

# Primary Cutaneous CD30(Ki-1)-Positive Pleomorphic Large Cell Lymphoma in a Patient with Generalized Lichen Myxedematosus

Doo-Hyun Chi, M.D., Sung-Tae Chung, M.D., Jee-Ho Choi, M.D.,  
Kyung-Jeh Sung, M.D., Kee-Chan Moon, M.D. Jai-Kyoung Koh, M.D.

*Department of Dermatology, Asan Medical Center,  
College of Medicine, University of Ulsan, Seoul, Korea*

A case of primary cutaneous CD30(Ki-1)-positive pleomorphic large cell lymphoma in a 51-year-old woman with generalized lichen myxedematosus is presented. Histopathological findings of the biopsy specimens from the lichenoid papules of the left forearm and the dorsum of the right hand were compatible with papular mucinosis. The mucinous material in the papillary dermis stained with alcian blue at pH 2.5 and was susceptible to hyaluronidase, but serum protein electrophoresis showed no abnormal bands and an abnormal paraprotein was not detected in our patient. Histopathological examination of a solitary, firm, purplish nodule on the right popliteal area showed diffuse and extensive infiltration in the reticular dermis composed of large, atypical, often hyperchromatic, sometimes multinucleated and markedly pleomorphic cells. The majority of the large atypical cells were CD30(Ki-1) positive. The TCR gene rearrangement analysis demonstrated the presence of a monoclonal rearrangement of the  $\gamma$ -TCR gene in the skin biopsy specimen of our patient. The organ-system survey revealed no evidence of internal organ involvement. We concluded that this was primary cutaneous CD30(Ki-1)-positive pleomorphic large cell lymphoma developing in a patient with generalized lichen myxedematosus. (*Ann Dermatol* 9:(1)55~58, 1997).

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*Key Words* : CD30(Ki-1)-positive large cell lymphoma, Lichen myxedematosus

Generalized lichen myxedematosus is a rare fibromucinous disorder of unknown etiology. It is characterized by the presence of infiltrative skin lesions with the mucinous deposition in the papillary dermis and the presence of a monoclonal protein<sup>1,3</sup>. In addition, extracutaneous manifestations, most often gastrointestinal<sup>1</sup>, were present and malignant disorders such as non-Hodgkin's lymphoma<sup>4</sup>, multiple myeloma<sup>5,6</sup> and Hodgkin's disease<sup>7</sup> have been described in scleromyxedematous patients.

We report a patient with generalized lichen myxedematosus associated with primary cutaneous CD30(Ki-1)-positive pleomorphic large cell lymphoma.

## REPORT OF A CASE

A 51-year-old woman had a 7-year history of a pruritic papular eruption on the extensor surfaces of the extremities that had become more generalized and sclerotic during the past 2 months. Examination revealed firm, densely grouped, red to flesh-colored, lichenoid papules especially on the dorsal regions of the hands and the extensor areas of the arms and legs (Fig. 1). Histopathological findings of two biopsy specimens from the left forearm and the dorsum of the right hand were compatible

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**Reprint request to** : Doo-Hyun Chi, M.D., Department of Dermatology Asan Medical Center College of Medicine, University of Ulsan Kangdong P.O. Box 145 Seoul, Korea Phone: (02)224-3460; fax (02)486-7831

**Fig. 1.** Lichenoid papules on the extensor surface of the left forearm.

**Fig. 2.** The mucinous material in the papillary dermis stained with alcian blue at pH 2.5 ( $\times 100$ ).

with papular mucinosis. The mucinous material in the papillary dermis stained with alcian blue at pH 2.5 (Fig. 2) and was susceptible to hyaluronidase digestion. The physical examination was normal except for the skin lesions. The family history, the past medical history and the review of systems were all insignificant. The results of the following studies were within normal limits or negative: a blood cell count, urinalysis, liver function test, immunoglobulins, fasting serum glucose and lipid profile. Serum protein electrophoresis showed no abnormal bands and an abnormal paraprotein was not detected in our patient. The clinical and

**Fig. 3.** A solitary, firm, nontender, purplish nodule on the right popliteal area.

**Fig. 4.** A biopsy specimen showing large pleomorphic lymphoid cells (haematoxylin and eosin,  $\times 400$ ).

histopathological findings were consistent with a diagnosis of generalized lichen myxedematosus and treatment was started with topical steroids and oral prednisone. 3 months later, there was some improvement, but 5 months later, a solitary, firm, purplish nodule was detected on the right popliteal area (Fig. 3). Histopathological examination of the nodule showed a diffuse and extensive infiltrate in the reticular dermis and subcutis with a Grenz zone in the upper dermis. The infiltrate composed of large, atypical, often hyperchromatic, sometimes multinucleated and markedly pleomorphic cells, scattered small lymphocytes, some

**Fig. 5.** The majority of the large atypical cells were CD30(Ki-1) positive ( $\times 400$ ).

eosinophils and rare plasma cells (Fig. 4). Mitotic figures, vascular proliferation, and necrosis were also observed, but ulceration or epidermotropism was absent. The majority of the large atypical cells were CD45(LCA), CD45RO(UCHL-1), and CD30(Ki-1) positive (Fig. 5), but negative for CD20(L-26). The TCR gene rearrangement analysis demonstrated the presence of a monoclonal rearrangement of the  $\gamma$ -TCR gene in the skin biopsy specimen of our patient (Fig. 6). The organ-system survey revealed no evidence of internal organ involvement. We concluded that this was primary cutaneous CD30(Ki-1)-positive pleomorphic large cell lymphoma developing in a patient with generalized lichen myxedematosus.

## DISCUSSION

Generalized lichen myxedematosus is a rare fibromucinous disorder characterized by the presence of infiltrative skin lesions with the deposition of mucinous material in the papillary dermis<sup>1,2</sup>. The disease primarily involves skin as generalized papular eruption with sclerosis, although systemic manifestations such as gastrointestinal or pulmonary involvement<sup>1,2</sup>, proximal myopathy<sup>8</sup>, inflammatory polyarthritis, endocrine abnormalities, neurologic dysfunction have been described<sup>1,9,10</sup>. A monoclonal paraprotein has been also detected in most patients with this disorder, most commonly of the Ig G- $\lambda$  type<sup>1,3</sup>. In addition, hematologic disorders, including non-Hodgkin's lymphoma<sup>4</sup>, multiple myeloma<sup>5,6</sup> and Hodgkin's disease<sup>7</sup>, have been reported in a few patients. Primary cutaneous CD30(Ki-1)-positive pleomorphic large cell lymphomas

**Fig. 6.** The TCR gene rearrangement analysis demonstrated the presence of a monoclonal rearrangement of the  $\gamma$ -TCR gene in the skin biopsy specimen.

have been recognized only recently as a distinct type of non-Hodgkin's lymphoma with a favorable prognosis<sup>11,12</sup>. In contrast to morphologically identical primary nodal counterparts, primary cutaneous CD30(Ki-1)-positive large cell lymphomas are uncommon in children and adolescents, and have a much better prognosis<sup>11</sup>. In contrast, primary cutaneous pleomorphic, anaplastic or immunoblastic lymphomas, which are completely negative for CD30, generally have a poor prognosis<sup>11,12</sup>. Both entities, lymphomatoid papulosis(Lyp) and cutaneous CD30(Ki-1)-positive large cell lymphoma share some features clinically and histologically. Clinical similarities include a benign, protracted course of disease and the possibility of progression to lymphoma<sup>13</sup>. However, our case differed from Lyp in several aspects. The clinical presentation was clearly different: multiple disseminated papules in Lyp, but a single nodule in our case. Histologically, both entities have in common large, atypical cells expressing CD30(Ki-1) antigen<sup>13</sup>. However, the architectural pattern of involvement, distribution of atypical cells and composition of the cellular infiltrate are quite different between Lyp and our case. In our patient, lichen myxedematosus was associated with primary cutaneous CD30(Ki-1)-positive pleomorphic large cell lymphoma, composed of T cell immunophenotypes and an abnormal paraprotein was not found. The significance of this coexistence is not clear at this time, and one may think that this association is coincidental, but it is speculated that as yet unidentified factors from malignant cells may stimulate fibroblasts to increase activity, thus aggravating the pre-existing papular

mucinosi to generalized lichen myxedematosus. Alternatively, an unidentified serum factor of the patient with scleromyxedema may play some role in the development of the malignant tumors<sup>14</sup>. Further studies will help us characterize in more detail the exact mechanism of this association.

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