

Case Report

Primary cutaneous diffuse large B cell lymphoma: A Case Report

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DOI: 10.5455/jrmds.20153120

ABSTRACT

We present a case of 58 year old male with swelling on temporal region. Histopathologic and immunohistochemistry findings are compatible with a diagnosis of cutaneous diffuse large B cell lymphoma. We present a brief review of primary cutaneous DLBCLs and its relation to prognosis.

Keywords: DLBCL, WHO-EORTC, PCDLBCL-LT, PCDLBCL-OTHER, MIC-2, bcl-2, CHOP

INTRODUCTION

The skin is the second most common site of extranodal lymphoma, following the gastrointestinal tract. Lymphoma may involve the skin as the primary and only site of involvement or may spread to skin as a secondary site of disease [1]. The annual incidence of cutaneous lymphoma is estimated to be from 0.5 to 1 per 100000 persons per year. Cutaneous T cell lymphomas account for approximately 60% of primary cutaneous lymphomas, while 20% are B cell lymphomas [2]. Primary cutaneous diffuse large B cell lymphoma is an aggressive cutaneous B cell lymphoma (CBCL) that accounts for approximately 6% of all cutaneous lymphomas [6].

Primary cutaneous lymphomas have favourable prognosis, whereas those that are disseminated at presentation have unfavourable ones [3].

CASE HISTORY

This is a case of 58 year old male patient who presented with history of swelling over right temporal region since 3 months, progressively increasing in size.

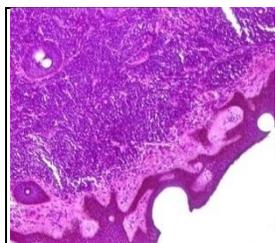


Figure 1a:

H&E Microphotograph
(4x)

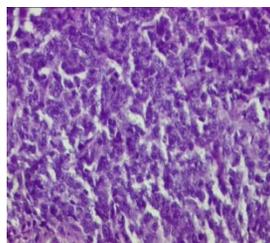


Figure 1b:

H&E Microphotograph
(40x)

On gross examination, it was a skin covered mass measuring 3x2x0.5 cm. On cut surface it was solid grey white. Histopathologic examination shows epidermis lined by stratified squamous epithelium. Underlying tissue shows a tumor composed of diffuse infiltrate of round to oval cells showing irregular vesicular nuclei, some with prominent nucleoli and scanty cytoplasm. At places mitotic activity is seen. (Fig: 1a & 1b). With immunostains these cells are diffusely positive for LCA, CD 20 and MIC2 and negative for EMA, AE1/AE3, S100 protein, Tdt, cyclin D1, CD34, CD3, C-kit. MIB-1 labelling index is 70-80% (Fig 2 & 3).

Bone marrow study was normal.

Figure 2: Immunohistochemistry photograph showing LCA +ve, CD20 +ve, CD3 -ve

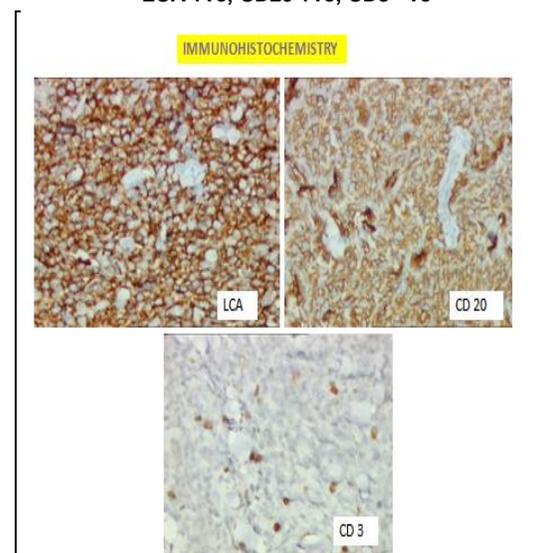
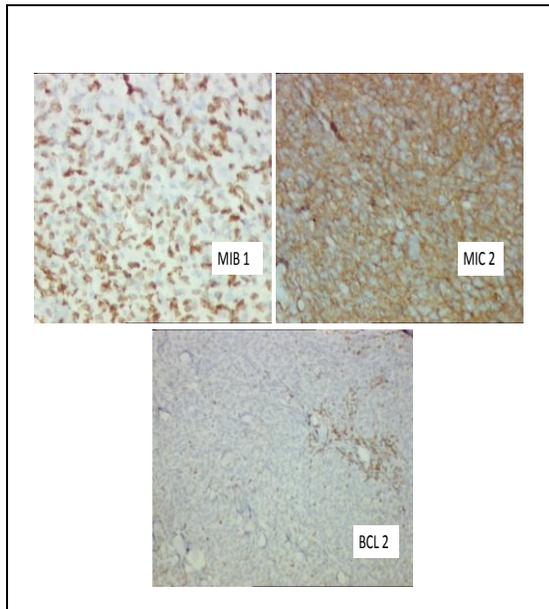


Figure 3: Immunohistochemistry photograph showing MIB1 labeling index 70-80%, MIC 2+ve, BCL2 -ve



On the basis of morphology and immunohistochemistry findings, the diagnosis given was primary cutaneous diffuse large B cell lymphoma.

DISCUSSION

Primary cutaneous B cell lymphoma differs clinically and has a different prognosis when compared to histopathologically similar systemic lymphomas. The world health organization – European organization for research and treatment of cancer (WHO-EORTC) defines PCBCLs as malignant lymphomas that are confined to skin at presentation after complete staging procedures. PCBCLs are categorized into five subtypes according to clinical behaviour: Primary cutaneous follicle center lymphoma; Primary cutaneous marginal zone B-cell lymphoma; Primary cutaneous diffuse large B cell lymphoma-leg type (PCDLBCL-LT), PCDLBCL-other and intravascular large B cell lymphoma [4].

PCDLBCL-LT presents as rapidly growing red or blue red tumours on one or both legs and often occurs in elderly women. In 10% of cases, lesions occur at sites other than leg. Histopathologic features of these lymphomas include a non-epidermotropic, diffuse infiltrate of centroblasts and immunoblasts in the dermis. Immunophenotypically neoplastic B cells express B cell associated antigens such as CD-20, CD79a, bcl-2, bcl-6 and MUM-1/RF4 protein [4]. Any

primary cutaneous large B cell lymphoma showing both bcl-2 and MUM-1 expression should be designated as PCDLBCL-LT independent of anatomic site [5]. PCDLBCL-LT type is characterized by predilection for the leg (72%), a high proportion of bcl-2 expression (85%), an advanced age at onset (mean age 76 years) and frequent relapses and extracutaneous dissemination. It is an entity with a poor prognosis particularly in patients with multiple tumours on the leg. The overall 5 year disease specific survival rate is 41% [6].

PCDLBCL-other includes Anaplastic or plasmablastic, T cell/Histiocyte rich large B cell lymphomas and the lymphomas which do not fulfil the criteria for PCDLBCL – LT type [4].

Primary cutaneous B cell lymphomas with the exception of large B cell lymphoma leg type and intravascular large B cell lymphomas are associated with an excellent prognosis. In primary cutaneous large B cell lymphoma, the location on leg, the round cell morphology (predominance of centroblasts and immunoblasts) and multiple skin lesions are identified as adverse prognostic factors [8].

In patients with DLBCL with the germinal center B cell subtype, positive expression of MIC-2 resulted in better 2 year event free survival and 2 year overall survival compared to negative expression of MIC-2 [7].

The t(14:18)(q32;q21) chromosomal translocation which leads to over expression of the bcl-2 antiapoptotic protein and is found in the majority of follicular lymphomas and in a lower percentage of systemic high grade DLBCLs, is not found in PCBCLs. The detection of t(14:18) translocation in cutaneous B cell lymphoma suggests the presence of systemic disease [8]. BCL-2 protein expression has been associated with poor prognosis in patients with noncutaneous DLBCLs. In one of the study the 5 year specific survival rates in bcl-2 positive and bcl-2 negative patients were 41% and 89% respectively [9].

In our case the prognosis is good because the single lesion is present on face, not associated with systemic disease and is MIC-2 positive and bcl-2 negative.

The standard treatment for patients with DLBCLs is cyclophosphamide, doxorubicin, vincristine and prednisone (CHOP). Rituximab, a chimeric monoclonal antibody against CD20 B cell antigen has

therapeutic activity in DLBCL. The rate of complete response was significantly better in the group that received CHOP plus rituximab than in the group received CHOP alone, 76% versus 63% [10].

After receiving chemotherapy, patient's condition is good without development of additional lesions, six months following treatment.

REFERENCES

- Burg G, Kaudewitz P, Klepzig K, Przybilla B, Braun-Falco O. Cutaneous B cell lymphoma. *Dermatol clin*1985;3:689-704.
- Porcia T Bradford, Susan S Devesa, William F Anderson, Jorge R Toro. Cutaneous lymphoma incidence patterns in the United States: A population based study of 3884 cases. *Blood* 2009;113(21):5064 – 73.
- Philip E, Leboit and Timothy H. Mc catmint. Cutaneous lymphomas and leukemias. *Lever's histopathology of skin*;32:805.
- Willemze R, Joffe ES, Burg G, Cerroni L, Berti E, Swerdlow H et al. WHO - EORTC classification for cutaneous lymphomas. *Blood* 2005; 105(10):3768-85.
- Plaza JA, Kacerovska D, Stockman DL, Buonaccors JN, Baillargeon P, Suster S et al. The histomorphologic spectrum of primary cutaneous diffuse large B cell lymphoma: A study of 79 cases. *Am J Dermatopathol* 2011; 33(7): 649-55.
- Florent Grange, Marie Beylot Barry, Phillipe Courville, Eve Maubec, Martine Bugot, Beatrice Vergier et al. Primary Cutaneous diffuse large B cell lymphoma, Leg type: Clinicopathologic features and prognostic analysis in 60 cases. *Arch Dermatol* 2007; 143(9):1144-50.
- Hong J, Park S, Park J, Jang SJ, Ahn HK, Sym SJ et al. CD 99 Expression and newly diagnosed diffuse large B cell lymphoma treated with rituximab- CHOP immunochemotherapy. *Ann Hematol* 2012;91(12):1897-906.
- Child FJ, Russell- Jones R, Woolford AJ, Calonje E, Photiou A, Orchard G et al. Absence of the t(14:18) chromosomal translocation in primary cutaneous B cell lymphoma. *Br. J. Dermatol* 2001;144(14)735-44.
- Grange F, Petrella T, Beylot M, Joly P, D'Incan M, Delaunay M et al. BCL-2 protein expression is the strongest independent prognostic factor of survival in primary cutaneous large B cell lymphomas. *Blood* 2004;103(10)3662-8.
- Bertrand coiffier M.D. CHOP chemotherapy plus Rituximab compared with CHOP alone in elderly patients with diffuse large B cell lymphoma. *N. Engl J. Med* 2002;346:235-42.

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Date of submission: 23/02/15

Date of acceptance: 05/03/15

How to cite this article: Gupta S, Ghorpade K, Patil V, Saraf A. Primary cutaneous diffuse large B cell lymphoma: A case Report. *J Res Med Den Sci* 2015;3(1):91-3.

Source of support: None

Conflict of interest: None declared