

Primary Cutaneous Large B Cell Lymphoma, Leg Type: Good Evolution of A Particular Location

Case Report

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Abstract

Primary cutaneous large B-cell lymphoma, leg type, is a rare and aggressive neoplasm as defined by the recently updated World Health Organization–European Organization for Research and Treatment of Cancer classification of cutaneous lymphomas. We present a case of a 39-year-old men who presented with a cutaneous lesion on her back. Skin biopsy revealed pathology consistent with this entity. The patient was treated with systemic chemotherapy with rituximab combined with doxorubicin, cyclophosphamide, vincristine, and prednisone with a good evolution. Here, we review the available literature and summarize clinical features and management of this uncommon subtype of non-Hodgkin lymphoma.

Keywords: B Cell Lymphoma; Leg Type; Cutaneous Lymphoma.

Introduction

Primary cutaneous B-cell lymphomas are a heterogeneous group of rare clonal B-cell lymphoproliferative disorders with distinct clinicopathological features compared with nodal counterparts [1]. Primary cutaneous diffuse large B-cell lymphoma-leg type (PCLBCL-LT) is a rare subtype which constitutes only 4% [2]. They have a predilection to the leg (72%), advanced age of onset (mean age, 76 years), high Bcl-2 expression (85%) and frequent relapses with extracutaneous dissemination [10-20%]. We report a case we report the case of a young patient of 39 years old who consulted for a hummocky tumor in the back whose histology was in favor of lymphoma B-type leg, with good response to rituximab combined with doxorubicin, cyclophosphamide, vincristine, and prednisone (R-CHOP).

Case Report

A 39 year old men, presented with asymptomatic erythematous tumors hummocky, with a hard texture to touch, gradually increasing size (Figure 1). Histopathological examination revealed diffuse atypical, pleomorphic cells, menues hyperchromatic nuclei anisokaryosis and many mitoses (Figure 2). Immunohistochemistry (IHC) revealed expression CD20 (Figure 3), le Bcl 6 (Figure 4), and focally Bcl2 (Figure 5), Proliferative index ki67 was very

high, anti-CD3 antibody labeled reaction cells, CD10-, CD4-, and CD30-. Based on the clinical, histopathologic and the IHC examination, a diagnosis of primary cutaneous diffuse large B-cell lymphoma, leg type (PCLBCL, LT) was established. Lymph nodes were free, all evolving in a context of apyrexia and conservation condition. Further staging evaluation, including bone marrow biopsy and computed tomographic imaging, was unremarkable. The patient was subsequently started on systemic chemotherapy with rituximab combined with doxorubicin, cyclophosphamide, vincristine, and prednisone (R-CHOP) for eight cycles with a good evolution keeping just a scarred macula pigment in the back.

He tolerated therapy well and remains free of disease approximately 1 year after her lymphoma diagnosis (Figure 6).

Discussion

Primary cutaneous B-cell lymphoma (PCBCL) belongs to a distinct group of B-cell lymphoproliferative disorders defined by its presentation in the skin, without evidence of extracutaneous spread at the time of diagnosis [3]. Extranodal involvement occurs in approximately 25% of non-Hodgkin lymphomas, with the gastrointestinal tract being the most common site of extranodal involvement, followed by the skin [4]. The annual incidence of cutaneous lymphomas is approximately 0.5 to 1 per 100,000

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Received: August 22, 2016

Accepted : October 03, 2016

Published: October 06, 2016

Citation: A Lahlou, S Elloudi, H Baybay, FZ Mernissi (2016) Primary Cutaneous Large B Cell Lymphoma, Leg Type: Good Evolution of A Particular Location. *Int J Clin Dermatol Res.* 4(3), 90-93. doi: <http://dx.doi.org/10.19070/2332-2977-1600024>

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Figure 1 . Erythematous tumors hummocky on the back.



Figure 2 : Dermal location of a diffuse and dense infiltrate, not epidermotropic, monomorphic (hematoxylin eosin saffron $\times 20$)

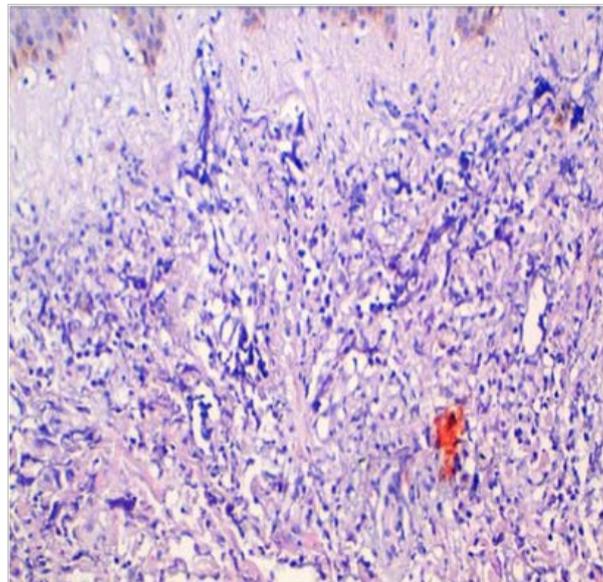


Figure 3 : Positive IHC with the anti-CD20 antibody

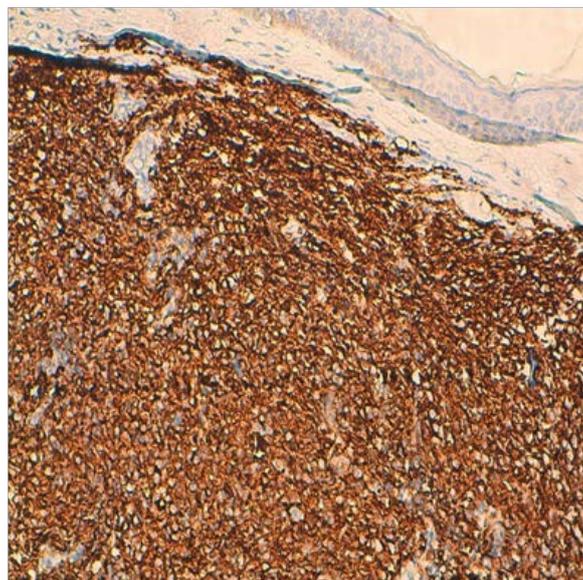


Figure 4 : Positive IHC with the anti-BCL2 antibody



Figure 5 : Positive IHC with the anti-BCL6 antibody

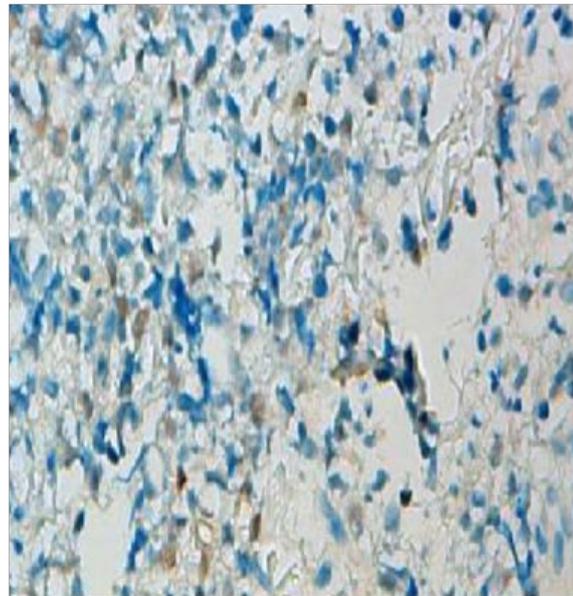


Figure 6 : Clinical improvement after treatment with R-CHOP



[3]. While B-cell lymphomas account for the majority of nodal lymphomas, PCBCls represent only 20% to 25% of all primary cutaneous lymphomas [5]. The pathogenesis of PCBCl is unclear. There is some speculation that PCBCl may represent a lymphoproliferative response to antigenic stimuli in the cutis, a skin-associated lymphoid tissue-related B-cell lymphoma [a process similar to mucosa-associated lymphoid tissue lymphomas in the gastrointestinal tract] [6]. In Europe, there is evidence linking *Borrelia*.

Burgdorferi infection to the development of PCBCl [4]. Jelic and colleagues reported borrelial serology in 12 of 22 (55%) PCBCl cases [7]. Another study evaluating cases of PCBCl in an area with endemic borrelia infections in the Scottish Highlands found *B. burgdorferi*-specific DNA in 35% of cases using polymerase chain reaction, with a statistically significant positive association between *B. burgdorferi* and PCBCl [8].

The subtype identified in our patient, PCLBCL, LT, represents approximately 20% of all PCBCls and 4% of all cutaneous lymphomas [9, 10]. It is more common in the elderly, with a median age in the mid 70s [11, 12]. But our patient was young. The male to female ratio ranges from 1:3 to 1:4 [10, 12, 13]. Patients with PCLBCL, LT present with red to bluish nodules or tumors on one or both lower legs. Only about 10% to 15% of these patients are noted to develop lesions outside of the lower extremities, as was the case with our patient, who presented with a tumor on the back [10]. Compared with other subtypes of PCBCls [primary cutaneous marginal zone lymphoma and primary cutaneous follicular center lymphoma], these tumors are more aggressive with worse outcomes, as they frequently disseminate to lymph nodes and visceral organs [1]. PCLBCL-LT typically realize tumors rapidly increasing in size, often multiple at diagnosis may be unilateral or bilateral in the lower limbs. Other areas may be affected, including the cephalic extremity and trunk as our patient, The extra leg location and uniqueness of the tumor are factors of good prognosis [13, 14]. Other factors such as young age in our patient may be involved. In fact studies have shown a survival rate at 3 years of 77% for other anatomical localization interesting seats against 43% for forms affecting lower limbs 4 and 87% in patients treated with rituximab and chemotherapy against 50% who received other types of treatment such as radiotherapy and chemotherapy [16, 17].

Conclusion

Lymphomas primitive leg skin types B have a poor prognosis, with a rapidly unfavorable evolution, we think that young age and topography outside the leg remains good prognosis if treated with R-CHOP chemotherapy as like our patient.

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