One Indurated Tumor on the Right Thigh in a 50-year-old Patient

Hsin-Chieh Tang Tun-Yuan Liu Chia-Chun Kao

CASE REPORT

One 50-year-old female complained of one painless skin lesion over the right thigh for 6 months. Dermatologic examination showed one ill-defined, indurated, erythematous to violaceous subcutaneous nodule on the medial side of the right thigh for 6 months (Fig. 1). She denied any systemic disease. Nobody had the similar skin lesions in her family. Other physical examination did not show any abnormal findings such as lymphoadenopathy or hepatosplenomegaly.

Pathologic examination revealed a nodular infiltration within middle to deep dermis (Fig. 2). More than 50% of them are large lymphoid cells with large irregular nuclei and prominent nucleoli (Fig. 3). Most of these large lymphoid cells (80%) were CD30 diffusely positive (Fig. 4). Other immunohistochemical stains including CD 3, CD45R0, CD20, CD15, CD56, CD68 and EMA (epidermal membrane antigen) were negative. Chest X-ray, computed tomography of the chest and abdomen, blood smear, and bone marrow biopsy did not have any abnormal findings.

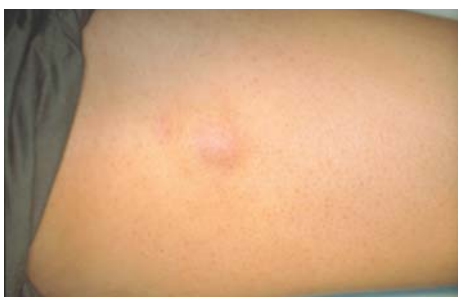

Fig. 1

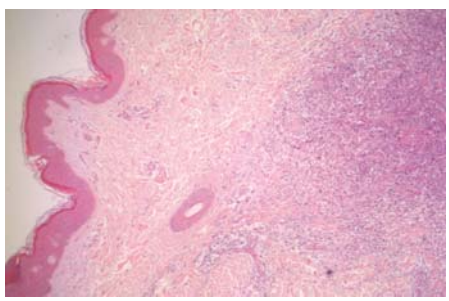

Fig. 2
H & E, x40

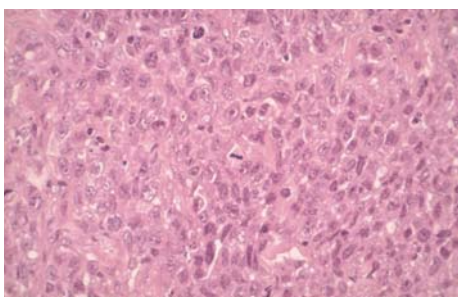

Fig. 3
H & E, x400

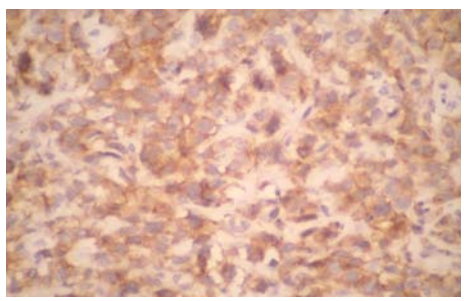

Fig. 4
CD30, x400

From the Department of Dermatology, Chung Shan Medical University Hospital
Accepted for publication: July 15, 2003
Reprint requests: Hsin-Chieh Tang, M.D., Department of Dermatology, No.110, Sec. 1, Jianguo N. Rd., Taichung, Taiwan, R.O.C.
TEL: 886-4-24739595 ext. 2338 FAX: 886-4-24739595 ext. 2332

DIAGNOSIS: CD30+ Primary Cutaneous Anaplastic Large Cell Lymphoma, Null Type

討　論

CD30，原名Ki-1，最早是由Stein等人所發現的一個細胞標記。由於CD30在一些淋巴球性或是組織球性的腫瘤，甚至是惡性黑色素瘤、精細胞瘤等皆會呈現出陽性反應，所以唯有仔細辨認病理下細胞型態的特徵及藉由特殊染色的方法，才能作出最後正確的診斷[1,2]。

在西元1985年，有一群作者描述了一個經常具有群聚性增生的大型多形性淋巴母細胞，並且在這些腫瘤細胞上都一致的表現出CD30這個分子。因此他們把它定義成一個新的淋巴瘤分類－退行發育大細胞淋巴瘤(anaplastic large cell lymphoma, ALCL)[1,2]。除了系統性(systemic ALCL, SALCL)的侵犯外，也可發生於皮膚，在皮膚上的臨床表現包括淋巴瘤樣丘疹症(lymphomatoid papulosis，LyP)、原發性皮膚ALCL(primary cutaneous ALCL, PCALCL)及繼發性皮膚ALCL(secondary cutaneous ALCL, SCALCL)。區分這一點對治療方式或預後來說相當重要，其預後由好到壞分別是LyP、PCALCL、SCALCL[3]。PCALCL好發在年紀較大的成年人，平均年齡是60歲。它佔整體皮膚淋巴瘤的比率將近9%。皮膚上的病灶通常是單一無症狀的腫塊，有時候可見到表皮會有潰瘍的情形出現；多發性的皮膚病灶比較少見，它們有可能是群聚的結節或是呈現廣泛的皮膚分佈[1,2]。

ALCL病理上的特徵通常是多形性的(pleomorphic)大型淋巴細胞，具有大且不規則的細胞核，明顯的核仁，與豐富、淡藍色的細胞質，有時可見到有絲分裂的情形。診斷ALCL的標準爲大型淋巴細胞的比例需超過30%，其中CD30陽性的比例需超過75%，絕大多數的病例會呈現T、NK細胞標記或是不呈現任何細胞標記的裸型細胞(null type)。但是如果染色夠多的話，或是做T細胞接受器重組(TCR rearrangement)的研究，可以發現很多的裸型細胞都可以歸類爲T細胞。由於大型淋巴細胞需與何杰金氏淋巴瘤(Hodgkin's lymphoma)作鑑別診斷，因此可染CD15來加以區分，一般R-S cell及Hodgkin's cell爲陽性反應，而在ALCL如同本病例爲陰性反應。

區分SALCL及PCALCL除了臨床上的分期之外，還包括了以下幾點。EMA在大部分的SALCL是陽性反應，而在PCALCL是陰性反應。另外有時SALCL會有第2及第5對染色體交換的情形，因此產生了新的融合基因NPM(nucleophosmin)-ALK(anaplastic lymphoma kinase)及新的蛋白質ALK；而在PCALCL則沒有這樣的情況[1,2]。

REFERENCES

1. Kinney MC, Kadin ME: The pathologic and clinical spectrum of anaplastic large cell lymphoma and correlation with ALK gene dysregulation. Am J Clin Pathol 111(Suppl. 1): S56-S67, 1999.
2. Stein H, Foss HD, Durkop H, *et al*.: CD30+ anaplastic large cell lymphoma: a review of its histopathologic, genetic, and clinical features. Blood 96: 3681-3695, 2000.
3. Bekkenk MW, Geelen FA, van Voorst Vader PC, *et al*.: Primary and secondary cutaneous CD30+ lymphoproliferative disorders: a report from the Dutch Cutaneous Lymphoma Group on the long-term follow-up data of 219 patients and guidelines for diagnosis and treatment. Blood 95: 3653-3661, 2000.