Congenital Cutis Laxa

A four- month- old male child presented with history of repeated episodes of fever, cough and fast breathing since early neonatal period. He had old man like facies with puffy eyes, prominent easily stretchable ears and everted nostril. Skin was loose and redundant, most marked over both the lower limbs. There was small paraumbilical hernia, left inguinal hernia and micropenis **(*Fig.*1)**. Abdomen was soft and distended with hepatosplenomegaly (liver 3 cm and spleen 2 cm below subcostal margin). Anteroposterior diameter of the chest was increased with bilateral crepitations and rhonchi on auscultation. Weight of the child was 3.6 kg (< 5th percentile), length- 53 cm (< 3rd percentile) and head circumference- 35.8 cm. *X*-ray chest revealed bilateral emphysematous lungs. Based on clinical and radiological features, a diagnosis of congenital cutis laxa was made.

Congenital cutis laxa is a rare hereditary connective tissue disorder resulting from abnormality of elastin fibres. It is characterized by loose, redundant skin all over body associated with other extra cutaneous manifestations in form of inguinal and umbilical hernia, pulmonary emphysema, gastrointestinal and bladder wall diverticulae and pulmonary artery branch stenosis. Our patient probably had type I autosomal recessive variant of cutis laxa. Absence of similar type of illness in other family members rules out the possibility of autosomal dominant type. The condition needs to be differentiated from Ehlers-Danlos Syndrome, where skin is hyperextensible and joints are hypermobile. Other condition with cutis laxa like skin changes is Costello syndrome which can be differentiated by presence of hyperkeratotic changes of palms and soles, abnormally flexible finger joints and presence of papillomata around mouth and nostrils. Absence of joint laxity rules out the possibility of type II congenital cutis laxa and Ehlers- Danlos Syndrome. There is no specific treatment for this condition. Prognosis of type I congenital cutis laxa is bad and majority of children die in early childhood due to pulmonary or cardiovascular complications.

(*a*) (*b*)

FIG. 1 *(a) Senile appearance of the face, puffy eyes and everted nostrils; (b) Loose skin folds over lower limbs, small paraumbilical hernia, right inguinal hernia and micropenis.*

MANISH KUMAR AND RAGHVENDRA SINGH
Department of Pediatrics, Chacha Nehru Bal Chikitsalaya (Affiliated with Maulana Azad Medical College), Geeta Colony, Delhi 110 031, India.
manishkp75@yahoo.com

Multifocal Cystic Osseous Tuberculosis with Lupus Vulgaris

A 6-year boy reported with multiple swellings on the dorsum of both hands and elbows and low grade fever for 6 months. He had received multiple antibiotics without any response. Examination revealed cervico-axillary lymphadenopathy with irregular soft swellings on the dorsum of both hands and elbows. Overlying skin was erythematous with ulcers having bluish thin undermined margins and serosanguinous ooze **(*Fig.* 1)**. Investigations revealed ESR 50mm, Mantoux 15mm and normal chest

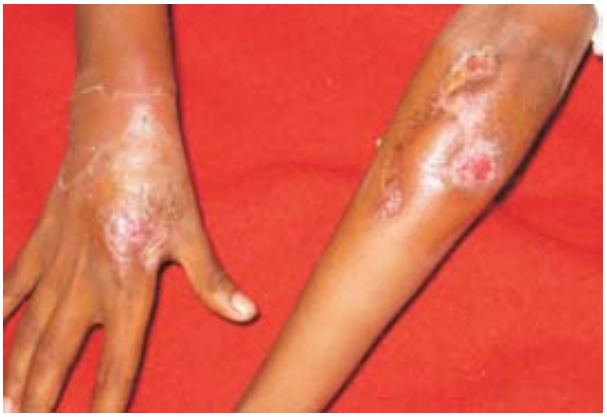

FIG.1 *Erythematous skin with characteristic ulcers.*

skiagram. *X*-ray hands revealed multiple lytic lesions and surrounding sclerosis (honeycombing) in metacarpals and multiple, oval, cystic lesions with variable sclerosis in radius and humerus **(*Fig*. 2)**. FNAC of lymph nodes and skin biopsy from the dorsum of hands revealed granulomatous pathology suggestive of tuberculosis. Patient responded to antitubercular treatment.

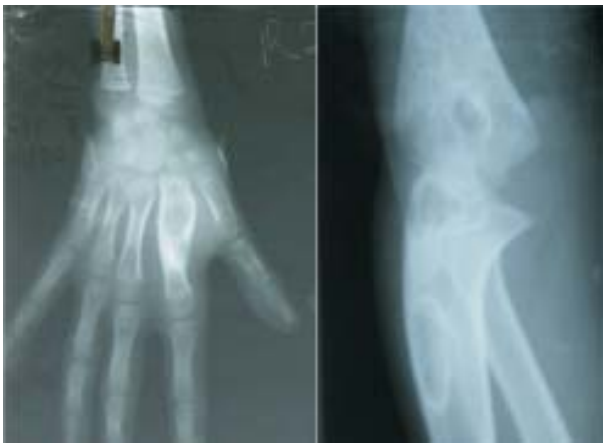

FIG. 2 *Lytic lesions in bones.*

Multiple cystic tuberculosis is a rare form of tubercular osteomyelitis. The cysts may be solitary or multiple, radiolucent, round to oval, situated in the peripheral skeleton near the metaphysis. The commonly affected sites in order of frequency are spine, tibia, ulna, radius, femur, fibula and humerus. The differential diagnosis includes bacterial or fungal infection, aneurysmal bone cysts, cartilaginous tumors and osteoid osteomas. Biopsy may be required to confirm such cases. ATT is the mainstay of treatment. Surgery may be required in a few cases with extensive disease.

REKHA HARISH, ASHU JAMWAL AND SUNIL DUTT SHARMA
Department of Pediatrics,
Govt Medical College, SMGS Hospital,
Jammu, J&K.
kkrhdang@gmail.com

Multiple Annular Erythematous Lesions with Trailing Scale

A 6-year-old boy presented with recurrent pruritic scaly lesions all over trunk for last 2 years. These lesions appeared spontaneously and resolved in 4-8 weeks without any sequale. Rest of the history was non-contributory and no other family member was having similar lesions. On examination, multiple, annular erythematous lesions with trailing scale at the inner border were present on trunk (***Fig.* 1**). Few annular lesions were found on tongue too. On enquiry, tongue lesions used to appear and disappear simultaneously with trunk lesions. Palm, sole, nail, scalp and other mucosa were unremarkable. Routine investigations and KOH mount did not reveal any abnormality. Clinically, the child was diagnosed as Erythema annulare centrifugum (EAC). Histopathological findings were consistent with the diagnosis. Involvement of tongue is very rare. The incidence of disease peaks in the fifth decade.

Erythema annulare centrifugum (EAC) is the most common figurate erythema and is characterized by solitary or multiple, annular or polycyclic lesions that grow slowly (2-3 mm/day); with a trailing scale at the inner border of the annular erythema. The majority of cases are idiopathic; some association with dermatophytosis, infections, drugs, foods (tomato) and malignancy have been noted. The

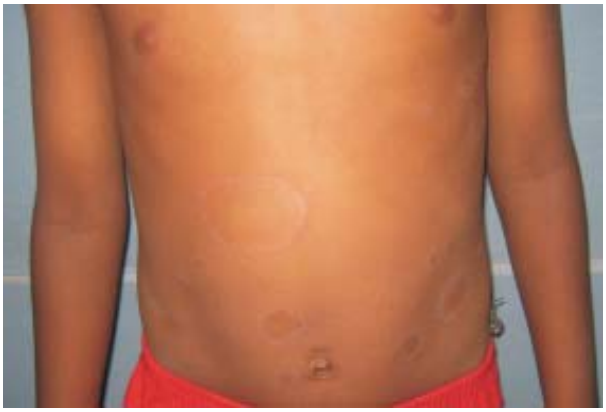

FIG.1 *Mutiple annular erythema with trailing scale at inner border.*

differential diagnosis includes those conditions that can have annular configuration and includes tinea, granuloma annulare, secondary syphilis, subacute cutaneous lupus erythematosus, erythema marginatum, erythema migrans, annular urticaria, and mycosis fungoides. The "trailing scale at inner border" of EAC is diagnostic. Most cases subside spontaneously.

AVIJIT MONDAL, *PIYUSH KUMAR AND #RAJESH KUMAR MANDAL
Departments of Dermatology, College of Medicine and JNM Hospital, Kalyani, Nadia;
**Katihar Medical College, Katihar, Bihar and #North Bengal Medical College.*
docpiyush@gmail.com