

Atypical Localization of a Diffuse Primary Cutaneous B Lymphoma with Large Cells of Leg Type

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Abstract

Cutaneous B-cell lymphomas are a malignant proliferation of lymphocytes of the B-cell type. Mutation occurring at different points in B cell development leads to different forms of lymphoma. Rarely, it presents as verrucous plaques or widespread garland-like lesions. They are typically red to bluish. They are mainly found on one or both legs but it affects other sites in 10–15%. It represents an aggressive subtype of PCBCL. Primary cutaneous diffuse large B-cell lymphomas of leg type are rare, occur mainly in very old women, preferentially localize in the lower limbs and in 10 to 15% of cases, they are localized in another part from the body. We describe a case in a 66 year old man with an atypical localization in the left shoulder.

Keywords: Cutaneous B lymphoma • Rare Lymphoma • Lymphoma • Atypical location

Abbreviations: PCBCLL: Primary Cutaneous B-Cell Lymphoma Lower Limb Type • GFELC: French Group for the Study of Cutaneous Lymphomas

Introduction

Cutaneous B lymphomas, less common than cutaneous T lymphomas, represent 20 to 25% of all cutaneous lymphomas [1,2]. The diffuse large-cell type B skin type leg usually occurs in women after age 70 and electively affects the lower limbs. Most often manifests as multiple, growing, purplish red, localized or scattered tumors. We report a case in a 66-year-old man with an atypical localization in the trunk.

Case Report

A 66-year-old patient with no history consults for confluent nodules gradually increasing in size in the left shoulder, which appeared 6 months ago without significant triggering factor or notion of insect bite, evolving in a context of conservation of the general condition. The dermatological examination on admission had objectified a firm purplish erythematous tumor measuring 6 cm in diameter centered in an ulceration, fixed relative to the deep plane surrounded by erythematous nodules of different confluent sizes taking the left shoulder (Figure 1), with presence of several papules and plaques of different size with a firm consistency in the central region of the back (Figure 2).

Dermoscopy showed irregular linear serpiginous vessels and a salmon-orange area (Figure 3). In addition, there were no clinically palpable lymphadenopathies.

The evolution was marked by a rapid increase in the size of the tumor in 15 days (Figure 4).

Histological examination of a biopsy performed at the periphery of the shoulder tumor showed a dense and diffuse dermal lymphocytic infiltrate reaching the hypodermis, consisting of large cells (Figure 5), with high mitotic

activity (marking of Ki67 > 70% of tumor cells) (Figure 6) a positive immunohistochemical staining with CD20 + BCL2 + BCL6 + MUM1 + and CD10– (Figures 7 and 8).

The evaluation of the extension by abdominal thoraco-thoracic computed tomography, an ultrasound of the lymph nodes and a biological evaluation made of NFS, LDH, Beta-2-microglobulin and EPP were completely normal as well as a bone marrow biopsy without signs of invasion confirming the primitive cutaneous character of large diffuse B-cell lymphomas. Polychemotherapy with cyclophosphamide, vincristine and prednisone, in combination with rituximab "RCHOP" was established after a normal pre-chemotherapy evaluation.



Figure 1. Purplish red tumor centered in ulceration, surrounded by confluent erythematous nodules of confluence taking the left shoulder about 6cm.

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Figure 2. Papules and plaques of different size with a firm consistency in the central region of the back.

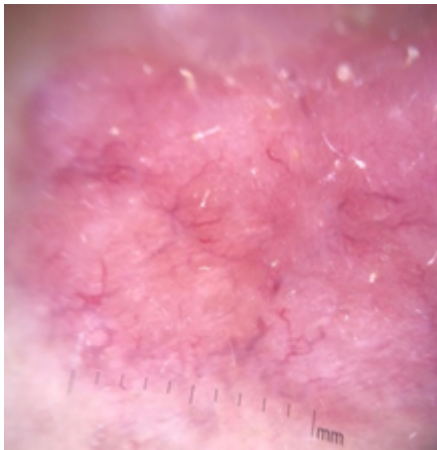


Figure 3. Irregular linear serpiginous vessels and a salmon-orange area objectified by dermoscopy.



Figure 4. Rapid increase in tumor size in 15 days comparing to Figure 1.

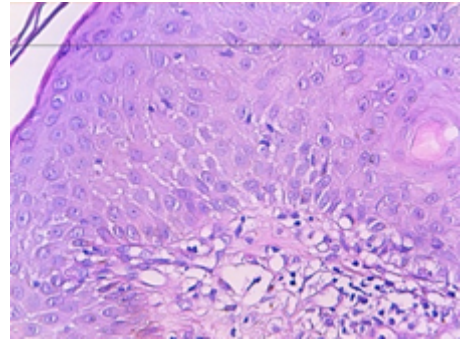


Figure 5. Dense and diffuse dermal lymphocytic infiltrate reaching the hypodermis, consisting of large cells.

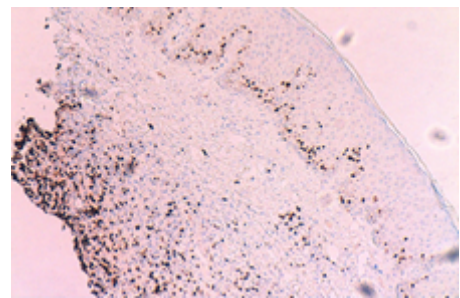


Figure 6. Ki 67 marks 70% of tumor cells in this immunohistochemistry image.

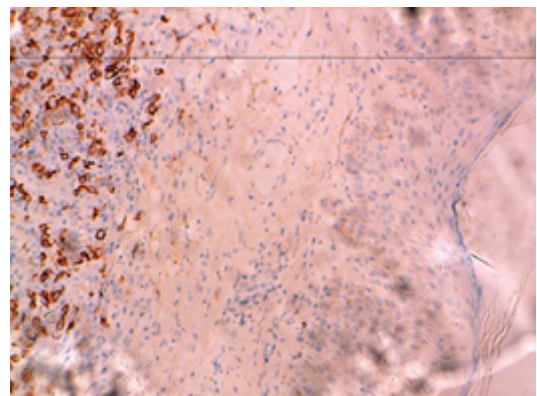


Figure 7. Histological image showing no epidermotropism with a positive immunohistochemical staining for CD20+.

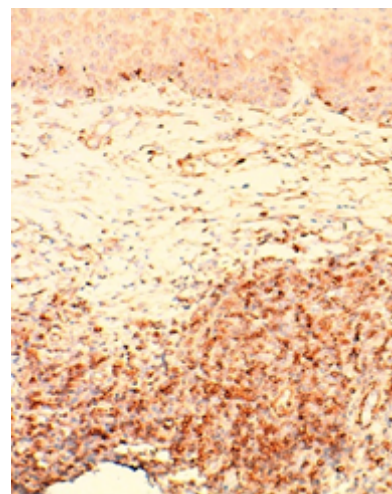


Figure 8. Immunohistochemical image showing positive staining with BCL2+.

Discussion

PCBCLL are defined as diffuse proliferations, consisting of layers of large B cells with rounded nuclei, immunoblasts and/or centroblasts [1,3], they occur mainly in the lower limbs in elderly subjects (Average age: 76 years), with a female predominance which is not the case for our patient. Clinically, they develop rapidly increasing, often multiple, purplish red in color, often ulcerated, localized or disseminated, which can affect the lower limb, both, or other anatomical regions. Pathology examination shows a diffuse and monotonous infiltrate of the dermis, often extended to the hypodermis, consisting of confluent layers of large round cells with a large central nucleolus (immunoblasts) or peripheral (centroblasts) with numerous mitoses [3,4] The immunohistochemistry allows to highlight a profile B (CD20+), bcl2, Mum-1/IRF4 + and FoxP1 positive, bcl6 is expressed in 50% to 68% of the cases but the expression of CD10 is exceptional (0-2%) [5]. This type of lymphoma is of intermediate malignancy. Our case describes an atypical location, and the site of the lesions is a prognostic factor, the specific 3-year survival rate is 77% in tumors that do not affect the lower limbs (compared to 43% for the lower limbs) [6]. The prognosis is linked to lymph node or visceral (especially cerebral) dissemination but also to skin progression responsible for deterioration of the general state or septic complications. However, these factors do not affect the treatment [7], which remains the same in both cases: RCHOP first-line chemotherapy in the absence of comorbidity, which is the case in our patient. The GFELC has clearly demonstrated the advantage of systematically associating rituximab with polychemotherapy of the CHOP type, which makes it possible to obtain a better result than the CHOP alone, in particular with regard to the relapse rate, while tolerance is not fundamentally changed. This scheme should therefore be preferred on the first line. Lenalidomide could represent an interesting solution in the second line [8].

Conclusion

PCBCLL is a rare entity with a poor prognosis; our case illustrates an atypical form in terms of presentation and location. The treatment is essentially based on multidrug therapy with a fairly satisfactory decline in the literature.

Conflicts of Interest

Author declares that there is no conflict of interest.

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