SKIN

BRIEF ARTICLE

# Bazex Syndrome Associated with Angioimmunoblastic T-cell Lymphoma

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## ABSTRACT

Bazex syndrome, also known as Acrokeratosis paraneoplastica, is a paraneoplastic disorder characterized by erythematous psoriasiform plaques involving the nose, ears, and acral sites. Although classically associated with squamous cell carcinomas of the upper aerodigestive tract, it has also been reported in association with adenocarcinoma, genitourinary tumors, multiple myeloma, and rarely, peripheral T-cell lymphoma and follicular lymphoma in-situ. Herein, we present a patient with Bazex syndrome associated with angioimmunoblastic T-cell lymphoma (AITL), a rare association not previously reported in the literature.

## INTRODUCTION

Bazex syndrome was first described by Bazex *et al*. in 1965 in association with malignancy of the upper aerodigestive tract.[1] The main features are the presence of symmetrical psoriasiform eruptions, nail dystrophy, and xerotic scaling usually accentuated on the acral surfaces, ears, and nose.[2]

Several mechanisms for development of Bazex syndrome have been proposed. One theory suggests that antibodies against the tumor cross react with the keratinocytes or basement membrane leading to damage of the basal layer of the skin. Alternatively, an immune reaction directed against tumor-like antigens in the epidermis could be responsible for the cutaneous eruptions. Association with cutaneous squamous cell carcinoma, Hodgkin's disease, peripheral T-cell lymphoma, and follicular lymphoma in-situ have been reported.[3] We present a unique case of Bazex syndrome associated with AITL. To our knowledge, this is the first reported case of Bazex syndrome seen in association with AITL.

## CASE REPORT

A 60-year-old male presented with a three-month history of fever, diarrhea, xerosis, and a 20-lb weight loss. The patient reported a several-week history of a non-pruritic rash on his face and extremities.

On physical exam, xerotic scaly plaques were noted on the ears, nose, and scalp, with ill-defined xerosis and adherent scale of the lower legs. Desquamating keratoderma was seen on the palms, soles, and dorsal hands, along with Beau's lines of the fingernails

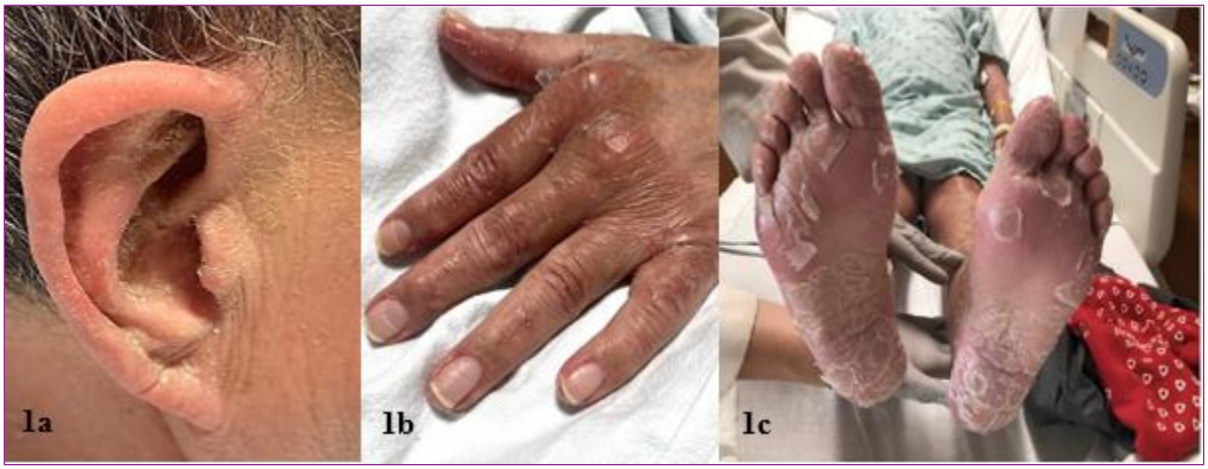


**Figure 1. (A)** Psoriasiform eruption of ear **(B)** Beau's lines on fingernails and scaling of hands **(C)** Xerotic scaly plaques on plantar feet

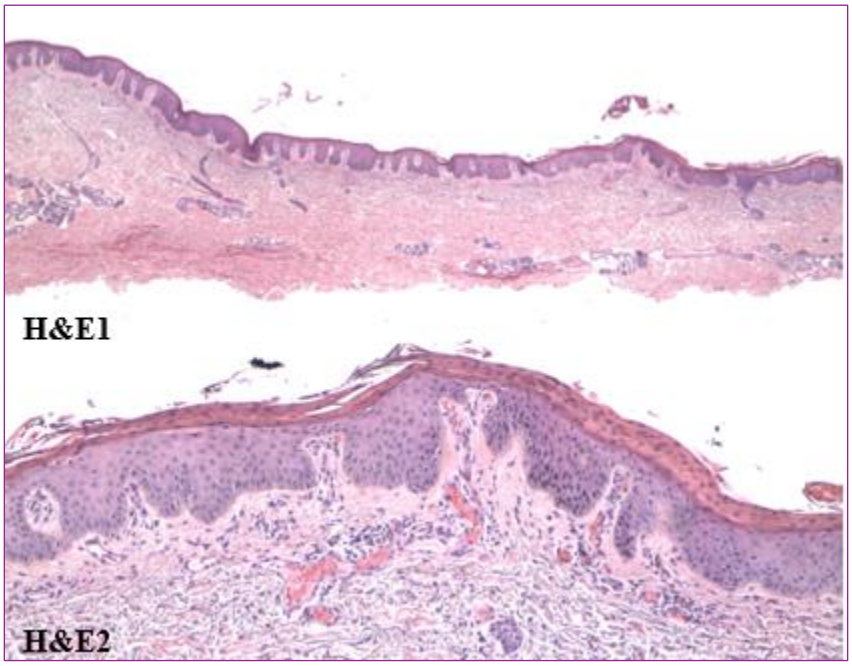


**Figure 2. H&E1 (20x):** Psoriasiform acanthosis with broad and blunt rete ridges, **H&E 2 (200x):** Confluent parakeratosis and underlying hypogranulosis

bilaterally (**Figure 1**). Diffuse lymphadenopathy was appreciated. CT scans revealed extensive lytic lesions throughout the spine, ribs, and scapula.

Lymphoma was suspected and a lymph node biopsy revealed AITL. A tangential biopsy and punch biopsy were performed, revealing psoriasiform acanthosis with broad and blunt rete ridges, along with hypogranulosis, confluent parakeratosis, and minimal inflammatory infiltrate (**Figure 2**). The histological findings and clinical picture were consistent with Bazex syndrome due to underlying AITL. The patient was then started

on topical steroids and urea with some improvement. However, the patient expired from angioimmunoblastic T-cell lymphoma shortly thereafter.

## DISCUSSION

Bazex syndrome is a paraneoplastic disorder of the skin characterized by erythematous psoriasiform plaques in an acral distribution that classically also involve the nose and ears.

Histopathologic features of Bazex syndrome are variable, but usually include acanthosis, hyperkeratosis, and parakeratosis. Inconstant features include variable spongiosis and dyskeratosis.[4-5] While the histologic features are not always diagnostic, the absence of diagnostic features of other conditions and clinicopathologic correlation are key to rule out other entities on the differential diagnosis. In our patient, the presence of confluent parakeratosis excluded the possibility of malignancy-associated ichthyosis. Furthermore, the absence of other diagnostic features of psoriasis (i.e., neutrophilic microabscesses), in conjunction with the patient's clinical presentation, excluded this possibility as well. The favored therapy for this paraneoplastic dermatosis is effective treatment of the underlying malignancy. For recalcitrant or palliative care, systemic treatments include corticosteroids, fluconazole, zinc, and cephalexin. Topical treatments include retinoids, zinc ointment, PUVA, and emollients.

Although Bazex syndrome is most commonly associated with squamous cell carcinoma of the upper aerodigestive tract, it has also been reported in association with adenocarcinoma, genitourinary tumors, and rarely, peripheral T-cell lymphoma and follicular lymphoma. Herein, we present the first case of Bazex syndrome seen in association with AITL.

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