Case Report

Primary Cutaneous B Cell Lymphoma- Leg Type (NEW EORTC - WHO Classification), with nasal sinuses involvement

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Abstract

Primary Cutaneous lymphomas of B cell origin are rare, there remains a controversy in truly classifying these lymphomas and an updated EORTC classification divides them on the basis of their distinct histopthological grounds rather than on the basis of their anatomic location as in WHO classification, while the new WHO- EORTC joint classification maintains some characteristics of both systems, We report an elderly gentleman who primarily had a typical Leg dominant Cutaneous lymphoma of B cell origin uniquely with involvement of nasal Sinusues , bearing the Immunohistochemical staining features of " Cutaneous lymphoma - Leg Type" befitting the new joint WHO-EORTC classification of Cutaneous B cell Lymphoma.

Introduction

Cutaneous lymphomas are uncommon skin tumours, with skin being either the primary site for the origin of the tumour or more often the secondary site. Approximately

65% of the cutaneous lymphomas are T cell in origin and only 20-25% are thought to originate from the B cell, with majority of these being diffuse large B cell type.[1] Most of the primary cutaneous lymphomas have an indolent behaviour.[2]

Primary Cutaneous B-cell lymphoma (PCBCL) as an entity was introduced in 1990s to characterize a group of lymphoproliferative disorders characterized by clonal proliferation of B lymphocytes primarily involving the skin.[3] According to the Revised European and American Classification of Lymphoid Neoplasms (REAL) / World Health Organization (WHO) classification the Non Hodgkin's lymphomas, lymphomas which have large cell morphology and diffuse growth pattern are designated as diffuse large B cell lymphoma (DLBCL).[4] The European Organization for Research and Treatment of Cancer (EORTC) divided primary cutaneous B-cell lymphomas (PCBL) in four groups, one of them being The Primary cutaneous diffuse large B-cell lymphoma, 'leg type' [5]; They are a heterogeneous group of lymphomas that primarily involve the skin but may have variable clinical, histopathologic, and immunologic phenotypes.

We are reporting a case of an elderly patient, with Primary Cutaneous Large B cell lymphoma (PCLBCL) rather unique in having primarily the involvement of both lower limbs along with simultaneous involvement of paranasal sinuses extending to involve the inferior orbital area.

Case Report

A 70 year old gentleman, presented in April 2007 with a history of painless skin nodules that developed over two months. Initially these nodules were small and on the shin of left leg but gradually they progressed to involve the right leg, right arm and left hand and right side of nose. The

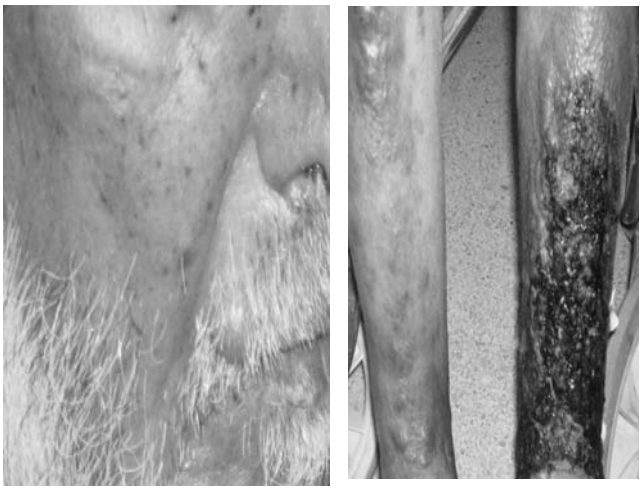

Figure 1: Appearance of the lesions on face and legs.

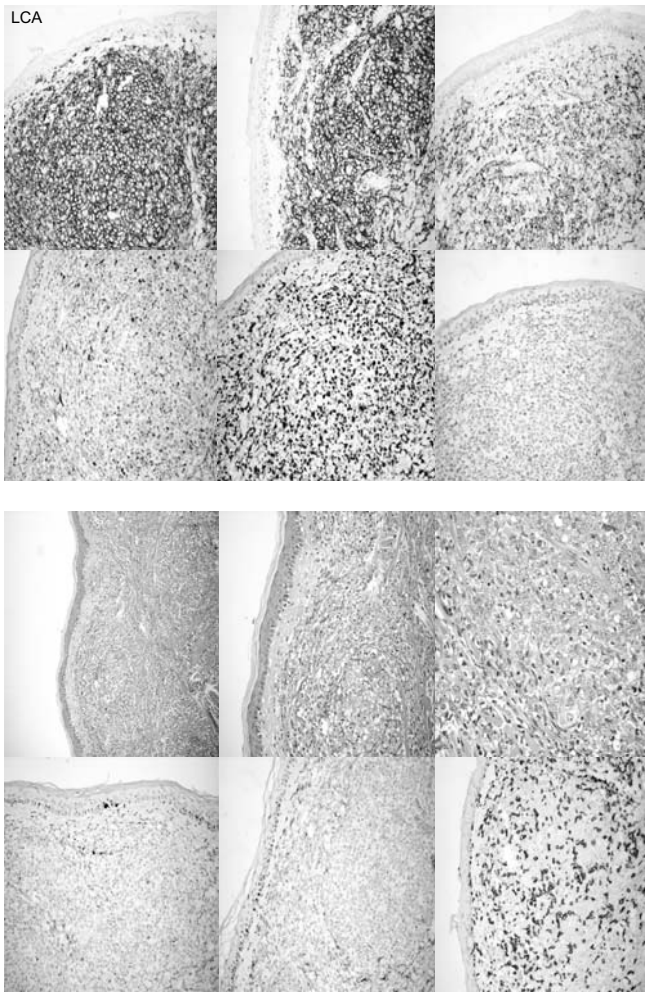

Figure 2. characterstic Immunocytochemical Stains and H & E stains: showing positivity to LCA, CD20, BCL2 and MUM 1, CD3 is characteristically Not stained.

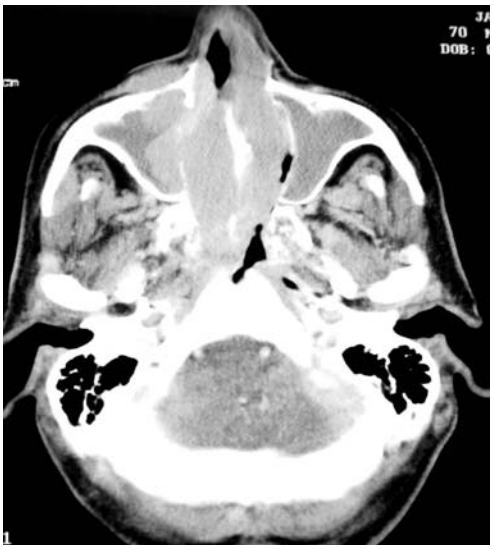

Figure: 3 CT scan of paranasal sinuses showing the extent of the lesion in paranasal sinuses.

skin covering these nodules first became black followed by development of blisters with serous discharge which finally became thick and crusted (Figure 1).

On examination there were bilateral blistering lesion at legs and on the dorsum of the right foot, crusted with mild serous discharge on bilateral shins, skin nodules on right side of nose, right and left forearm and swollen left middle finger. Rest of the systemic examination was unremarkable, his overall ECOG performance status was 2.

Laboratory investigations at the time of presentation were: Blood Urea Nitrogen: 19mg/dl, serum Creatinine: 0.9mg/dl, total Billirubin: 1.0 mg/dl, Gamma Glutamine transferease : 11 IU/l, Alanine Aminotransferase: 9 IU/l, Alkaline Phosphatase: 78 IU/l, serum Albumin: 2.7 mg/dl, serum Calcium: 7.6 mg/dl, serum Lactic Dehydrogenase:1373 I.U/l. Complete blood counts were Hb: 9.0 gm/dl, WBC: 7.1 x 10^9/l and Platelets: 262x 10^9/dl.

Biopsy specimen of leg lesions gave the diagnosis of diffuse large B cell lymphoma, with the immunohistochemical stains being positive for LCA, CD20, CD79, Bcl-2, Bcl-6, MiB 1, MUM-1 and characteristically negative for CD3 (Figure 2), (Figure 3).

CT scans of paranasal sinuses, chest and abdomen showed an enhancing soft tissue mass involving nasal cavity, paranasal sinuses, nasopharynx on right side with orbital extension and abdominal lymphadenopathy.

Discussion

Primary B cell Lymphoma of the leg, which was identified as a distinct entity in the EORTC classification, has now been incorporated in the new WHO-EORTC classification of cutaneous B cell Lymphomas as 'Primary cutaneous large B cell lymphoma, Leg Type', ending the longstanding controversy of placement and categorization of the primary cutaneous B cell Lymphomas.[6] The sub classification of PCBCL is now made on morphological features rather than anatomic site of presence and this is important to remember, as the type in question is apt to express Bcl-2, MUM-1, however and interestingly expression of the said immophenotypic markers was not associated with worse prognosis, as opposed to other types of Cutaneous B cell lymphomas namely follicular Cutaneous B cell lymphoma (PCFCLs).[7]

There however is a growing consensus that the primary cutaneous DLBCL of leg is distinct in its behaviour as opposed to the DLBCL occurring at the other sites in that these patients are of older age group, more frequently females, have a short duration of skin lesion before diagnosis.[2,8]

In a European multi centre study of 145 patients with primary cutaneous DLBCL, round-cell morphology, location on the leg, and multiple skin lesions at diagnosis were found to be independent adverse prognostic factors and these neoplasms are highly sensitive to radiotherapy[9] and hence, it is the treatment of choice for localized disease at presentation or relapse. Combination chemotherapy with Cyclophosphomide, vincristine, Adriamycin and prednisone (CHOP), with or without addition of Rituximab, is preferred in patients with the tumour involving the leg, multiple skin lesions or with systemic involvement.[1,9]

Our case is interesting in being a rare, histopathologicaly fitting the new WHO-EORTC classification of 'Primary Cutaneous diffuse large B-cell lymphoma, leg type' and with clinical involvement of Para nasal sinuses, as for primary Cutaneous lymphomas it's very unusual to involve other organ sites. The patient was managed in line of the recommended guidelines and he responded well to the treatment. This case reminds the practicing physicians of diversity with which lymphoma can present.

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