Primary cutaneous B cell lymphoma

Ishwara Bhat, C. Janaki, G. Sentamilselvi, V. R. Janaki
Department of Dermatology, Madras Medical College, Chennai - 600003, India.

Address for correspondence: Prof. Dr. V. R. Janaki, 1-D, Lakshmi Apartments, 122, Fourth Street, Abhirampuram, Chennai - 600018, India.
E-mail: dr_janakivr@rediffmail.com

ABSTRACT

A 54-year-old man, a road layer by occupation, presented with a 'leonine facies' and multiple tumors that were more commonly present over the exposed parts of the body. On investigation, he turned out to be a case of primary cutaneous B cell lymphoma with a distinctive histopathology. Chemotherapy was given with a good therapeutic response.

Key Words: Primary cutaneous B cell lymphoma, Grenz zone

INTRODUCTION

Cutaneous lymphomas, arising from T or B lymphocytes, constitute an uncommon but nevertheless important entity, with the so-called Skin Associated Lymphoid Tissue (SALT) being either the origin (primary) or the target (secondary) in the neoplastic process.[1] The primary cutaneous lymphomas, by definition, present clinically on the skin with no extracutaneous disease at the time of diagnosis or for 6 months thereafter.[2] Of these, primary cutaneous lymphomas of B cell origin are less common, their incidence being one-third that of their T cell counterpart.[3] Although several classifications like Kiel, Lukes and Collins, Rappaport and Revised European and American Lymphoma (REAL)[4] classification have been used, the one proposed by the European Organisation for Research and Treatment of Cancer (EORTC)[3] is the latest one and is being followed widely. The follicular centre cell type described in the EORTC classification is the commonest type of primary cutaneous B cell lymphoma and shows a characteristic histopathology[5] and expression of cell surface markers like CD 19, CD 20, CD 79a.[1,6] One such case is described below.

CASE REPORT

A 54-year-old male, who had been a road layer for 30 years, presented with a one-year history of progressively enlarging swellings over the face, chest and forearm. The lesions were mildly photosensitive.

On examination, multiple tumor masses of firm consistency were found infiltrating the forehead and malar areas to produce a 'leonine' appearance (Figure 1). The overlying skin was hyperpigmented and with a 'peau d'orange' appearance. Similar lesions were present on the neck, upper back and dorsal aspect of forearms. No significant lymphadenopathy was detected clinically.

Histopathology of the involved skin showed a normal epidermis with absence of epidermotropism. A clear subepidermal cell free Grenz zone was seen in the papillary dermis. The mid and lower dermis were densely infiltrated by small, round lymphocytes in a 'bottom heavy' pattern (Figure 2). Radiological and bone marrow aspiration studies revealed no extracutaneous pathology.

Immunohistochemistry for cell markers was done. A final diagnosis of primary cutaneous B cell lymphoma or follicular centre cell type was made based on immunohistochemistry with Bcl-2 antibodies.

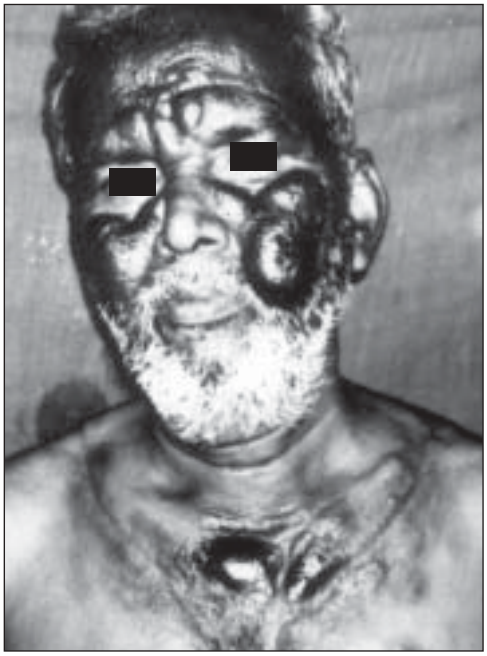

Figure 1: Tumor masses producing leonine facies and involving the upper chest

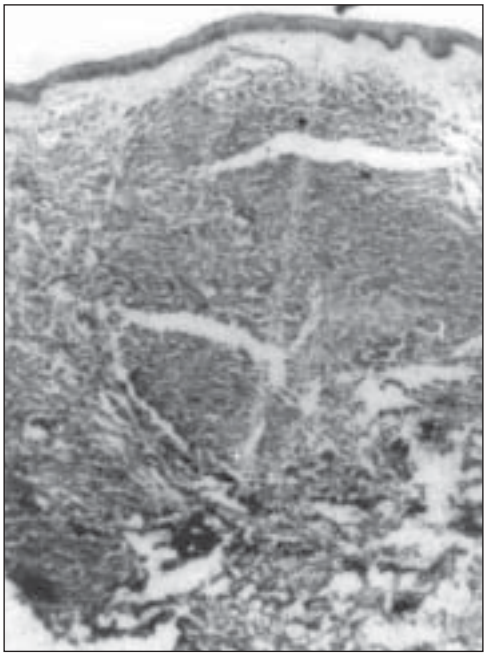

Figure 2: Histopathology showing the subepidermal grenz zone and a diffuse infiltrate in the lower dermis

Chemotherapy with cyclophosphamide, adriamycin, vincristine and prednisolone was started. An excellent response with near total resolution of lesions was seen (Figure 3). The patient is being followed up at regular intervals.

DISCUSSION

The follicular centre cell type of primary cutaneous B cell lymphoma with its classical histopathology[5] and cell markers, as seen in this case, has a favorable prognosis with good therapeutic response, a fact well documented.[2,7] Interestingly, with the patient's history of lifelong exposure to coal tar and sunlight, and with the lesions being limited to the exposed areas of the body, the possibility of these environmental factors playing a carcinogenic role in primary cutaneous B cell lymphoma, although hitherto unreported, needs to be further explored.

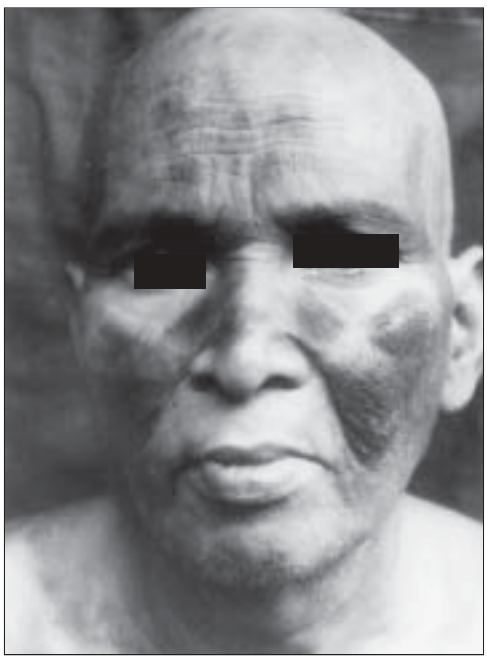

Figure 3: Resolution following chemotherapy

REFERENCES

1. Giannotti B, Santucci M. Skin associated lymphoid tissue (SALT) related B cell lymphoma (primary cutaneous B cell lymphoma): A concept and clinicopathological entity (editorial). Arch Dermatol 1993;129:353-5.
2. Sterry W, John S. Other systemic lymphomas with skin infiltrates. In: Freedberg IM, Fitzpatrick TB, et al, editors. Dermatology in general medicine. 5th ed. New York: McGraw-Hill; 1999. p. 1250-8.
3. Willemze R, Kerl H, Sterry W, et al. EORTC classification of primary cutaneous lymphomas: A proposal from the Cutaneous Lymphoma Study Group of the European Organisation for Research and Treatment of Cancer. Blood 1997;90:354-71.
4. Harris NL, Jaffe ES, Stein H, Barks PM, Char JK, Cleary ML, et al. A revised European–American classification of lymphoid neoplasm: A proposal from the International Lymphoma Study

Group. Blood 1994;84:1361-92.
5. Mackie RM. Cutaneous lymphomas and lymphocytic infiltrates. In: Champion RH, Burton JL, Burns DA, Breathnach SM, editors. Rook/Wilkinson/Ebling's Textbook of dermatology. 6th ed. London: Blackwell Science; 1998. p. 2393-5.
6. Willemze R, Meijer CJ, Sentis HJ, Scheffer E, Von Vloten WA, Toonstra J, et al. Primary cutaneous large cell lymphoma of follicular centre cell origin: A clinical follow-up study of 19 patients. J Am Acad Dermatol 1987;16:518-26.
7. Rijlaarsdam JV, Toonstra J, Meijer OW, Noordijk EM, Willemze R. Treatment of primary cutaneous B cell lymphoma of follicular centre cell origin – A clinical follow up of 55 patients treated with radiotherapy or polychemotherapy. J Clin Oncol 1996;14:549-5.