Ectodermal dysplasia is a rare condition occurring in an estimated 1/100,000 live births. It embrases several abnormalities whose common denominator is a morphological alteration of ectoderm derived organ and tissues.

Case 1, an 11-year-old Hindu boy born of first degree consanguinity, presented with inability to sweat, heat intolerance and absence of teeth. Positive findings on examination were sparse fine hair over scalp, absent eyebrows and lashes, frontal bossing, saddle nose, prominent chin, protruding lips, hyperpigmentation over periorbital and perioral area with photophobia, cone shaped teeth which were two in number. Skin was thin and dry, nails were normal, systemic examination was normal. Case 2 was preterm female baby delivered at 8 months gestation, younger sibling of Case 1 and was seen on first day for sparse and scanty hair, wrinkled eyelids and absence of eyebrows and eyelashes, nails were normal.

The facial features of individuals suffering from ectodermal dysplasia are consistently similar.The typical facies is characterised by frontal bossing, malar hypoplasia, flattened nasal bridge, reccessed colemella, thick everted lips, wrinkled hyperpigmented periorbital skin and prominent low set ears as was seen in Case 1. Skin over entire body is dry and hypopigmented, hair is sparse, unruly and lightly pigmented and eyebrows and eyelashes are sparse or absent, as was noted in both the cases. Anodontia or hypodontia with widely spaced peg shaped teeth are consistent features as was seen in Case 1.

In most of the cases, hypohidrotic ectodermal dysplasia is inherited as x-linked recessive trait with full expression only in male, however an autosomal recessive mode of inheritence may be operative in some families,[1] as probably occurred in the reported family.

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Reference

1. Esterly NB. Ectodermal dysplasia. In: Behrman RE, Kliegman RM, Nelson WE, Vaughen VC III, editors. Textbook of pediatrics. Philadelphia: Saunders, 1992: 1629.

ECTHYMA GANGRENOSUM WITHOUT BACTEREMIA

To the Editor,

A 30-year-old otherwise healthy housewife presented with two coin-sized punched out ulcers covered with grey-black eschar and surrounded by an erythematous halo over her right scapular region of 2 weeks duration without any preceding history of trauma or drug intake. It started as a painless red macule which enlarged, evolved into a haemorrhagic bulla which ruptured forming a gangrenous ulcer. Ulcers were non-tender and mobile over the underlying structures. Her vital parameters and systemic examination were normal and there was no evidence of bacteremia in the patient.

Haemogram, urinalysis, X-ray chest, blood sugar, VDRL test, ELISA for HIV, peripheral blood smear and blood culture were normal. Smear and culture from the skin lesions revealed isolation of P aeruginosa; and biopsy from the ulcer revealed bacterial vasculitis with a dense bacillary infiltration of the media and adventitia of blood vessels consistent with the diagnosis of ecthyma gangrenosum.

The four major dermatologic manifestations of severe systemic Pseudomonas aeruginosa infection are ecthyma gangrenosum, vesicular lesions,

cellulitis, and maculopapular plaques or nodules. Ecthyma gangrenosum is considered by many authors as pathognomic of Pesudomonas sepsis, though occurring in only 1.3% to 6% of patients with Pseudomonas bacteremia.[1] Although usually caused by P aeruginosa, it has been described in a case of P seudomonas cepacia endocarditis.[2] Lesions can occur anywhere but are usually found in the anogenital region, buttocks, extremeties, abdomen and axillae. In non-bacteremic ecthyma gangrenosum, the lesion is actually located at the site of entry of the organism into the skin; as opposed to classic ecthyma gangrenosum where the lesions represent a blood-borne matastatic seeding.[1]

This rare case of non-bacteremic ecthyma gangrenosum is consistent with earlier reports[1] having female predominance and better prognosis as compared to patients with Pseudomonas bacteremia, though occurring at a rare site.

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References

1. Huminer D, Siegman-Igra Y, Morduchowicz G, et al. Ecthyma gangrenosum without bacteremia-report of six cases and review of the literature. Arch Intern Med 1987; 147: 299-301.
2. Mandell JN, Feiner HP, Price NM, et al. Pseudomonas cepacia endocarditis and ecthyma gangrenosum. Arch Dermatol 1977; 113: 199-202.

SILENT RENAL CARCINOMA PRESENTING AS CUTANEOUS METASTASIS

To the Editor,

Cutaneous metastasis of renal adenocarcinoma is rare.[1] A 70-year -old woman came with history of restricted movement of left upper arm of 3 months duration. A hard mass was noticed measuring 8x8 cm in left scapular region fixed to underlying muscles. Skin over the mass was pinchable and normal. Patient was anaemic, not jaundiced and there was no generalized lymphadenopathy. There was no organomegaly. Bowels and micturition were normal. Abdomen was scaphoid, soft, and no mass was palpable. Renal angles were free. Biopsy of left scapular mass revealed clear cell type of carcinomatous cells in sheets, and glandular pattern; separated by thin fibrous septa. The nuclei were centrally placed. Ultrasound of abdomen revealed 6.4x4.1 cm size hypoechoic mass arising from the lower pole of left kidney. General condition of the patient deteriorated and she became unfit for surgery. The diagnosis of silent renal cell carcinoma of left kidney with cutaneous metastasis was made.

M Sikinder Hayath, M Janaki,
S V Ranga Reddy, Siva Kumar, G Baleswari,
A Bala Thimmaiah, O Joji Reddy
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Reference

1. Connor DH, Taylor HB, Helwig EB. Cutaneous metastasis of renal cell carcinoma. Arch Pathol 1963; 76: 339-46.

PAUCITY OF IMMUNE COMPLEXES IN SKIN LESIONS OF LICHEN PLANUS

To the Editor,

This is with reference to the article entitled "Paucity of immune complexes in skin lesions of lichen planus"[1] published recently in the Journal. We wish to share our experience on direct immunofluorescence (IMF) in lichen planus(LP). As reported by the authors we have also observed colloid bodies showing IgM, IgA, IgG and C3 deposits, however, in addition to the above we have consistently observed a