

Lymphomatoid papulosis : A Case Report.

Watchai Mingbunjersuk MD,

Nopadon Noppakun MD.

ABSTRACT:

MINGBUNJERSUK W, NOPPAKUN N. LYMPHOMATOID PAPULOSIS: A CASE REPORT. THAI J DERMATOL 2011; 27: 135-141.

DIVISION OF DERMATOLOGY, DEPARTMENT OF MEDICINE, FACULTY OF MEDICINE, CHULALONGKORN UNIVERSITY, BANGKOK, THAILAND.

Lymphomatoid papulosis is one of CD30 positive cutaneous lymphoproliferative diseases. The clinical presentation is recurrent reddish-brownish papulonodular lesions that tends to spontaneously regress leaving hyperpigmented scars. Lesions usually occur over the trunk and the extremities. There is an association between lymphomatoid papulosis and malignancies.

We reported a 39-year-old man presenting with multiple papulonodular lesions on his trunk and extremities. The histopathology revealed dense dermal infiltration with atypical monomorphous mononuclear cells that have pleomorphic large vesicular nuclei and some cells have hyperchromatic nuclei and atypical mitotic figure. The atypical cells expressed CD4 and CD30, suggesting the diagnosis of lymphomatoid papulosis. The lesions were spontaneously healed within 2 weeks.

Key words: Lymphomatoid papulosis, CD30 positive cutaneous lymphoproliferative disease

บทคัดย่อ :

วัฒน์ชัย มิ่งบรรเจิดสุข, นกตล นพคุณ รายงานผู้ป่วยโรคลิ้มโฟมาตอยด์ แปนปูโลสิส วารสารโรคผิวหนัง 2554; 27: 135-141.

หน่วยผิวหนัง ภาควิชาอายุรศาสตร์ คณะแพทยศาสตร์ จุฬาลงกรณ์มหาวิทยาลัย

โรคลิ้มโฟมาตอยด์ แปนปูโลสิสเป็นโรคในกลุ่มโรคผิวหนังเนื้องอก ลิ้มโฟโพลิเฟอเลติฟ ซีดัสลามิบัให้ผลบวก ซึ่งประกอบด้วย โรคลิ้มโฟมาตอยด์ แปนปูโลสิส โรคมะเร็งต่อมน้ำเหลืองที่แสดงออกทางผิวหนังชนิดเซลล์ใหญ่ผิดปกติและโรคบอเคอร์โรค ซีดัสลามิบัให้ผลบวก โดยมีลักษณะทางคลินิกคือ มีตุ่มหรือก้อนสีแดงซึ่งสามารถหายได้เองโดยมักทิ้งแผลเป็นรอยดำไว้ เป็นๆหายๆมักพบได้บริเวณลำตัวและแขนขาทั้งสองข้าง อย่างไรก็ตามพบว่าโรคลิ้มโฟมาตอยด์ แปนปูโลสิสสามารถพบได้ร่วมกับโรคมะเร็งบางชนิดได้

คำสำคัญ: โรคลิ้มโฟมาตอยด์ แปนปูโลสิส โรคผิวหนังเนื้องอก ลิ้มโฟโพลิเฟอเลติฟ ซีดัสลามิบัให้ผลบวก

Lymphomatoid papulosis is one of CD30 positive cutaneous lymphoproliferative diseases, including lymphomatoid papulosis, primary cutaneous anaplastic large cell lymphoma, and borderline CD30⁺ lesions. The clinical presentation is recurrent reddish-brownish papulonodular lesions that tends to spontaneously regress leaving hyperpigmented scars. Lesions usually occur over the trunk and the extremities. There is an association between lymphomatoid papulosis and malignancies.

Case Report

A 39-year-old man, without medical history, was referred to our unit for treatment of papulonodular lesions on the trunk and the extremities. The lesions were slightly tender and pruritic, and had healed spontaneously without scarring. He had a 3 weeks

history of self-healing reddish papules on his trunk and extremities.

Clinical examination revealed multiple groups of 2-4mm. erythematous papules on his the trunk, both arms, both hands and legs. No mucosal involvement was found. The patient was in good health, and the general examination was completely normal.

Histopathological examination of a skin biopsy taken from a left hand lesion revealed dense atypical monomorphous mononuclear infiltrate in the dermal papillae and extends in to the deep dermis. The cells have pleomorphic large vesicular nuclei and some cells have hyperchromatic nuclei and atypical mitotic figure. There is no evidence of epidermotrophism.

Immunohistochemistry revealed the atypical cells were positive for CD4, CD30 and TIA1. They were negative for CD3, CD5, CD8, CD20, CD34, S-100

protein, PAX5, HMB45, GranzymeB and ALK. Minority of cells were positive for EMA. A complete examination revealed no evidence of systemic disease. He was diagnosed as Lymphomatoid papulosis type C. Two weeks later, the main lesions resolved spontaneously without scarring. No treatment was given. Follow-up for 4 months did not show any recurrence or associated disease.

Discussion

Lymphomatoid papulosis was originally described by Macaulay in 1968 as a self-healing rhythmical paradoxical papular eruption with histologically malignant but clinically benign.¹ Lymphomatoid papulosis most commonly occurs between the age of 20 and 40 year old. The disorder was characterized by recurrent crops of papulonecrotic or papulonodular lesions predominantly affecting the trunk and extremities. They can be resolved spontaneously after 2-12 weeks, usually leaving slightly depressed oval scars.²

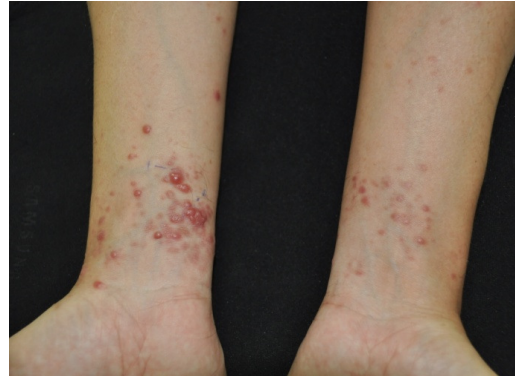
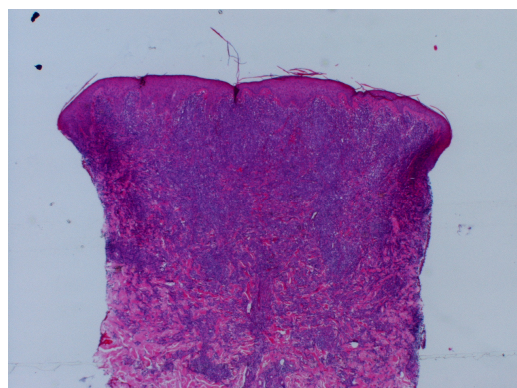


Figure 1

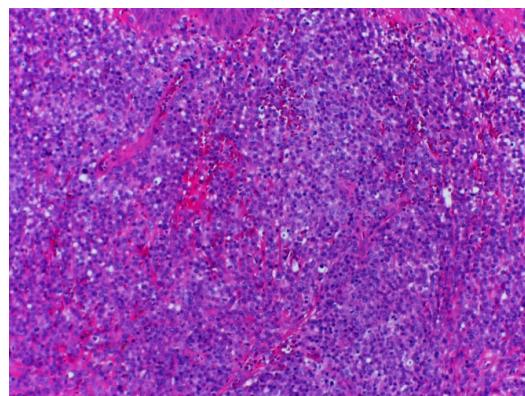
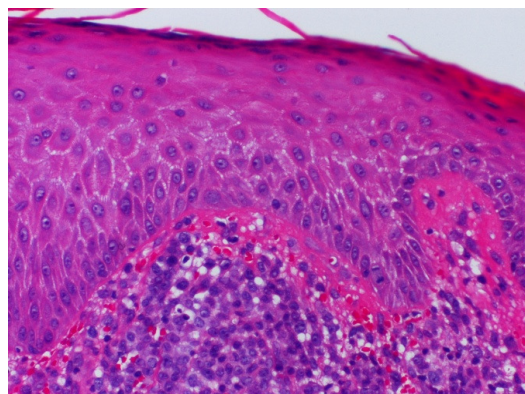


Figure 2

Figures 1,2 Multiple discrete erythematous papules on both forearms and dorsum of hand.

**Figure 3****Figure 5****Figure 4**

Figures 3,4 Multiple discrete erythematous papules and some coalescent to plaque on both forearms and leg.

**Figure 6****Figure 7**

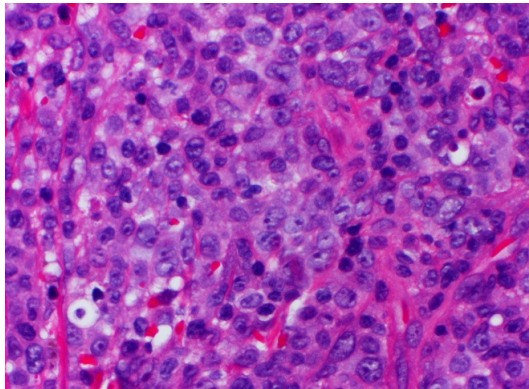


Figure 8

Figure 5-8 Sheet of dense atypical monomorphous mononuclear cells infiltrate in the dermal papillae, upper and deep dermis.(haematoxylin-eosin stain. Original magnification x40,

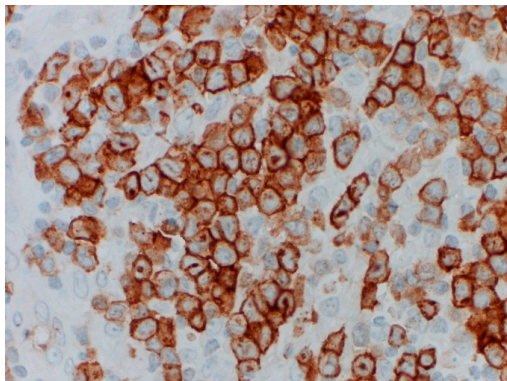


Figure 9 Staining for CD30 shows positive atypical cells in sheets.

Histologically, lymphomatoid papulosis has been divided into type A, type B and type C³ depending on the predominant cell type presents in the infiltrate. Type A is characterized by large CD30⁺ atypical cells intermingled with a prominent inflammatory infiltrate. The large tumor cells have polymorphic convoluted

nuclei with a minimum of 1 prominent nucleolus and resemble Reed-Sternberg cells when binucleate, as is seen in Hodgkin's disease. Type B is characterized by smaller atypical cells with hyperchromatic cerebriform nuclei resembling the atypical lymphocytes in mycosis fungoides. Epidermotropism is more common in this variant. Type C (diffuse large cell type) is characterized by sheets of CD30⁺ anaplastic large cells indistinguishable from Anaplastic Large Cell Lymphoma (ALCL). However, some authorities view this histologic variant as borderline ALCL.^{4,5,6}

There is an association between lymphomatoid papulosis and malignancies such as Hodgkin's disease, mycosis fungoides and cutaneous CD30+ large cell lymphomas. Basarab et al, reported lymphomatoid papulosis can occur precede, concurrent and after onset of mycosis fungoides. There are 10-20% of lymphomatoid papulosis patients who developed malignant lymphoma. However, the precise risk of development is unclear. The patients should be informed about the possible changes that might indicate malignancy such as persistence of lesions, increased size of lesions, B symptoms and lymph node enlargement.^{7,8,9}

There are various treatments to suppress the disease including systemic corticosteroids, low dose weekly metotrexate, UV phototherapy and topical chemotherapy. There are the possibility of progression to Hodgkin's disease, anaplastic large cell lymphoma or mycosis fungoides, The cumulative risk of

transformation into lymphoma may be higher than 20%, therefore long-term surveillance is essential in all case of lymphomatoid papulosis. The patients need to be examined regularly.^{5,10,11,12}

Clinical and histologic differences between cutaneous CD30+ Large cell lymphoma, borderline cases and lymphomatoid papulosis are summarized in table1.¹¹

Table1: Clinical and histological finding in CD30+large cell lymphoma and lymphomatoid papulosis

Features	CD30+ LCL*	Borderline	LYP**
Clinical			
Extent	Solitary>regional	Regional	Regional/diffuse
Lesion	Nodule, tumor	Nodule	Pupule, papulonodule
Regression	Infrequent	Frequent	Always
Extracutaneous DS.	25-30%	Absent	Absent
Histologic			
Wedge shape infiltrate	Absent	Occasional	Regularly
Large sheet CD30+ cells	Always	Small cluster	Absent
Infiltration of subcutis	Present	Absent	Absent
CD30 positivity	>75% tumor cells	Small cluster	Scatter CD30+ cells

* large cell lymphoma

** lymphomatoid papulosis

References

1. Macaulay WL. Lymphomatoid papulosis: A continuing self-healing eruption, clinically benign-histologically malignant. Arch Dermatol 1968;97:23-30.
2. el-Azhary RA, Gibson LE, Kurtin PJ, Pittelkow MR, Muller SA. Lymphomatoid papulosis: A clinical and histopathologic review of 53 cases with leukocyte immunophenotyping, DNA flow cytometry, and T-cell receptor gene rearrangement studies. J Am Acad Dermatol. 1994;30(2 Pt 1):210-8.
3. El Shabrawi-Caelen L, Kerl H, Cerroni L. Lymphomatoid Papulosis: reappraisal of Clinicopathologic Presentation and Classification Into Subtypes A, B, and C. Arch Dermatol 2004;140:441-7.
4. Saggini A, Gulia A, Argenyi Z, et al. A Variant of Lymphomatoid Papulosis Simulating Primary Cutaneous Aggressive Epidermotropic CD8+ Cytotoxic T-cell Lymphoma. Description of 9 Cases. Am J Surg Pathol. 2010;34(8):1168-75.
5. De Souza A, Camilleri MJ, Wada DA, et al. Clinical, histopathologic, and immunophenotypic features of lymphomatoid papulosis with CD8 predominance in 14 pediatric patients. J Am Acad Dermatol. 2009;61(6):993-1000.
6. Stein H, Foss HD, Dürkop H, et al. CD30⁺ anaplastic large cell lymphoma: a review of its histopathologic, genetic, and clinical features. Blood. 2000;96(12):3681-95.
7. Tan AW, Giam YC. Lymphomatoid papulosis associated with recurrent cutaneous T-cell lymphoma. Ann Acad Med Singapore. 2004;33(1):110-2.
8. Cabanillas F, Armitage J, Pugh WC, et al. Lymphomatoid papulosis: a T-cell dyscrasia with a propensity to transform into malignant lymphoma. Ann Intern Med 1995;122: 210-7.

9. Kadin ME, Levi E, Kempf W. Progression of lymphomatoid papulosis to systemic lymphoma associated with escape from growth inhibition by transforming growth factor-beta and CD30 ligand. *Ann NY Acad Sci* 2001; 941: 59-68.
10. Vonderheid EC, Sajjadian A, Kadin ME. Methotrexate is effective therapy for lymphomatoid papulosis and othe primary cutaneous CD30-positive lymphoproliferative disorders. *J Am Acad Dermatol* 1996;34:470-81.
11. Marie-France D, Lynne JG, Marshall EK, Howard K. K: Is it lymphoma or lymphomatoid papulosis?. *J Am Acad Dermatol* 1997;36:765-772
12. Willemze R, Beljaards RC. Spectrum of primary cutaneous CD30 (Ki-1)-positive lymphoproliferative disorders: A proposal for classification and guidelines for management and treatment. *J Am Acad Dermatol* 1993;28:973-980.