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Journal

Dermatology Online Journal, 30(2)

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Publication Date

2024

DOI

10.5070/D330263580

Acrokeratosis paraneoplastica (Basex syndrome) with trachyonychia preceding the diagnosis of squamous cell carcinoma of the lung

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Abstract

Acrokeratosis paraneoplastica (Basex syndrome) is a rare paraneoplastic condition hallmarked by psoriasiform lesion development on acral surfaces, most often related to an underlying squamous cell carcinoma. Patients may also present with nail plate changes. Successful management of this condition can be accomplished by treating the underlying malignancy.

Keywords: acrokeratosis paraneoplastica, bazex syndrome, cancer, lung, malignancy, paraneoplasia, paraneoplastic, psoriasiform

Introduction

Acrokeratosis paraneoplastica, commonly referred to as Bazex syndrome, is a rare paraneoplastic condition characterized by the development of psoriasiform plaques on acral surfaces, such as ears, hands, and feet [1, 2]. Bazex syndrome is often seen in association with squamous cell carcinoma (SCC) of the upper gastrointestinal tract [1, 2]. Less commonly, Bazex syndrome has been reported in association with adenocarcinomas of genitourinary tract, breast cancer, sarcoma, or hematologic malignancies. Bazex syndrome can be difficult to treat, but generally resolves after successful treatment of the underlying malignancy.

Case Synopsis

A 62-year-old man presented to dermatology clinic for evaluation of a rash on his hands and feet, present for ten months. The affected areas were itchy and scaly. He noted fingernail brittleness that started around the same time as his rash. He had recently been admitted for bacterial and fungal infections of the hands and nails. Treatment with systemic antibiotics and topical antifungals did not improve his symptoms.

Physical examination revealed well-demarcated, erythematous hyperkeratotic scaling plaques limited to the palms (**Figure 1A**) and soles. Nail changes included trachyonychia in 9 of 10 fingernails (**Figure 2A**). He did not have any skin changes on the nose, ears, or penis. Tangential biopsy of the palm demonstrated spongiotic and psoriasiform dermatitis, with prominent superficial perivascular lymphohistiocytic infiltrate with occasional eosinophils (**Figure 3**). Periodic acid-Schiff staining for fungal elements was unremarkable.

Two months prior to his initial dermatology visit, he was diagnosed with SCC of the left mainstem bronchus after presenting with left sided chest pain and dyspnea. Chemotherapy (carboplatin/paclitaxel) was started around the time of his initial dermatology visit. This was not tolerated and he was then treated with radiation therapy with a dose of 6,000cGy over 63 days.

A diagnosis of Bazex syndrome was rendered. He was started on clobetasol 0.05% ointment twice daily under occlusion, which led to gradual improvement in pruritus and scaling. At 11 weeks post-radiation, the proximal half of previously affected nail plates lacked pathology, with evident Beau lines visible (**Figure 2B**). At this time, he still had some pruritus

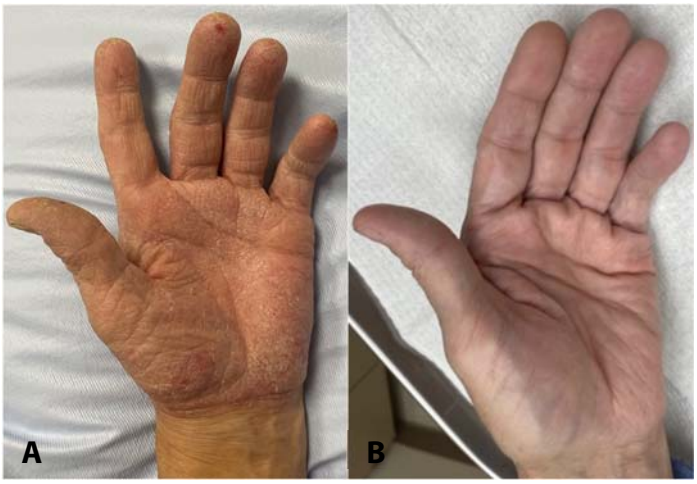

Figure 1. *Skin manifestations of acrokeratosis paraneoplastica, or Basex syndrome.* ***A)*** *Well-demarcated erythematous scaling plaques to the palm before, and* ***B)*** *52 weeks after radiation treatment.*

and scaling, relieved with as-needed use of clobetasol. At 23 weeks post-radiation (no image available), his palms were clear and the affected nail plates were relatively normal appearing, with some pitting (**Figure 2C**). After 52 weeks post-radiation, his palms remained clear (**Figure 1B**) and nail plates appeared healthy (**Figure 2D**), and he had no further scaling, pruritus, or need for topical corticosteroids. A follow-up computed tomography scan of the chest at 52 weeks was negative for cancer.

Case Discussion

The diagnosis of Bazex syndrome is made based on history and clinical presentation. Bazex syndrome should be suspected when a patient presents with new onset psoriasiform plaques on the nose or helices of the ears, as involvement of these sites would be exceedingly rare for other psoriasiform conditions [2]. Bazex syndrome may also be considered when a patient develops new psoriasiform changes isolated to other acral surfaces or the nails, or in a patient with psoriasiform plaques that are resistant to traditional treatments [1, 2]. Suspicion for Bazex syndrome should prompt a workup for an underlying malignancy. In our case, the patient first developed cutaneous and nail findings 10 months prior to the diagnosis of his underlying malignancy; however, he was initially diagnosed with and treated for bacterial and fungal infections for months without relief. In a majority of cases (63-79%), cutaneous findings preceded the diagnosis of cancer by an average of 11 months [1-3]. Early recognition of paraneoplastic conditions is essential to allow for earlier malignancy diagnoses and to reduce morbidity and mortality.

There are no features specific to Bazex syndrome on histopathology. Common histologic findings include parakeratosis, acanthosis, spongiosis, dyskeratosis, and vacuolar changes of the epidermal basal layer. There are variable inflammatory infiltrates which can include eosinophils [4]. The presence of dyskeratosis and interface changes can help to differentiate this condition from psoriasis, which is often suspected owing to similar clinical appearance [2]. Our case displayed spongiotic and psoriasiform changes with superficial perivascular lymphohistiocytic infiltrate and occasional eosinophils (**Figure 3**). Although the mechanism behind these histopathologic changes is poorly understood, it has been proposed that the

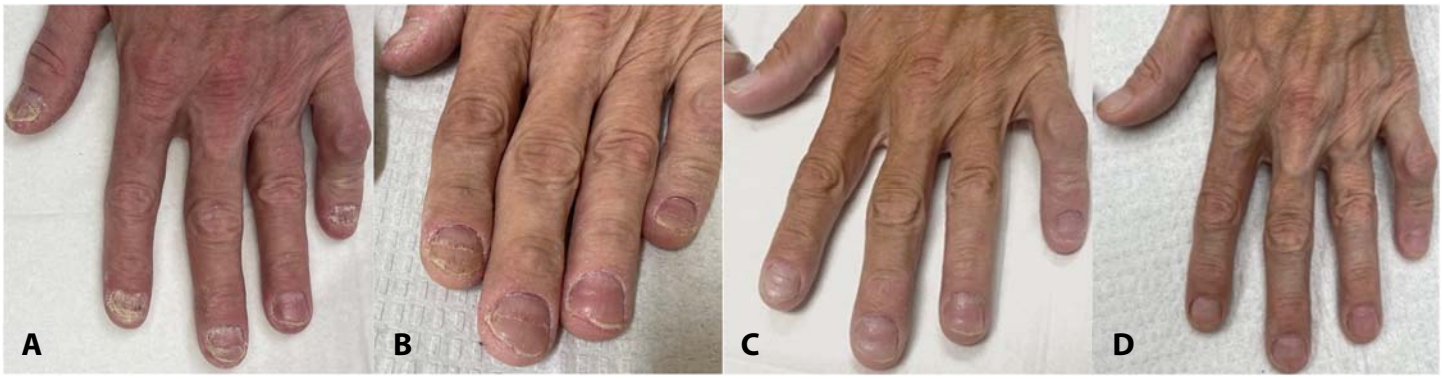

Figure 2. *Nail manifestations of acrokeratosis paraneoplastica, or Basex syndrome.* ***A)*** *Onychorrhexis of nails before treatment, and* ***B)*** *with evidence of healthy proximal nail plate growth at 11 weeks post-radiation.* ***C)*** *Maintenance of non-pathologic nails at 23 weeks, and* ***D)*** *52 weeks post-radiation.*

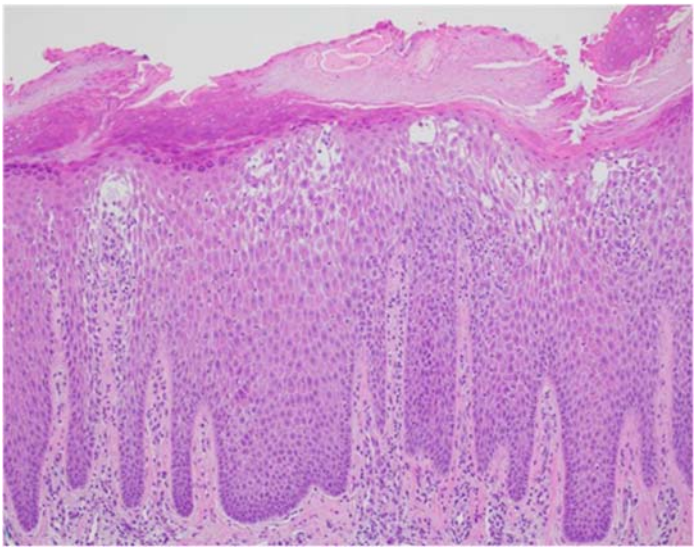

Figure 3. *Histopathology of palmar tangential biopsy. Prominent spongiosis with intraepidermal vesiculation, psoriasiform hyperplasia, parakeratosis and serum in the stratum corneum. Within the dermis, there is a superficial perivascular lymphohistiocytic inflammatory infiltrate with occasional eosinophils. H&E, 100×.*

psoriasiform changes may relate to direct keratinocyte stimulation via growth factors secreted by SCC such as epidermal growth factor, which shares structural and functional similarities to transforming growth factor α and insulin-like growth factor 1 [2]. We were unable to measure these factors in our patient.

Nail changes are present in 75% of patients with Bazex syndrome [2]. The most common nail findings in Bazex syndrome include thickening/onychauxis (35%), subungual hyperkeratosis (31%), yellow/brown discoloration (27%), onycholysis (27%), longitudinal ridging (25%), and loss of nail plate (13%), [3]. Our patient's nail plates were severely affected, clinically consistent with trachyonychia. Many of these features are shared with more common nail diseases such as psoriasis and onychomycosis, the latter of which was one of our patient's previous diagnoses, contributing to a delay in accurate diagnosis. Although onychomycosis commonly affects a few nails or just one hand, Bazex syndrome typically affects all fingernails. Our patient's clinical course has been followed and nail changes have been documented, demonstrating healthy nail plate growth that is sustained after successful treatment of his associated malignancy.

Effective management for Bazex syndrome includes treatment of the underlying malignancy, as this leads to resolution of cutaneous symptoms in greater than 74% of cases [2]. Topical treatments are generally ineffective, and a lack of response to topical therapy may be a feature of the disease [1]. Topicals may be used for symptomatic relief and our patient found relief of pruritus with clobetasol use. Even with treatment of the cancer, nail symptoms commonly persist despite resolution of skin disease [3]. At one-year post-radiation, our patient no longer had scaling, pruritus, or a need for topical corticosteroids. His nails remained clear. Recurrence of the cutaneous symptoms following the treatment of the cancer should prompt evaluation for recurrence of malignancy [1,2].

Conclusion

Early recognition of paraneoplastic phenomena can be life-saving and new-onset acral psoriasiform plaques or nail changes should prompt suspicion for Basex syndrome. If Basex syndrome is suspected, an appropriate workup for malignancy should be completed. Basex syndrome can be successfully managed by treatment of underlying malignancy.

Potential conflicts of interest

The authors declare no conflicts of interest.

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