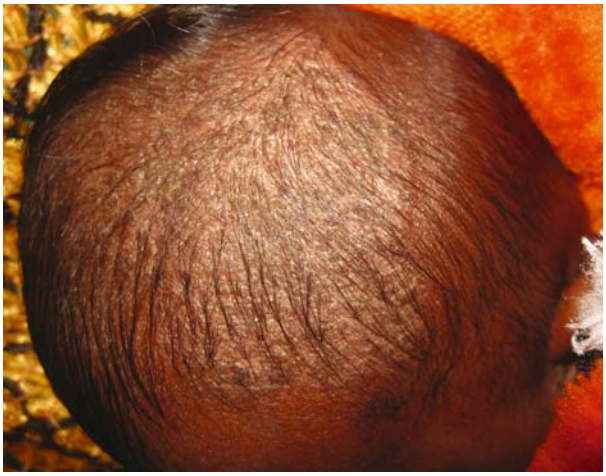

FIG. 1 *Well-defined lesion on scalp covered with crust.*

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.

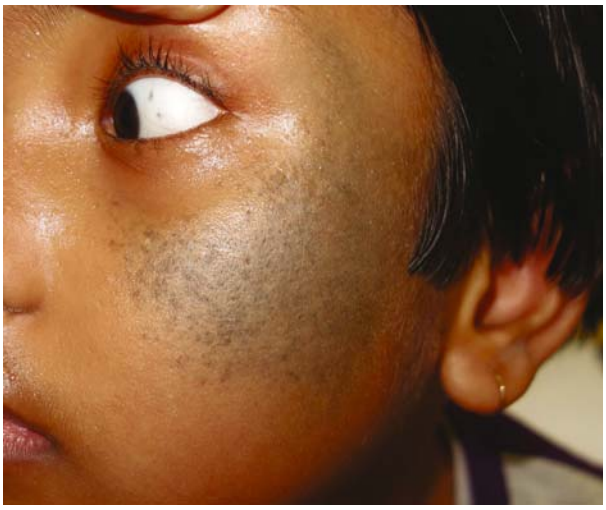

FIG. 1 *Mottled blue-grey macules of Nevus of Ota.*

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, 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 β-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

hemorrhagic crust. Differential diagnoses include arthropod bites, leishmaniasis, ecthyma gangrenosum (*vide infra*), pyoderma gangrenosum, *Mycobacterium marinum* infection, and papulonecrotic tuberculid. Two related terms need to be differentiated from ecthyma: *Ecthyma gangrenosum* (a gangrenous ulcer with a central eschar surrounded by an erythematous halo) a pseudomonal infection that occurs in immuno-suppressed or gravely ill patients, and *Ecthyma contagiosum* (solitary pustular lesions on hands) resulting from the direct contact of damaged skin with animal infected by a virus of Parapoxvirus group.

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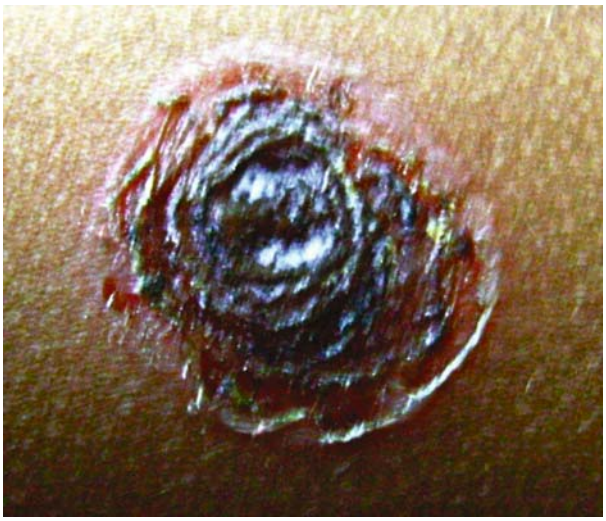

FIG. 1 *Solitary ulcer with central adherent crust and yellowish-brown dried up exudates at the ulcer's margin.*

Congenital Triangular Alopecia

A 9-month-old child presented with two localized areas of hair loss over the occipital region, noticed by the parents since the age of three months. Parents did not report any history of trauma, redness, swelling, erosion or ulceration prior to the onset of hair loss. None of the family members had similar complaints. The patient had been treated with topical antifungal and steroids for two months without any improvement. Examination revealed two well demarcated triangular patches of non-scarring alopecia, in the occipital area; the base of the triangles was towards the vertex (***Fig.* 1**). The overlying skin was smooth and shiny with no scaling or erythema. Exclamation mark hairs were absent. Examination of nails did not show ridging or pitting. The triangular shape of alopecia, absence of exclamation hair and lack of response to any treatment in past led us to the diagnosis of Congenital triangular alopecia. Counselling of the parents was done about the poor prospect of hair regrowth and option of hair transplantation in future.

Congenital triangular alopecia or Brauer nevus, is a benign, non progressive, circumscribed and non-scarring form of alopecia usually present over the temporal region. Other conditions that should be considered under differential diagnosis are alopecia areata (round or oval patches of rapid and complete hair loss with exclamation point hairs, pitted nails and tapered fractures on the hair counts), aplasia cutis (solitary or multiple well demarcated oval or circular atrophic fibrotic scars or ulcerations), tinea capitis (itchy, scaly or erythematous ring lesions with greyish, lustreless, brittle and broken hairs) and trichotillomania (irregular areas of incomplete hair loss in occipital or parietal areas of scalp).

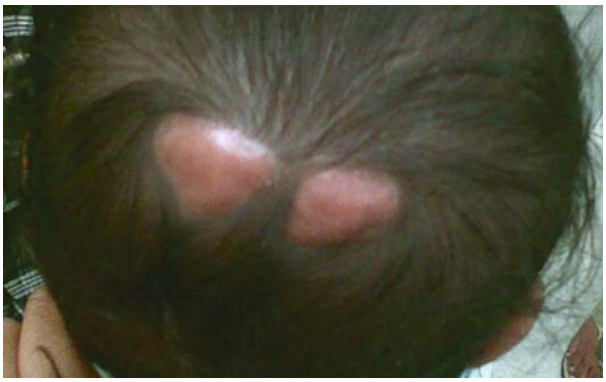

FIG.1 *Congenital triangular alopecia.*

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