

Urticaria Multiforme

A 10-month-old girl presented with erythematous pruritic rash on cheeks, ears and trunk along with swelling of the feet for last 12 hours (*Fig. 1a*). She had a fever with coryza for two days and received three doses of paracetamol. Rashes were discrete, round, blanchable, and a few were 'target lesions' (*Fig. 1b*). There was no evidence of mucosal involvement, arthritis or involvement of any other system. Complete blood count, erythrocyte sedimentation rate and C-reactive protein were normal. A diagnosis of urticaria multiforme was made and she was treated with hydroxyzine and ranitidine. The rashes gradually disappeared in next 4 days.

Urticaria multiforme, a form of acute annular urticaria, is a benign, self-limiting cutaneous hyper-sensitivity reaction mediated by histamine. It is characterized by the acute and transient onset of blanchable, arcuate, annular, polycyclic, erythematous wheals and facial or acral edema. It usually occurs in children between 4 months and 4 years. Viral illness, immunization and drugs like antibiotics or paracetamol could be the inciting factors. Erythema multiforme, urticarial vasculitis (both having fixed lesions, dusky, purpuric centers often with necrosis and blistering



FIG.1 Erythematous rash on cheeks, ears and trunk.

which resolve slowly over weeks with post inflammatory depigmentation; and mucosal erosions); and serum sickness (high grade fever, myalgia, arthralgia, lymphadenopathy and frequent neutropenia in addition to skin and mucosal involvement) are important differential diagnoses.

***SUMANTRA SARKAR** AND #**ABHISHEK DE**
 Departments of Pediatrics and #Dermatology,
 IPGME&R and SSKM Hospital,
 Kolkata, West Bengal, India.
 *sumantra.com@gmail.com

Adams Oliver Syndrome

A 2-month-old girl, born to a non-consanguinously married parents, presented to us with presence of an open wound at birth which healed with scarring and hair loss, along with abnormalities involving toes of both feet. The lesion was an atrophic scar tissue with a rough and heterogeneous appearance. Physical examination revealed localized alopecia with dimensions of 1.8×4 cm (*Fig. 1a*), along with prominent scalp veins. Cutis marmorata was present all over body. Oligodactyly and digital nubbins were present in bilateral feet (*Fig. 1b*).

Based on the classical clinical presentation of aplasia cutis congenita of scalp, cutis marmorata, prominent scalp veins, and limb abnormality, a diagnosis of Adams



FIG.1 (a) Alopecia cutis congenita of scalp, prominent scalp veins; (b) Oligodactyly and presence of digital nubbins.

Oliver syndrome was considered. Histopathology of hairless atrophic patch showed loss of rete ridges, collagen deposition and loss of skin appendages. Hemogram, liver function tests, kidney function tests, serum electrolytes and Chest X-ray were normal.

Adams Oliver Syndrome is a rare and clinically heterogeneous anomaly characterized by the combined occurrence of congenital scalp defects and terminal transverse limb defects. It includes aplasia cutis congenita, variable limb defects, and associated anomalies ranging from skin tags to lymphedema. Other system anomalies and malformations such as cardiovascular, respiratory and orofacial defects have also been reported. With conservative therapy to prevent secondary infection and consequent tissue damage, most small defects of scalp heal well during the first few months of

life. Larger and obvious scars can be treated with plastic surgical reconstruction. The lesions of cutis marmorata may fade with time during first year of life due to skin thickening and maturation.

***ANJALI MADAN, KABIR SARDANA AND
VIJAY KUMAR GARG**

*Department of Dermatology,
Maulana Azad Medical College,
New Delhi, India.*

**anjalimadan85@gmail.com*

Facial Angiofibromas

A nine-year-old boy presented to us with recurrent episodes of convulsions over the preceding two years. His family and birth history was unremarkable. He had learning difficulties and poor scholastic performance. Cutaneous examination revealed multiple non-tender, dark brown and skin-coloured telangiectatic papules of varying size over the face (**Fig. 1**). In addition, he had three oblong hypopigmented macules over the upper back. Systemic examination including ophthalmoscopy was normal. Computerized tomographic (CT) scan of brain revealed multiple calcified subependymal nodules of different size. Based on the features, a diagnosis of tuberous sclerosis was established. Subsequent echocardiography and ultrasonography of the abdomen were normal.

The distinctive facial lesions in the present patient were angiofibromas, also referred by a misnomer – adenoma sebaceum. These are pink, dark brown, or skin-colored telangiectatic papules, often observed in the nasolabial folds and on the cheeks and chin. Pathologically, they are hamartomas, and composed of fibro-vascular tissue. They usually present after 2 years of age and gradually increase in size and number until adolescence. Besides tuberous sclerosis, angiofibroma may also be seen in multiple endocrine neoplasia type I and Birt-Hogg-Dube syndrome. Treatment of these skin lesions is usually required for cosmetic concerns, and different forms of laser being the best option.



FIG. 1 (a) Dark brown and skin-coloured papules of varying size over the face; and (b) Close-up showing typical lesions of angiofibromas.

Common differential diagnoses of angiofibroma include, trichoepithelioma (small, firm, flesh-coloured papules, begin during early puberty), acne (polymorphic lesions, presence of comedones, also present in other acne-prone areas, usually noted during adolescence), syringoma (small, skin-colored, dermal papules, typically on the lower eyelid; occurs between 20 and 40 years of age), and sebaceous hyperplasia (<3 mm yellowish telangiectatic papules with central umbilication; common in elderly people).

**ABHIJIT DUTTA, *SUDIP KUMAR GHOSH AND
#RAJESH KUMAR MANDAL**

*From Departments of Pediatric Medicine and #Dermatology,
North Bengal Medical College; and *Dermatology, RG Kar
Medical College; Kolkata, West Bengal, India.
dr.adutta@yahoo.co.in*