CASES FOR DIAGNOSIS

Acral Psoriasiform Lesions

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Clinical History

The patient was a 55-year-old man who was admitted for study of a constitutional syndrome; in addition, he reported a 3-month history of skin lesions. There was no past history of interest except that the patient was a drinker and smoked more than 40 cigarettes per day.

Physical Examination

The patient presented symmetrical, erythematous-violaceous plaques on the distal phalanges of the fingers and toes; the plaques had a hyperkeratotic and desquamative surface (Figures 1 and 2). There was also slight reddening and desquamation of the lobes of the ears. In addition, the patient was asthenic, with mild jaundice of the skin and mucosas, and presented hepatomegaly and palpable right lateral cervical lymph nodes that formed a mass of approximately 4 by 3 cm (Figure 3).



Figure 1.

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Accepted for publication September 26, 2008.

Additional Tests

Computed tomography of the neck revealed a tumor in the oropharynx with lymph node masses in the internal jugular chain and posterior cervical space. Biopsy of a lateral cervical lymph node was compatible with metastasis from an undifferentiated carcinoma of nasooropharyngeal origin.



Figure 2.



Figure 3.

What Was the Diagnosis?

Diagnosis

Paraneoplastic acrokeratosis of Bazex.

Clinical Course

The patient started the first cycle of adjuvant chemotherapy, but died after 15 days due to neutropenic sepsis.

Discussion

Paraneoplastic acrokeratosis of Bazex is a rare paraneoplastic syndrome that mainly affects men over 40 years of age; it is usually associated with squamous cell carcinoma of the upper respiratory and digestive tracts. Karabulut et al² performed a review of 133 patients with this syndrome and found that the primary tumor was a squamous cell carcinoma of the oropharynx or larynx in 74 cases (more than 50% of the patients), undifferentiated carcinoma of the oropharynx or larynx in 19 cases, and adenocarcinoma—most commonly of the lung, prostate, or esophagus—in 15 cases.

The pathogenesis of Bazex syndrome is not fully understood. Possible mechanisms include cross-reactivity between tumor antigens and the skin and secretion by the tumor of factors analogous to epidermal growth factor, which could be responsible for epidermal hyperproliferation.³

The characteristic skin changes are symmetric, nonpruritic, erythematous or violaceous, scaly plaques; the main differential diagnosis is with psoriasis. The rash develops in 3 phases: it initially develops on the acral tissues, affecting the fingers and toes, although the lesions can also appear on the ears and nose, and the early plaques are hardly raised or infiltrated and have poorly defined borders; in the second stage, the skin lesions extend to the elbows, knees, and pretibial regions, and are frequently associated with palmoplantar keratoderma over the pressure areas; in the third stage, if the tumor has not been found and treated, the rash extends to the trunk and scalp.

Nail changes are observed in 75% of cases and usually appear early; there is subungual hyperkeratosis, onycholysis, longitudinal and horizontal streaks, and yellowish discoloration. Paronychia can be the first sign

of nail involvement. Onychomadesis has been reported occasionally,⁴ as has the presence of tense, subepidermal blisters over the typical lesions of paraneoplastic acrokeratosis.⁵

Histology is nonspecific: there may be hyperkeratosis, parakeratosis, acanthosis, and a perivascular lymphocyte infiltrate, though there are also reports of vacuolar degeneration of the basal layer, dyskeratosis, and melanophages in the dermis.

Complete resolution of the lesions requires treatment of the tumor. However, the nail changes frequently persist. Traditional dermatological treatments (topical corticosteroids, keratolytics, emollients, etc) have not been found to be effective; best results are achieved with phototherapy (psoralen-UV-A).⁶ Reappearance of the skin lesions may indicate recurrence of the tumor.

In conclusion, we would like to draw attention to the importance to the dermatologist of recognizing this syndrome, as early diagnosis of the skin lesions can improve the management of the underlying tumor and thus improve the patient's quality of life.

Conflicts of Interest

The authors declare no conflicts of interest.

References

- Bazex A, Salvador R, Dupré A, Christol B. Syndrome para néoplasique à type d'hyperkératose des extrémités. Guérison après le traitement de l'epithélioma laryngé. Bull Soc Fr Derm Syphiligr. 1965;72:182.
- 2. Karabulut A, Sahin S, Sahin M, Eksioglu M, Ustun H. Paraneoplastic acrokeratosis of Bazex (Bazex's syndrome): Report of a female case associated with cholangiocarcinoma and review of the published work. J Dermatology. 2006;33:850-4.
- 3. Mutasim DF, Meiri G. Bazex syndrome mimicking a primary autoimmune bullous disorder. J Am Acad Dermatol. 1999;40:822-5.
- 4. González-Herrada CM, Frías J, Casanova JM, et al. Acroqueratosis paraneoplásica de Bazex. Aportación de un nuevo caso. Actas Dermosifiliogr. 1984;75:301-6.
- Cabanillas M, Pérez-Pérez L, Sánchez Aguilar D, Fernández-Redondo V, Toribio J. Acroqueratosis paraneoplásica con lesiones ampollosas asociada a carcinoma epidermoide esofágico. Actas Dermosifiliogr. 2006;97:196-9.
- Gill D, Fergin P, Kelly J. Bullous lesions in Bazex syndrome and successful treatment with oral psoralen phototherapy. Australas J Dermatol. 2001;42:278-80.