Nicolau's Syndrome

An 11-year-old girl, suffering from Dengue fever, developed pain and swelling in the dorsum of left hand following intravenous Ceftriaxone. The swelling resolved over 8 days and the affected area turned reddish violet and eventually black over next 20 days. On cutaneous examination, a single necrotic eschar 5×7 cm with a surrounding tough, fibrotic margin was present on the dorsum of the left hand (*Fig.* 1). On the basis of history and clinical features, we diagnosed Nicolau's Syndrome. Debridement of the area of necrosis was done along with oral antibiotics. Her wound healed in three weeks (*Fig.* 2).

Nicolau's Syndrome – also known as Embolia Cutis Medicamentosa – is a rare, cutaneous, adverse complication of drug administration. Inflammation, embolism, thrombosis and vasospasm are the various mechanisms proposed. Direct or indirect vessel damage involving any or all these mechanisms ultimately leads to peripheral arterial occlusion. Differentials to be considered are acute compartment syndrome (severe pain, pallor, swelling, paresthesia and poikilothermia), subcutaneous fat necrosis (seen exclusively in newborns), and gangrene (distally located). Prevention is by taking care to avoid accidental intra-arterial or para-arterial injection or injection of the drug into an arteriole. Debridement is the mainstay of therapy.



FIG. 1 Necrotic eschar on dorsum of left hand.



Fig. 2 Healed lesions after 3 weeks.

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Erythema Annulare Centrifugum

An 11-year-old boy presented with recurrent, self-healing, asymptomatic eruptions involving trunk, over the last three years. The eruptions used to start as small red papules, progressing centrifugally to form annular plaques with a central clearing. No systemic features or mucosal lesions were present. Physical examination revealed multiple erythematous annular and polycyclic plaques, with trailing scaling at their inner margins (Fig. 1). The lesions were present exclusively on trunk; rest of the muco-cutaneous examination was non-contributory. There was no lymphadenopathy. KOH mount of scales did not reveal any fungal hyphae. Blood investigations were noncontributory. Histopathology from the erythematous margin showed mild hyperkeratosis, focal parakeratosis, and perivascular lymphocytic infiltrate in the superficial as well as deep dermis. The patient was diagnosed with erythema annulare centrifugum (EAC).

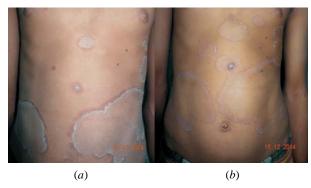


FIG. 1 Annular erythematous plaques with trailing scales on trunk; on presentation (a), and after 2 weeks (b).

EAC is one of the figurate or gyrate erythemas, others being erythema marginatum (transiently seen in acute rheumatic fever), erythema migrans (rash of localized Lyme disease caused by *Borrelia burgdorferi*) and erythema gyratum repens (usually associated with visceral malignancy, pulmonary tuberculosis, lupus erythematosus and azathioprine). EAC presents as asymptomatic annular,

arcuate, circinate, or polycyclic erythematous plaques with indurated margin and a trailing scale noted on the inner aspect of the advancing edge. Rapid progression is typically seen. The condition is recurrent and the course may last 4-6 weeks to many years. It has been documented in association with infections, drugs (Chloroquine, Hydroxychloroquine, Piroxicam, salicylates, Amitriptyline, Hydrochlorothiazide etc), pregnancy, and malignancy. The differential diagnoses include tinea corporis (itchy, papules/ pustules at the margin and fungal hyphae on KOH mount), subacute cutaneous lupus

erythematosus, and other figurate erythemas. Topical steroids usually cause resolution of the lesions of EAC, but they do not prevent new lesions or recurrence. A search for, and treatment of the underlying disorder is warrented, but an exhaustive workup for occult malignancy for EAC alone is not recommended.

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Congenital Platelike Osteoma Cutis

An 11-month-old infant was evaluated for gradually enlarging localized swellings over the left side of the chest, present since birth. The perinatal, developmental and family history were unremarkable. Cutaneous examination revealed two well-defined porcelain white-colored, platelike hard, subcutaneous swellings over the left upper lateral chest wall, measuring 15×20 mm and 5×10 mm (*Fig.* 1). The swellings were non-tender, free from underlying structures and without any visible discharge. There was no clinical evidence of rickets. Serum and urinary levels of calcium and phosphorus, and serum parathyroid hormone levels were normal. A chest radiograph revealed two prominent spicules of calcification in the soft tissue of left lateral chest wall at the level of 10th rib. A diagnosis of congenital plate-like osteoma cutis was confirmed on excisional biopsy that revealed dermal ossification with multiple osteocytes (Fig. 2).

Four types of osteoma cutis have been identified – congenital plaque- or plate-like, late-onset osteoma, widespread osteomas, and multiple miliary facial osteomas. Plaque-like osteoma is present since birth. Although the scalp and extremities are commonly affected, any site may be involved. In osteoma cutis, bone arises in skin and soft tissues through membranous ossification, purportedly effected by osteoblastic differentiation of dermal fibroblasts. Clinical diagnosis is confirmed on plain radiography and histopathology of the excised specimen. Serum calcium and parathyroid hormone levels aid in ruling out Albright's hereditary osteodystrophy. Surgical excision is the mainstay of treatment.

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FIG. 1 Porcelain white-colored, plate-like, subcutaneous swellings over the left upper lateral chest wall.

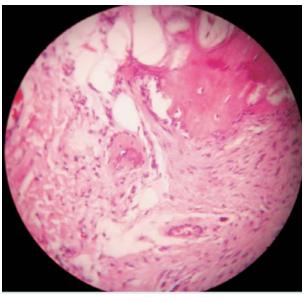


FIG. 2 Dermal ossification with multiple osteocytes in ovalshaped lacunae (hematoxylin & Eosin, 400×).