

Non-Pruritic Acral Rash in a Middle-Aged Male

A 56-year-old man presented with progressive rash on the face, hands and feet for over 6 months. He was treated with topical steroids cream and oral antihistamine pills, but no obvious improvement was achieved. His clinical history also included progressive hoarseness, stridor and a 10kg weight loss for 1 year. A diagnosis of primary squamous cell carcinoma (SCC) of the larynx was made a month before.

On physical examination, the patient had symmetrical desquamative, intensive violaceous erythema on the nose and cheek (Fig. 1) and in the helix and lobule of the ears (Fig. 2). His fingers were swollen and thickened with fine scales. The central palmarplantar skin was relatively spared. Prominent nail dystrophy, onycholysis, hyperkeratotic subungual debris and loss of cuticles were observed on all finger nails (Fig. 3). There was a 6 cm lump representing the cervical lymphadenopathy on the right neck. Skin biopsy from the right hand revealed non-specific findings including hyperkeratosis, acanthosis and slight dermal infiltration of lymphocytes. Laboratory tests including blood counts, liver function, chemistry panel and ANA (anti-nuclear antibody, antibodies against SS-A/Ro and SS-B/La) panel were unremarkable.

What is the likely diagnosis?

- A. Acrokeratosis paraneoplastica
- B. Dermatomyositis
- C. Cutaneous lupus erythematosus
- D. Hyperkeratotic eczema
- E. Palmoplantar psoriasis

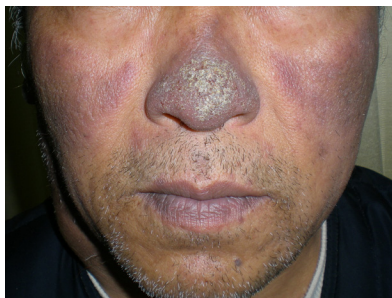


Fig. 1. Intensive violaceous erythema with scales and crust seen on the nose and cheek.



Fig. 2. Induration and violaceous erythematous swelling with scales on the right ear helix and lobule.



Fig. 3. Prominent nail dystrophy comprising yellow discoloration, vertical ridging and splitting, onycholysis, hyperkeratotic subungual debris and loss of cuticles.

Discussion

In the context of the patient's known malignancy, the typical clinical features and pathological finding were consistent with acrokeratosis paraneoplastica of Bazex.

Acrokeratosis paraneoplastica belongs to paraneoplastic skin diseases, and is most frequently associated with SCC—usually of the upper aerodigestive tract, and metastatic cervical lymphadenopathy.¹ Skin lesions tend to precede the tumour diagnosis by 1 year.² Symptomatic improvement can be achieved by effective treatment of the underlying tumour, and the relapse of skin lesions is associated with the recurrence of the tumour.³ Our patient received systemic chemotherapy and palliative radiation therapy but died 6 months later.

Answer: A

Options B, C, D and E are possible differential diagnoses for the initial clinical presentation. The commonest skin signs of dermatomyositis are Gottron's papules/sign, heliotrope rash and nail fold telangiectasia. Histology usually demonstrates interface dermatitis.⁴ These clinical and histological features were not identified in our patient. Moreover, he showed no muscle weakness and negative laboratory findings. Thus, dermatomyositis was excluded. Systemic lupus erythematosus (SLE) was ruled out because the patient had no symptoms of fever, oral ulcer, photosensitivity, joint pain and negative laboratory findings. Discoid lupus erythematosus (DLE) is another differential diagnosis. However, the lesions of DLE typically demonstrate atrophy, scarring, and areas of both hyperpigmentation and depigmentation. Besides, histological examination of DLE usually shows hydropic degeneration of the basal layer of the epidermis and patchy perivascular and periadnexal lymphoid inflammatory infiltrate. Clinically, the skin lesions of our patient were non-pruritic, progressively thickening (without improvement to topical steroids therapy) and atypically distributed on the ears and nasal tip which are not consistent with that of eczema or psoriasis. In contrast, prominent nail changes and unusual distribution of violaceous erythematous swelling with scales on the ear and nasal tip are the hallmark of acrokeratosis paraneoplastica.

Acrokeratosis paraneoplastica is rare and only a limited number of cases have been reported so far in the literature. A

case is presented here to highlight that prompt recognition of acrokeratosis paraneoplastica may lead to the earlier detection and potential treatment of an underlying malignancy.

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