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Letter

Acitretin amelioration of Acrokeratosis Paraneoplastica (Bazex Syndrome) in cases of incurable squamous cell carcinoma of the hypopharynx

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Abstract

BACKGROUND

Acrokeratosis paraneoplastica (Bazex Syndrome) is a rare paraneoplastic syndrome and dermatosis that only arises in patients with underlying malignancy and uncommonly resolves with systemic therapy.

OBJECTIVE/METHODS

We present a patient with acrokeratosis paraneoplastica that improved significantly with acitretin. We present evidence to justify costs of therapy for insurance purposes. Additionally, there is a single report of acitretin use for Bazex syndrome in the French language.

RESULTS

We present a case of acrokeratosis paraneoplastica in a patient with incurable stage IV squamous cell carcinoma of the hypopharynx that significantly improved on acitretin.

CONCLUSION

Although acrokeratosis paraneoplastica most often is cured by treatment of the underlying squamous cell carcinoma, this case highlights the potential benefit of early initiation of acitretin during malignancy work up and staging. This therapy may also be valuable for patients in which the primary malignancy is unresectable or incurable.

Case synopsis

A man in his 60s presented with a seven month history of nail thickening in conjunction with a scaling dermatitis over his nose, ears, and dorsal hands (Figure 1).



Figure 1. Initial presentation (Left) and two month improvement (Right).

He complained of a burning sensation in the affected areas. The violaceous plaques spread to his lower extremities and back, with later extension to his ears, nose, and scalp. Soon after, he developed a neck mass and was later diagnosed with squamous cell carcinoma of the hypopharynx, which was found to be stage IV. He was also diagnosed with acrokeratosis paraneoplastica and owing to significant burning parasthesias of his skin, treatment with acitretin was initiated. He underwent two months of therapy prior to starting cisplatin and radiotherapy. He was seen in follow up at that time with considerable improvement of skin lesions on the hands, feet, elbows, knees, and nails. After three months and substantial improvement, the dose was decreased from 25 mg daily to 10 mg three times weekly.



Figure 2. Initial presentation (Left), and two month improvement (Right).

A woman in her 50s presented with new onset pain in her hands after being diagnosed with stage IVA incurable squamous cell carcinoma of the hypopharynx with adjacent metastases. She had just begun treatment with concomitant cisplatin and radiation therapy when the erythema, scaling, and brittle nails first developed (Figure 2). She had no involvement of her face or ears. She did have dry gangrene present on her 3rd digit that developed at the same time. She was diagnosed with acrokeratosis paraneoplastica and started on acitretin at 10 mg three times weekly. When she returned two months later, she also noted substantial improvement in her burning parasthesias, erythema, brittle nails, and dry gangrene.

Discussion

Bazex syndrome is a rare paraneoplastic syndrome and dermatosis that usually affects white men age 40 or older [1, 2]. It is found as a paraneoplastic syndrome in squamous cell carcinomas of the upper respiratory or digestive tract, but has been reported in other malignancies as well, including breast, lung, prostate gland, esophagus, stomach, colon, and bladder [1, 2, 3]. No cases have been reported without an underlying malignancy [4]. Initially, there is an erythematous, hyperkeratotic desquamating eruption that affects acral sites such as fingers and toes, followed by involvement of the ears, nose, and periungual hyperkeratosis with onycholysis and ungula dystrophy [1]. Eventually, erythematous hyperkeratotic lesions spread to the trunk and extensor surfaces [3]. In 2/3 of cases, the cutaneous manifestations precede the diagnosis of cancer by about one to three years and the primary malignancy tends to become symptomatic during the palmoplantar manifestation [3]. Although pathogenesis is unknown, one theory hypothesizes cross reactivity between the tumor and skin antigens, leading to activation of the cellular immune system against the skin; another theory hypothesizes that substances secreted by the tumor could play a role [3, 5].

Generally, histopathologic findings are nonspecific, but include spongiosis and interface change in the epidermis and hyperkeratotic epithelium with focal areas of parakeratosis, acanthosis, dermal perivascular infiltrate, and diffuse lymphocytic infiltrate in the upper dermis [1, 2].

In 90-95% of patients, treatment of the underlying malignancy results in complete resolution of the lesions, whereas response to topical and systemic treatments is inconsistent [1]. Some patients have improved with the use of dexamethasone [4]. Elevations of liver enzymes may occur in up to 15% of patients receiving acitretin, although it is usually transient and reversible [6]. We note two unique aspects about these cases. First, the cancers were not adequately treated by the chemotherapy. Second, although the acitretin was started several months prior to initiation of therapy, substantial improvement still occurred in our first patient and the skin involvement in the second patient did not develop until after the initiation of her chemotherapy. Acitretin has the benefit of being both non-immunosuppressive and commonly used in the treatment of psoriasiform and palmoplantar diseases.

Conclusions

Although acrokeratosis paraneoplastica is generally treated by targeting the underlying malignancy, these cases highlight the benefit of early initiation of acitretin during malignancy work up and the benefit in cases of incurable primary malignancy.

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Author Contributions

Dr(s) Benjamin Kaffenberger, Polina Vaynshtok had full access to all of the data in the study and take responsibility for the integrity of the data and the accuracy of the data analysis. *Study concept and design*: Benjamin Kaffenberger, Polina Vaynshtok. *Acquisition, analysis, and interpretation of data*: Benjamin Kaffenberger, Polina Vaynshtok. *Drafting of the manuscript*: Polina Vaynshtok. *Critical revision of the manuscript for important intellectual content*: Benjamin Kaffenberger. *Statistical analysis*: None. *Obtained funding*: None. Administrative, technical, or material support: Benjamin Kaffenberger. Study supervision: Benjamin Kaffenberger.

