Spontaneous regression of primary cutaneous diffuse large B-cell lymphoma, leg type with significant T-cell immune response

Paul M. Graham, DO,[a] Adam S. Richardson, DO,[a] Brian L. Schapiro, MD,[b] Mark D. Saunders, MD,[c] and Daniel M. Stewart, DO[a]
Ann Arbor and Williamsburg, Michigan

We report a case of histologically confirmed primary cutaneous diffuse large B-cell lymphoma, leg type (PCDLBCL-LT) that subsequently underwent spontaneous regression in the absence of systemic treatment. The case showed an atypical lymphoid infiltrate that was $CD20^+$ and $MUM\text{-}1^+$ and $CD10^-$. A subsequent biopsy of the spontaneously regressed lesion showed fibrosis associated with a lymphocytic infiltrate comprising reactive T cells. PCDLBCL-LT is a cutaneous B-cell lymphoma with a poor prognosis, which is usually treated with chemotherapy. We describe a case of clinical and histologic spontaneous regression in a patient with PCDLBCL-LT who had a negative systemic workup but a recurrence over a year after his initial presentation. (J Am Acad Dermatol 2018;4:305-9.)

Key words: B cell; lymphoma; primary cutaneous diffuse large B-cell lymphoma; leg type; regression.

INTRODUCTION

Primary cutaneous B-cell lymphoma is a lymphoma confined to the skin, with no evidence of extracutaneous disease. The 4 major subtypes of cutaneous B-cell lymphoma include primary cutaneous follicle center lymphoma (PCFCL), primary cutaneous marginal zone lymphoma (PCMZL), primary cutaneous diffuse large B-cell lymphoma, leg type (PCDLBCL-LT), and primary cutaneous diffuse large B-cell lymphoma, other (PCDLBCL-other).[1,2] PCDLBCL-LT is an aggressive subtype of primary cutaneous B-cell lymphoma that presents at a median age of 76 with a slight female predominance. Diagnosis relies on clinical and histologic features and a characteristic MUM-1 immunophenotype.

PCDLBCL-LT most commonly presents as red-to-purple nodules of the lower extremities. Extracutaneous spread typically occurs to regional lymph nodes and bone marrow with the central nervous system being the most common location of visceral involvement.[3,4]

Abbreviations used:

PCFCL:	primary cutaneous follicle center lymphoma
PCMZL:	primary cutaneous marginal zone lymphoma
PCDLBCL-LT:	primary cutaneous diffuse large B-cell lymphoma, leg type
PCDLBCL-other:	primary cutaneous diffuse large B-cell lymphoma, other
R-CHOP:	rituximab, cyclophosphamide, doxorubicin, oncovin/vincristine, and prednisone

CASE REPORT

A 79-year-old white man presented with a plaque on his left leg that was present for 1 week. Physical examination found a 20- × 12-cm irregular, rectangular-shaped, erythematous, reticulated plaque on his left anterior shin with prominent follicular accentuation (Fig 1).

Biopsy found an atypical lymphoid infiltrate in the upper dermis composed of cells with scant cytoplasm and large round-to-oval nuclei with prominent nucleoli, numerous mitotic figures, and apoptotic bodies. The atypical cells were positive with CD20, MUM-1, Bcl-2 and Bcl-6 stains. A CD3 stain showed a minor component of small, round T lymphocytes. Systemic workup was negative, and the findings were compatible with a PCDLBCL-LT.

From the Departments of Dermatology[a] and Dermotopathology,[b] St. Joseph Mercy Hospital, Ann Arbor and Base of the Bay Dermatology, Williamsburg.[c]
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Correspondence to: Paul M. Graham, DO, 5333 McAuley Dr, Suite 5003, Ypsilanti, MI 48197. E-mail: pmgraham86@gmail.com.

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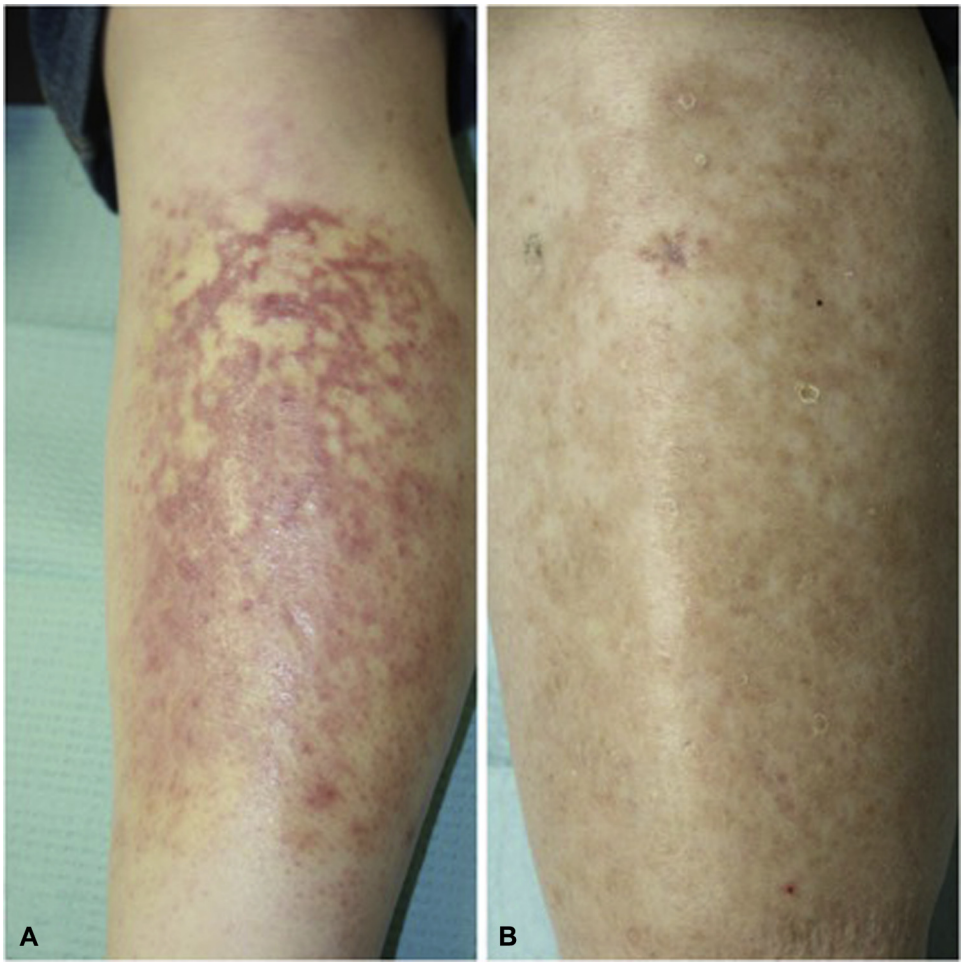

Fig 1. **A**, Five days after initial presentation: reticulated erythematous to violaceous plaque of the left anterior lower leg. **B**, Forty-five days later.

Approximately 1 month later, significant improvement of the lesion was seen in the absence of any systemic therapy. The plaque spontaneously resolved, and a punch biopsy of the area found a superficial and deep perivascular inflammatory infiltrate comprised of mature lymphocytes. The atypical, pleomorphic lymphoid infiltrate seen in the previous biopsy was not identified. Immunohistochemistry showed significant $CD3^+$ T cells with a minority of $CD20^+$ B cells. These histologic findings were consistent with spontaneous regression of the patient's PCDLBCL-LT.

A computed tomography scan of the head, chest, abdomen, and pelvis were negative for lymphoma and lymphadenopathy. A bone marrow aspirate and biopsy of the right iliac bone was negative for malignancy. The oncology department recommended 4 cycles of rituximab, cyclophosphamide, doxorubicin, oncovin/vincristine and prednisone (R-CHOP) therapy followed by local radiation. The patient declined treatment and remained disease free for more than a year but ultimately presented more than a year later with a new plaque confirmed as a recurrence of his PCDLBCL-LT.

DISCUSSION

Four subtypes of cutaneous B-cell lymphoma currently recognized include PCFCL, PCMZL, PCDLBCL-LT, and PCDLBCL-other. These subtypes are based on histologic morphology and immunohistochemistry (Table I).[1,4] PCDLBCL-LT accounts for 2.6% of primary cutaneous non-Hodgkin lymphomas with a female/male ratio of 1:6.[3,7] PCDLBCL-LT occurs on the lower extremities, trunk, head/neck, and upper extremities in 66.6%, 7.8%, 2%, and 2% of cases, respectively, with the remaining 21.6% of cases being disseminated.

Histopathologic evaluation is the primary method used in the diagnosis of PCDLBCL-LT. Biopsy results show diffuse monotonous large B cells separated from the epidermis by a Grenz zone and numerous mitotic figures (Fig 2, *A*). The atypical lymphoid cells