A Solitary Facial Nodule of a 48-year-old Man

Yu-Tzu Tai[1] Horng-Shin Lin[1] Yu-Ping Hsiao[1] Han Chang[2] Jen-Hung Yang[1]

CASE REPORT

A 48-year-old man presented with a reddish skin tumor over face for three months. He denied any systemic disease and history of insect bite. The skin examination revealed a reddish dome-shaped elastic firm nodule, 1.2 cm in diameter, over left zygomatic area (Fig. 1). Other physical exam did not show any abnormal findings such as lymphadenopathy or hepatosplenomegaly. The nodule was partially excised for pathological examination. The histopathology revealed superficial and deep patches of atypical lymphoid cell infiltrates around the appendages with a characteristic lymphoepithelial pattern and Grenz zone. The infiltrate is composed of small lymphocytes, centrocyte-like cells, and plasma cells admixed with a small proportion of large blastoid cells (Fig. 2). Immunohistochemical stains showed that the infiltrate was strongly positive for CD20 and Bcl-2 (Fig. 3). A complete blood count, liver and renal function, lactate dehydrogenase, and serum immunoglobulin analysis were normal, and hepatitis B/C serology and HIV tests were negative. A positron emission tomography (PET) scan did not show any evidence of systemic involvement. After total excision of the lesion, he was regularly followed up without local recurrence at our clinic for more than half a year.

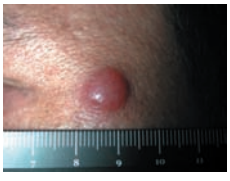

Fig. 1

A reddish, dome-shaped and elastic firm nodule about 1.2 cm in diameter over left zygomatic area of face.

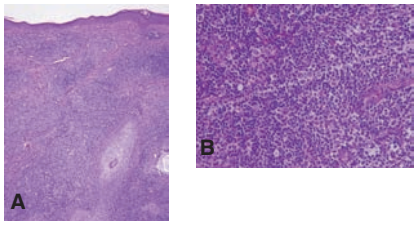

Fig. 2

(A) Lobular and diffuse lymphoid infiltrates throughout the dermis (bottom-heavy) and sparing of the epidermis. (H&E, original magnification x100)

(B) The infiltrate is composed of centrocyte-like cells, lymphoplasmacytoid cells and immunoblast-like cells. (H&E, original magnification x400)

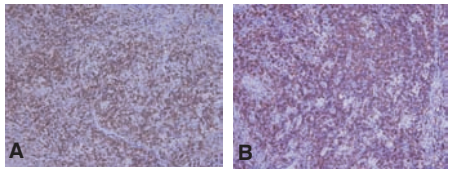

Fig. 3

Immunohistochemical features:

(A) Most of the neoplastic cells express CD20 and (B) react positively for Bcl-2. (original magnification x200).

From the Department of Dermatology[1] and Pathology,[2] Chung Shan Medical University Hospital, Taichung City, Taiwan, R.O.C.

Corresponding author: *Han Chang, No. 110, Sec. 1, Jianguo N. Rd., Taichung City 402, Taiwan, R.O.C.*
TEL: 886-4-24730022 FAX: 886-4-24753984 E-mail: changhan@csmu.edu.tw
Funding source: none
Conflict of interest: none declared

Received: August 04, 2008
Revised: October 02, 2008
Accepted: October 22, 2008

DIAGNOSIS: Primary Cutaneous Marginal Zone B Cell Lymphoma

DISCUSSION

Primary cutaneous marginal zone B cell lymphoma (PCMZL) is a low-grade malignant B-cell lymphoma without extracutaneous involvement. This type of lymphoma has been reported to represent 2% to 16% of all cutaneous lymphomas.[1] Patients with PCMZL are mainly adults older than 40 years with a male predominance. Lesions often involve the trunk and arms, consisting of deep red to violaceous infiltrated plaques, nodules or tumors, and are multifocal in approximately 75 percent of cases. A small number of patients presented with lesions on the head and neck area. When affecting this area, lesions tended to be hard and deeply infiltrated.[2] Our case illustrated an unusual clinical presentation.

Histopathlogically, PCMZL typically is "bottom-heavy" and characterized by a dense, lymphocytic infiltrate distributed mainly in the reticular dermis and often extending into subcutaneous tissue. There is no epidermotropism, and a Grenz zone can be observed in most cases. Adnexal involvement was usually present, with eventual formation of lymphoepithelial complexes. Cytologically, PCMZL is characterized by a polymorphous infiltrate that includes centrocyte-like cells, monocytoid B cells and lymphoplasmacytoid cells with a variable proportion of large (blastoid) cells. The centrocyte-like cells are small cells with irregular nuclei but usually have more cytoplasm than centrocytes. Monocytoid B cells are small to medium-sized cells with round or indented nuclei and abundant pale cytoplasm, and the large cells resemble centroblasts or immunoblasts.[3] Reactive follicle centers are present in approximately 70% of cases.[4] Our case shows a typical histological presentation of PCMZL. Besides, the small neoplastic B cells react positively for Bcl-2, which facilitates differentiation from pseudolymphoma.

Regarding the treatment of PCMZL, patients presented with solitary or localized skin lesions were treated with either surgical excision or local radiotherapy, which will often result in sustained complete remissions. Cutaneous relapses after remission had been reported in nearly one-half of patients, but dissemination to extracutaneous sites is exceedingly rare.[4] The skin relapses are much more common in patients with multifocal skin lesions than in patients with solitary or localized skin lesions. Head and neck presentation and large cell transformation may be associated with a worse prognosis.[2] However, it has been accepted the notion that the indolent clinical behavior and excellent prognosis with a 5-year-survival rate close to 100% in the cases of PCMZL.

When a solitary reddish infiltrated nodule was noted over the face, PCMZL should be put into differential diagnosis lists with systemic survey and long-term follow up.

REFERENCES

1. Fink-Puches R, Zenahlik P, Back B, *et al.*: Primary cutaneous lymphomas: applicability of current classification schemes (European Organization for Research and Treatment of Cancer, World Health Organization) based on clinicopathologic features observed in a large group of patients. Blood 99: 800-805, 2002.
2. Servitje O, Gallardo F, Estrach T, *et al.*: Primary cutaneous marginal zone B-cell lymphoma: a clinical, histopathological, immunophenotypic and molecular genetic study of 22 cases. Br J Dermatol 147: 1147-1158, 2002.
3. Cho-Vega JH, Vega F, Rassidakis G, *et al.*: Primary cutaneous marginal zone B-cell lymphoma. Am J Clin Pathol 125: S38-S49, 2006.
4. Hoefnagel JJ, Vermeer MH, Jansen PM, *et al.*: Primary cutaneous marginal zone B-cell lymphoma: clinical and therapeutic features in 50 cases. Arch Dermatol 141: 1139-1145, 2005.
5. Li C, Inagaki H, Kuo TT, *et al.*: Primary cutaneous marginal zone B-cell lymphoma: a molecular and clinicopathologic study of 24 Asian cases. Am J Surg Pathol 27: 1061-1069, 2003.