Case Report: Atypical Ulceronecrotic Lymphomatoid Papulosis Treated Successfully with Brentuximab Vedotin

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Abstract:
A 68-year-old Caucasian woman presented with a six-month history of a vesicular and ulcerative eruption on the bilateral hands associated with significant pain and disability. She also had a worsening ulcerative and crusted eruption on the chest and back with similar symptoms. Because of concern for eczema herpeticum, tissue PCR for HSV and VZV were performed and were negative. Skin biopsies performed from multiple sites demonstrated an atypical lymphocytic infiltrate with increased CD4:CD8 of 4:1 and large cells with CD30-positivity. She was started on methotrexate 20mg weekly and an oral prednisone taper with only modest response. Brentuximab vedotin at 1.2mg/kg every three weeks was initiated, with complete resolution of all lesions. Treatment was continued for a total of six cycles and the patient achieved a sustained remission of nearly one year.

Case Presentation:
The patient is a 68-year-old Caucasian female who presented with a 10-year history of a waxing and waning polymorphic eruption. Lesions were red, scaly and pruritic plaques located on trunk and extremities. Also present were vesicles and ulcerative papules most prominent on the hands and wrists, but also scattered throughout the trunk and arms. Patient had recurrent skin infections secondary to skin breakdown. Severe pain from ulcerations on the hands resulted in disability. Many non-diagnostic biopsies were performed. However, repeat biopsy of a lesion on the left breast demonstrated ulcerated and necrotic skin with atypical cellular infiltrate. The infiltrate stained positively with CD3, CD45, CD4, CD5, CD7, and CD8 with a CD4:CD8 ratio of 4:1. A scant number of CD20 positive B-cells were observed. CD30 marked many of the enlarged mononuclear cells forming loose aggregates within the dermis. CD15 marked some of these cells as well. CD1A highlighted mild hyperplasia of Langerhans cells in the epidermis and dermis. No significant reaction for CD56, EMA, pancytokeratin, CD31, and S100 was observed. Direct immunofluorescence demonstrated no patterned deposition for albumin, IgG, IgM, C3, and C5b-9. Nonspecific C5b-9 was demonstrated in endothelia. CBC and CMP were non-contributory. Given the clinical history and biopsy demonstrating CD30+ lymphoproliferative disorder the patient was diagnosed with Ulceronecrotic CD30+ Lymphoproliferative disorder.

Clinical Response to Treatment:

Figure 1. Punch-out ulcerative and necrotic papules on bilateral hands (i-c) and red scaly pruritic plaques with honey-colored crusts on trunk and extremities (d)

Figure 2. Resolution of all lesions on bilateral hands with residual post-inflammatory hyperpigmentation

Discussion and Conclusion:

LyP proves a diagnostic challenge with one study demonstrating an average of 45 months from onset of symptoms to diagnosis. Histologically, LyP is divided into five subtypes that most commonly demonstrate an infiltrate of CD30-positive T-cells. However, CD30-negative variants, CD8-positive cytotoxic T-cell variants, and angioinvasive variants are described. There is much overlap in the histological presentation of these subtypes. LyP has an excellent prognosis with a 5-year disease specific survival of 100%. However, there is debate in the literature regarding the individual patient’s risk of developing a secondary cutaneous lymphoma or systemic lymphoma. Depending on the study, 10-60% will develop another lymphoma such as systemic ALCI, Hodgkin’s, or mycosis fungoides.

Commonly prescribed treatments include: MTX, PUVA, topical steroids. Effective treatment for severe forms of LyP has been lacking.

Our patient completed 6 cycles of Brentuximab vedotin at reduced dose of 1.2mg/kg with complete resolution of the lesions on the hands. Methotrexate was continued at 20mg per week with folate supplementation.

We demonstrate that reduced-dose of 1.2mg/kg was as effective as the full dose of 1.8mg/kg.

In conclusion, severe LyP cases have difficult to treat with historical treatment routes. This case demonstrates an atypical presentation of LyP and its successful treatment with BV with excellent results.

References: