Lymphomatoid papulosis successfully managed with excimer laser maintenance therapy

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INTRODUCTION

Lymphomatoid papulosis (LyP) is a chronic $CD30^+$ cutaneous lymphoproliferative disorder characterized by recurrent papulonodular and papulonecrotic eruptions. It is considered a low-grade variant of cutaneous T-cell lymphoma (CTCL). Although the disease course is frequently benign, 4% to 25% of cases are associated with secondary cutaneous or nodal lymphoid malignancies, including Hodgkin lymphoma, mycosis fungoides, and cutaneous or nodal anaplastic large cell lymphoma.[1,2] There are five documented LyP histologic subtypes (A-E). The most common subtype, type A (histiocytic type), is characterized by a wedge-shaped infiltrate of atypical $CD4^+$ lymphocytes, eosinophils, and histiocytes.[3] Type B (lymphocytic type), which can resemble mycosis fungoides, is characterized by a monomorphous infiltrate of small-to-medium–sized lymphocytes with cerebriform nuclei.[3] Type C can resemble anaplastic large cell lymphoma, presenting with sheets of large $CD30^+$ cells.[3] Types D and E are rare, characterized by histopathologic similarities to primary cutaneous aggressive epidermotropic $CD8^+$ cytotoxic T-cell lymphoma (type D) and angiodestructive infiltrates of small-to-medium–sized lymphocytes that are $CD30^+$ and $CD8^+$ (type E).[4,5] Unfortunately, few efficacious treatment options for LyP are currently available, none of which are curative. LyP is also often misdiagnosed on presentation, and symptoms can persist for 1 to 3 years before it is correctly diagnosed and appropriately treated.[2]

Abbreviations used:

CTCL:	cutaneous T-cell lymphoma
LyP:	lymphomatoid papulosis
NB-UVB:	narrow-band ultraviolet B
PUVA:	psoralen with ultraviolet A

CASE REPORT

We present a now 55-year-old woman with a longstanding history of atopic dermatitis who originally presented in 2002 after developing new, erythematous, papulonodular lesions on her trunk and extremities during pregnancy. At that time, biopsies of the patient's back found spongiotic dermatitis, whereas biopsies on her legs found dense, nodular, wedge-shaped infiltrates of lymphocytes, neutrophils, and eosinophils in the papillary and reticular dermis. The lymphoid component was heterogeneous and included clusters of enlarged mononuclear cells with atypical, pleomorphic nuclei that were strongly $CD30^+$ and $CD4^+$ on immunohistochemical staining. These findings, in conjunction with the patient's clinical presentation of relapsing and remitting papulonodular lesions, were consistent with LyP. The patient was given clobetasol, with little improvement to the nodules, although her eczematous areas responded. Although the patient's disease course remained relatively stable for 3 years following her pregnancy, it later accelerated in 2007. The patient re-presented to the clinic at this time with new crops of erythematous, excoriated papules on her left jawline, bilateral forearms, and right upper arm. She noted that these lesions were often pruritic and sensitive to contact with fabric from her clothing. Repeat biopsies were again consistent with LyP.

The patient started treatment with narrow-band ultraviolet B (NB-UVB) using excimer laser

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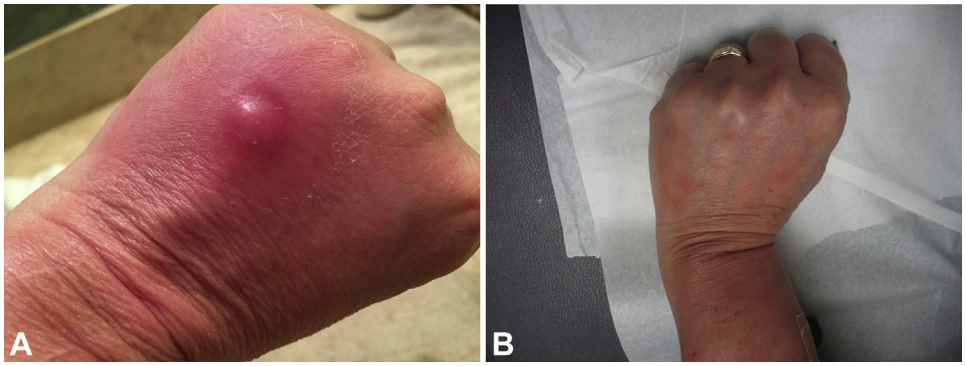

Fig 1. A, Lymphomatoid papulosis nodule on dorsal surface of patient's left hand before treatment with excimer laser. **B**, Lymphomatoid papulosis nodule resolution after treatment sessions with excimer laser.

(XTRAC; PhotoMedex, Inc., Boston, MA). She was treated twice per week with a starting dose that varied (319 mJ to 519 mJ) depending on the size of each nodule. From 2009 to 2012, she continued excimer treatment twice per week to new nodules at low doses ranging from 319 mJ to 464 mJ.

Within the first 2 weeks, the patient noticed decreased pruritus, pain, discomfort, and size of nodules. Most nodules cleared within 6 to 10 treatments (Fig 1), whereas untreated nodules persisted for many months. The patient experienced temporary darkening of the skin in the treatment areas (her skin type is Fitzpatrick IV). This finding may represent a combination of tanning in the treatment areas and postinflammatory hyperpigmentation from the LyP and/or the treatment itself. She also experienced blistering on occasion when treated at doses greater than 464 mJ. The patient continued to have new outbreaks with similar frequency to those before initiating treatment. She is currently on a maintenance excimer dose of 403 mJ. Large, painful nodules are treated at 638 mJ (319 mJ double pulsed) as needed based on the patient's request for treatment. The 638-mJ dose has often resulted in slight blistering but significantly decreases nodule size and discomfort for the patient.

DISCUSSION

Excimer laser involves the dissociation of xenon and chloride gas, which creates a 308-nm monochromatic light capable of targeted induction of apoptosis and reduced proliferation of T lymphocytes.[6] This process is mediated by DNA absorption of UVB radiation with subsequent DNA breakage. Excimer has been used successfully to treat localized vitiligo and psoriasis, with lower cumulative doses of UVB and fewer treatment sessions required for clearance of psoriasis compared with whole-body NB-UVB.[6] When compared with whole-body NB-UVB for the treatment of LyP, excimer laser has multiple benefits, which include the ability to tailor treatment to individual nodules depending on size (such as the 319-mJ double-pulsed dose used for larger nodules in this case), treatment of difficult-to-reach areas of the body, such as the postauricular region or the scalp, and utility in patients for whom whole-body NB-UVB is physically burdensome. To our knowledge, only 2 cases using excimer laser to treat LyP have been reported.[7,8] Of these, one case involved treatment of a solitary lesion and the second was limited to 13 total treatments. Our case, however, shows that long-term maintenance therapy with excimer for LyP is possible. Both methotrexate and PUVA (psoralen with ultraviolet A therapy) have been used to treat LyP. However, both have limitations when compared with excimer laser; methotrexate with undesirable side effects, particularly for patients with few LyP lesions who prefer targeted therapy, and PUVA with increased risk of skin cancer. In our patient's case, methotrexate was contraindicated, as she was pregnant at the time of LyP onset. A review of LyP therapies determined that both methotrexate (n = 79) and PUVA (n = 19) have recurrence rates of 63% and 84%, respectively.[9] Other treatment options in the literature, such as intralesional corticosteroid injections, have not been found effective for the treatment of LyP. Although treatment with excimer in our patient was not curative, our case shows that for LyP patients, eximer laser potentially represents an excellent option for maintenance therapy that can be tailored to individual lesions and patient preference.

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