A Case of Idiopathic Lymphocytoma Cutis

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The pseudolymphoma of the skin has the architectural and cytological features of a neoplastic proliferation of lymphoid tissue but pursue a benign course. Cutaneous B cell pseudolymphoma (CBPL) shares many histopathologic and clinical features with cutaneous B cell lymphoma (CBCL). Therefore, the differentiation between CBPL and CBCL is often very difficult, but it is important because each of them has a different therapeutic consequence. Recently, immunoglobulin gene rearrangement is considered as a reliable technique for differentiation of CBPL with CBCL.

We herein report a case of idiopathic lymphocytoma cutis, showing a typical nodular infiltrate of lymphocytes that formed a follicular germinal center resembling reactive lymph nodes with numerous tingible bodies, and that revealed a polyclonality in the immunoglobulin gene rearrangement. (Ann Dermatol 16(2) 71~75, 2004)

Key Words: Cutaneous B cell pseudolymphoma, Idiopathic lymphocytoma cutis, Immunoglobulin gene rearrangement

INTRODUCTION

Cutaneous pseudolymphoma neither refers to a specific disease nor implies anything about the cause but simply implies a process of accumulation of lymphocytes in the skin in response to a variety of stimuli[1]. Depending on the predominant cell type in the infiltrate, cutaneous pseudolymphoma is divided into T cell and B cell pseudolymphoma. Cutaneous B cell pseudolymphomas (CBPL) includes idiopathic lymphocytoma cutis, borrelial lymphocytoma cutis, tattoo-induced lymphocytoma cutis, post-zoster scar lymphocytoma cutis, and some persistent nodular arthropod-bite reactions[1].

Herein, we present a case of CBPL, which had no known cause, idiopathic lymphocytoma cutis, and showed a polyclonality in B cell receptor gene rearrangement.

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CASE REPORT

A 62-year-old woman presented with a 2-month history of several, asymptomatic, erythematous to violaceous papules and plaques involving the forehead and left cheek without pruritus or tenderness (Fig. 1). On physical examination, there was no cervical lymphadenopathy and she did not complain of fever, night sweat or weight loss. In her past history she had developed similar lesions on the forehead and nose 6 years ago, which resolved spontaneously without treatment. There was no trauma, insect bite or specific medication history.

Histological examination of a biopsy specimen taken from the plaque revealed a nodular infiltrate of lymphocytes mimicking a lymphoid follicle with a germinal center in the dermis and subcutis (Fig. 2A). Most cells were small lymphocytes admixed with a small number of histiocytes and eosinophils. Abnormal mitosis was absent. Some lymphoid follicles had numerous tingible bodies, macrophages containing phagocytosed lymphocytes (Fig. 2B). The

epidermis was uninvolved and epidermal appendage was preserved.

Immunohistochemistry showed the infiltrated cells strongly expressed CD20 antigen (Fig. 3). Other immunohistochemical stains including CD3, CD4 and CD56 antigen were all negative for tumor cells. Thus, there was a predominance of B lymphocytes in the infiltrate. B cell receptor gene rearrangement test demonstrated the polyclonality of tumor cells.

Based on these clinical and histologic findings, this case was diagnosed as idiopathic lymphocytoma cutis. She applied topical steroid ointments for 3 months but the lesion has shown no improvement yet.

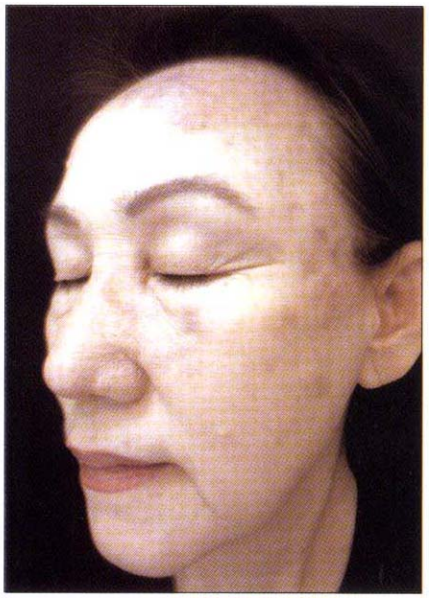


Fig. 1. Several, erythematous to violaceous papules and plaques on the forehead and left cheek.

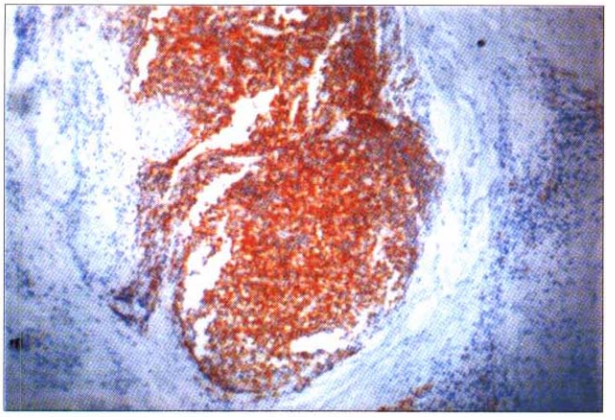
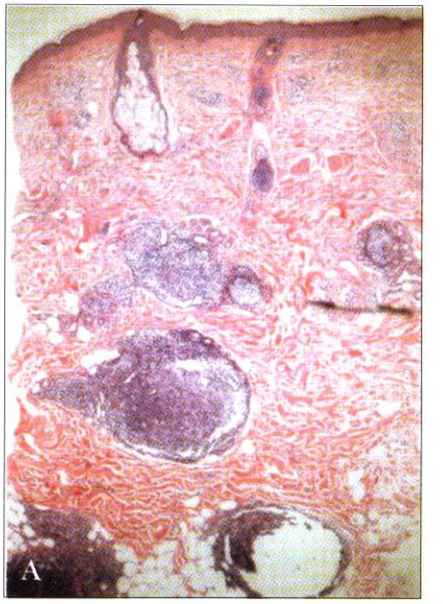


Fig. 3. The infiltrated tumor cells strongly expressed CD20 antigen.



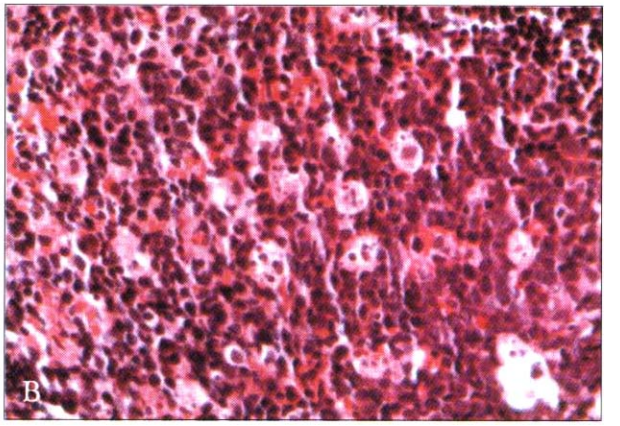


Fig. 2. A. Nodular infiltrate of lymphocytes mimicking a lymphoid follicle with germinal center in the dermis and subcutis (H & E, ×10). B. The follicles composed of small lymphocytes and stippled by multiple tingible bodies (H & E, ×400).

DISCUSSION

Cutaneous pseudolymphoma is a benign cutaneous lymphoid infiltrate that simulates lymphoma clinically and histologically. Pseudolymphoma can be classified according to the predominant lymphocytic cell of the infiltrate (Table 1)[1,2]: cutaneous T cell pseudolymphoma (CTPL) and cutaneous B cell

Table 1. Classification of Cutaneous Pseudolymphoma

Cutaneous T cell pseudolymphomas (CTPL)
Bandlike CTPL (major pattern)
Idiopathic CTPL
Lymphomatoid drug eruption
Lymphomatoid contact dermatitis
Actinic reticuloid
Clonal CTPL
Nodular CTPL (minor pattern)
Anticonvulsant-induced pseudolymphoma syndrome
Persistent nodular arthropod-bite reactins
Nodular scabies
Acral pseudolymphomatous angiokeratoma
Cutaneous B cell pseudolymphomas (CBPL) ; nodular pattern
Idiopathic lymphocytoma cutis
Borrelial lymphocytoma cutis
Tattoo-induced lymphocytoma cutis
Post-herpes zoster scar lymphocytoma cutis
Lymphocytoma cutis caused by antigen injection/acupuncture
Persistent nodular arthropod-bite reaction
Lymphomatoid drug eruption
Acral pseudolymphomatous angiokeratoma
Clonal CBPL

Cited from reference 1

pudolymphoma (CBPL). The histologic appearance of the infiltrate in CTPL can be either band-like or nodular, whereas the predominant appearance of the infiltrate in CBPL is nodular[1,3].

In most cases, the cause of CBPL is unknown: idiopathic lymphocytoma cutis. It is the most common example of CBPL. The most favored sites of involvement include the face, chest and upper extremities[1,3]. The female-to-male ratio is 3 : 1. Clinically, the patients with CBPL typically exhibit one or a few erythematous to violaceous nodules or plaques similar to cutaneous B cell lymphoma (CBCL)[1-4]. In our case, there were several erythematous to violaceous papules and plaques on the face typical for cutaneous pseudolymphoma, and she did not have any history of the associated causes or underlying diseases.

Histopathologic examination of CBPL lesion reveals a dense nodular infiltrate of lymphocytes admixed with a variable number of histiocytes, eosinophils and plasma cells. The infiltrate tends to favor the papillary dermis and often to diminish in the deeper dermis (top-heavy)[1-5]. But our case did not show the typical top-heavy pattern of infiltration. In the follicular pattern of CBPL, well-defined reactive lymphoid follicles, primary or secondary, are characteristic. These primary follicles are composed of homogeneous small lymphocytes, whereas the secondary follicles are compartmetalized into germinal centers surrounded by a mantle zone. Germinal centers are composed of follicular center cells including small cleaved and large lymphocytes, follicular dendritic cells, and tingible body macrophages, whose cytoplasm contains the debris of small

Table 2. Summary of Cutaneous Pseudolymphomas in Korean Literatures

Case	Patient	Clinical finding	Type	Cause	Histologic finding	Ig gene rearrangement	Reference
1	30/M	Solitary nodule in upper arm	B cell	Arthropod bite reaction	Nodular infiltrates	Polyclonality	4
2	63/F	Solitary nodule on the abdomen	B cell	Repeated trauma following scratching	Diffuse infiltrates	Polyclonality	
3	65/M	Several papules on the ear and neck	B cell	Hair dye	Nodular infiltrates	ND	7
4	58/M	Eczematoid lesion on the face and neck	T cell	Photosensitivity (Actinic reticuloid)	Perivascular, periadnexal infiltrates	ND	8
5	52/M	Multiple nodules on the face and abdomen	T cell	Phenytoin	Nodular infiltrates	ND	9
6	47/M	Generalized papulovesicular eruption	T cell	Carbamazepine	Band-like infiltrates	ND	10
7	65/M	Several papules on the face and hand	T cell	Photosensitivity (Actinic reticuloid)	Perivascular, periadnexal infiltrates	ND	11
8	27/M	Facial edema, generalized erythema	T cell	Carbamazepine	Band-like infiltrates	ND	12
Our case	62/F	Several papules and plaques on the face	B cell	Idiopathic	Nodular infiltrates	Polyclonality	

ND: not done

lymphocytes that have undergone apoptosis[5]. The germinal center and tingible body macrophages are typically seen in follicular CBPL[5].

Immunohistochemical findings reveal a B-cell predominance with variable numbers of T cells. The most important feature is the presence of B lymphocytes with polyclonal light chains (a mixture of λ and κ light chains in the B cells)[1]. In our case, most tumor cells expressed the CD20 antigen (pan B-cell marker), but were negetive for CD3, CD4 (T cell marker) and CD56 antigen (NK cell marker).

In the differential diagnosis of CBPL, CBCL is the most difficult but important. Especially, differentiating CBPL from marginal zone lymphoma can be difficult. Clinically, both of them affect women more often than men, and they appear as single or multiple cutaneous nodules on the face, arms or trunk[6]. But marginal zone cells, small to medium-sized cells with abundant pale cytoplasm and irregular nuclear contours and expressing pan B cell marker, were the significant features favoring a diagnosis of marginal zone lymphoma. Also, confluent sheets or zone of plasma cells, as opposed to solitary plasma cell in CBPL, were seen in marginal zone lymphoma[6]. Recently, molecular analysis of lymphoid gene rearrangement by Southern blot or polymerase chain reaction technique has proved to be a useful technique for differentiating them[1,4,7]. There are eight cases of cutaneous pseudolymphoma in Korean literature (Table 2).

Clinically, in this case, localized papules and plaques were presented and on the past history, the similar lesions had also developed 6 years ago, which showed spontaneous resolution. These findings suggested that the disease had benign characteristics. Histologic findings showed the nodular infiltrate of lymphocytes mimicking a lymphoid follicle with

germinal center in the dermis and subcutis. Most of the tumor cells were B lymphocytes which expressed CD20 antigen. Numerous tingible bodies were scattered in the germinal center. The polymerase chain reaction of the immunoglobulin gene rearrangement showed a polyclonal pattern, so this case was diagnosed for CBPL.

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