Cutaneous B-cell lymphoma with histologic features of mycosis fungoides

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Histologic examination of skin biopsy specimens from a 57-year-old man with a pruritic rash on his chest, abdomen, and thighs revealed a dense atypical dermal lymphoid infiltrate bordering the dermoepidermal junction and scattered intraepithelial lymphocytes. Histopathologic and clinical features were suggestive of mycosis fungoides. Immunophenotyping studies, however, identified $CD20^+$ B lymphocytes with aberrant expression of CD43. Clonal immunoglobulin gene rearrangement was demonstrated, and no clonal T-cell gene rearrangement was identified. Morphologic and immunophenotypic features were most consistent with extranodal marginal zone B-cell lymphoma. Systemic evaluation identified involvement of the bone marrow and possibly the peripheral blood, spleen, and splenic lymph nodes. Cutaneous manifestations were treated with narrowband ultraviolet B phototherapy for 3 months, resulting in complete resolution of the pruritus and rash. Primary and secondary cutaneous B-cell lymphomas seldom mimic mycosis fungoides histologically. Immunophenotyping studies were critical in correctly classifying this rare example of an epidermotropic B-cell lymphoma. (J Am Acad Dermatol 2010;62:320-3.)

Key words: lymphoid tissue; phototherapy; pruritus.

CASE REPORT

A 57-year-old man presented to the dermatology clinic in our tertiary care academic medical center with a 5-year history of an intermittently pruritic rash on the chest, abdomen, and thighs. The rash appeared to improve with topical 1% hydrocortisone, moisturizing lotions, and exposure to sunlight. The patient had no history of exposure to contactants. He denied having had any fevers, chills, night sweats, loss of appetite, weight loss, fatigue, nausea, or vomiting. His medical history was clinically significant for hyperlipidemia and gastroesophageal reflux disease treated with atorvastatin and rabeprazole. The patient had no family history of cancer or skin disorders. Physical examination revealed multiple erythematous papules on the chest (Fig 1), extending onto the abdomen and both thighs.

Three skin biopsies were obtained from the right aspect of chest, right side aspect of chest, and left thigh. Microscopic evaluation of all 3 specimens demonstrated a dense bandlike infiltrate of small monomorphic lymphocytes admixed with scattered larger cells in the papillary dermis and superficial reticular dermis (Figs 2 and 3). The inlfiltrate abutted the dermoepidermal junction and was associated with vacuolization of keratinocytes near the basal cell layer. Rare small lymphocytes extended into the mid epidermis. Immunohistochemiclal analysis of paraffin sections from two of the biopsy specimens (right side of chest and left thigh) revealed identical immunophenotypic features. The atypical dermal lymphocytes expressed positive immunohistochemical staining for CD20 and bcl-2, but were negative for CD5, bcl-6, CD10, CD23, and CD43 (Fig 4). Scattered plasma cells within the infiltrate showed a polytypic staining pattern for κ and λ immunoglobulin light chains. A moderate number of $CD3^+$ T lymphocytes were scattered throughout the infiltrate (Fig 5). Some intraepithelial lymphocytes expressed CD3, but most expressed CD20. Polymerase chain reaction analysis of frozen tissue from the right chest specimen demonstrated a clonal immunoglobulin gene rearrangement. No T-cell receptor gene rearrangement was detected by polymerase chain reaction or Southern blot analysis. The histologic, immunophenotypic, and molecular findings were diagnostic of a low-grade cutaneous B-cell lymphoma with epidermotropism.

As part of a staging evaluation, a peripheral blood sample was sent for a complete blood cell count,

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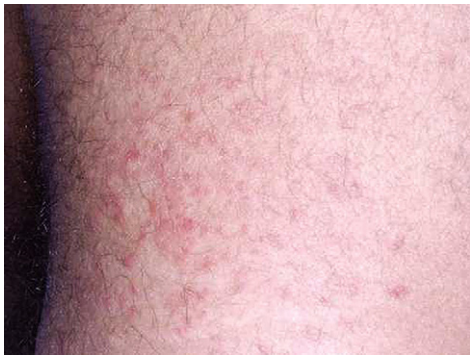

Fig 1. Multiple erythematous papules were present on the chest.

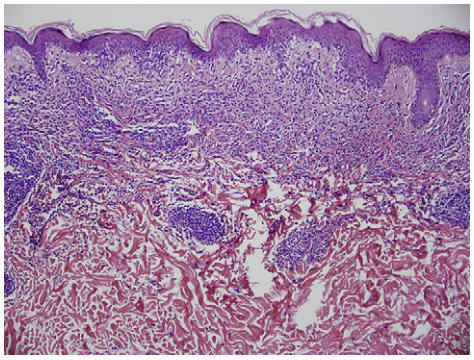

Fig 2. Skin biopsy specimen from left thigh with dense bandlike lymphocytic infiltrate in papillary dermis. (Hematoxylin-eosin stain; original magnification: ×100.)

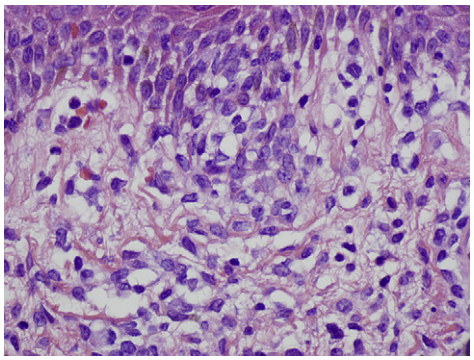

Fig 3. Atypical lymphocytes fill papillary dermis and extend into epidermis. (Hematoxylin-eosin stain; original magnification: ×600.)

peripheral smear, and flow cytometry. Findings included a white blood cell count of 5.2 K/μL (reference range [RR], 4.2-10.2) with a normal differential; hemoglobin of 13.6 (RR, 13.2-17.5); hematocrit of 38.8% (RR, 38.7-48.3); and a platelet count of 183 K/μL (RR, 151-355). Rare atypical lymphoid cells were identified on peripheral smear; however, flow cytometry did not identify a clonal B lymphocyte population. Computed tomograms of the chest,

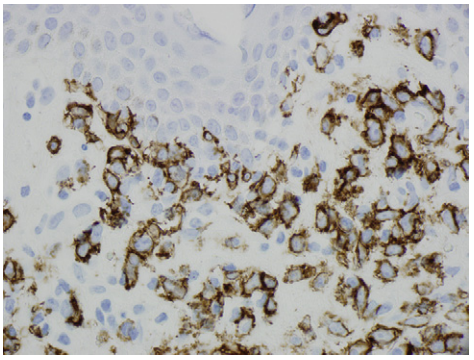

Fig 4. $CD20^+$ atypical lymphocytes with epidermotropism. (Original magnification: ×600.)

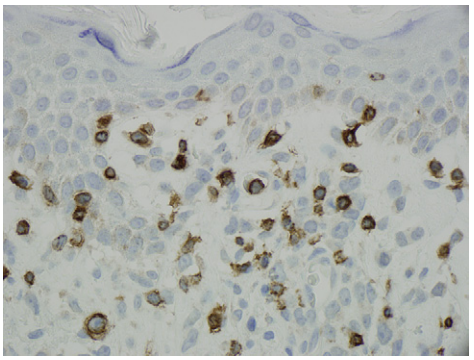

Fig 5. $CD3^+$ lymphocytes admixed with atypical B-cell infiltrate. (Original magnification: ×600.)

abdomen, and pelvis revealed an enlarged spleen and low-grade adenopathy in the splenic hilum, mesentery, and retroperitoneum.

A bone-marrow biopsy was conducted for histologic examination and immunophenotyping studies. The bone-marrow core biopsy specimen revealed a prominent sinusoidal infiltrate of atypical $CD20^+$ B cells. Flow cytometric analysis of the bone-marrow aspirate confirmed a monoclonal B-lymphocyte population expressing CD19, CD20, CD22, CD45, FMC-7, and λ immunoglobulin light chain. This population was negative for CD5, CD10, CD11c, CD23, and CD38. Histologic and immunophenotypic findings in the skin and bone marrow were diagnostic of an extranodal marginal zone B-cell lymphoma with widespread cutaneous disease.

The patient was evaluated by specialists both in dermatology and hematology and by specialists in oncology, with dermatology follow-up. Aside from pruritus, the patient was asymptomatic for lymphoma, and systemic chemotherapy was deferred in the absence of systemic symptoms. For the cutaneous disease, the patient received 21 treatment sessions over 3 months of narrowband ultraviolet (UV) B phototherapy, ranging from 175 to 785 mJ/cm^2, which produced complete resolution of