

Case Report

A rare case of primary cutaneous diffuse large B cell lymphoma, leg type with metastasis

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ABSTRACT

Primary cutaneous diffuse large B cell lymphoma, leg type (PCDLBCL-LT) is a rare and aggressive type of primary cutaneous B cell lymphoma (PCBCL), which represents 10-20% cases of PCBCL. It has a 40-50% recurrence rate and 5 year survival rate of 50%. Here, we present a case of an 86 year old female who presented to us with complaints of slightly tender annular plaques with an oedematous base present over bilateral lower limbs and pitting oedema. Histopathological examination from the annular lesion showed normal epidermis, grenz zone and a dense lymphoid infiltrate involving almost the entire dermis. Immunohistochemistry confirmed histological findings, atypical cell were positive for CD20 and MUM1 protein with focal expression of BCL 6 which is rare. Based on the above findings, we made a diagnosis of diffuse large B cell lymphoma-leg type and started her on palliative radiotherapy. As PCBCL-LT is rare and aggressive lymphoma, we present this case to review literature and summarise its clinical features.

Keywords: Lymphoma, Radiation, Immunohistochemistry, Cutaneous lymphoma

INTRODUCTION

Primary cutaneous lymphomas are a rare group of non-Hodgkin lymphomas that include a heterogenous group of cutaneous T-cell lymphomas (CTCLs) and cutaneous B-cell lymphomas (CBCLs).¹ PCDLBCL-LT is a rare and aggressive subtype of PCBCL and represents 10-20% cases of PCBCL.² The basis for making a diagnosis is a clinico-pathological correlation, including the use of several immunohistochemical markers and molecular biological methods.

CASE REPORT

An 86 years old female came with complaints of swelling over bilateral lower limbs since the past 8 months. The lesions were asymptomatic initially, which later increased in size to form annular plaques with fluid filled lesions over it. There was no history of fever, weight loss or night sweats. Cutaneous examination revealed multiple erythematous, slightly tender annular plaques with an oedematous base present over bilateral lower limb. Pitting oedema was present till mid shin. Rest of the cutaneous and systemic examination was within normal limits. Laboratory investigation showed low haemoglobin (9 gm%), normal blood sugars, raised serum LDH levels (553 U/l) and raised uric acid levels 8 mg/dl. Histo-

pathological examination from the annular lesion showed normal epidermis, Grenz zone and a dense lymphoid infiltrate involving almost the entire dermis. PET scan revealed extensive metabolically active dermal and subdermal lesion in bilateral calves, more on the right side suggestive of the primary neoplasm. Isolated metabolically active lymph nodes in right axilla and lower activity in the left inguinal lymph node suggested metastasis. Immunohistochemistry confirmed the histopathology findings by showing atypical lymphoid cells which were positive for CD20, MUM1 and focally positive for BCL-6. The lymphoid cells were negative for CD10, CD56. CD3 stain showed admixed T lymphoid cells.



Figure 1: Annular erythematous tender plaques with an oedematous base on bilateral lower limbs.

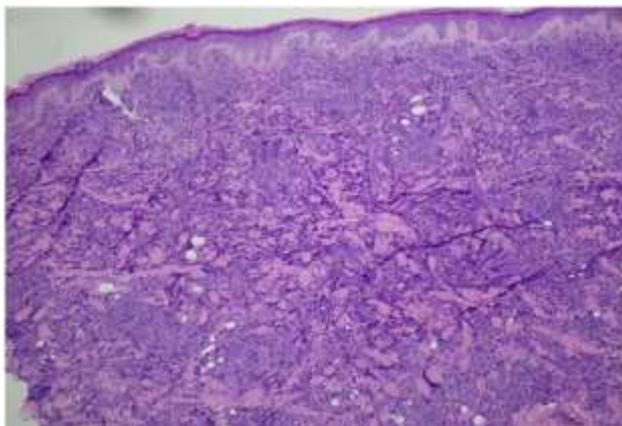


Figure 2: Histopathology showing no evidence of epidermal involvement and a Grenz zone; there is a dense lymphoid infiltrate involving almost the entire dermis.

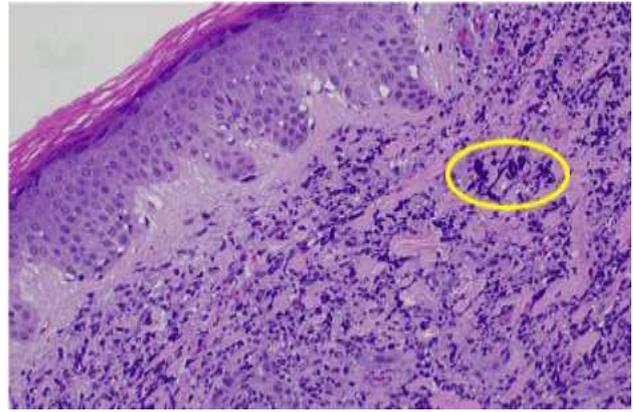


Figure 3: Histopathology-higher magnification shows hyperchromatic nuclei with atypical mitoses (yellow circle).

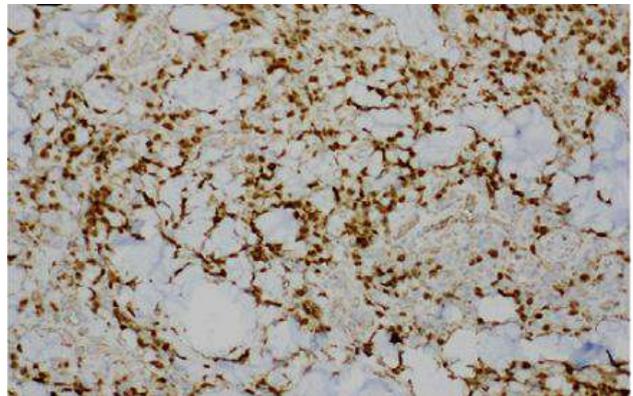


Figure 4: Immunohistochemistry; the atypical lymphoid cells are positive for CD20, mum1 and focally positive for BCL-6.; they are negative for CD10 and CD56; CD3 reveals few admixed T lymphoid cells; MIB1 labelling index is 70-80%.



Figure 5: Post treatment: after 2 cycles of palliative radiation using 8 Gy of external beam radiation therapy.

Based on the clinical, histopathological, immunohistochemistry and radiological findings we made a diagnosis of diffuse large B cell lymphoma-leg type and started her on palliative radiotherapy considering the age and systemic metastasis. Till now she has received 2 cycles of 8 Gy using external beam radiation therapy and has had a good response.

DISCUSSION

PCBCL is a group of lymphoproliferative disorder which involves the skin without any other organ involvement.¹ According to the new 2008 WHO-european organization for research and treatment of cancer (WHO-EORTC) classification, PCBCL has three main subtypes: primary cutaneous marginal zone lymphoma (PCMZL), primary cutaneous follicular centre lymphoma (PCFCL) and primary cutaneous diffuse large B cell lymphoma leg Type (PCDLBCL-LT).¹

PCDLBCL-LT is a rare and aggressive subtype of PCBCL and represents 10-20% cases of PCBCL. Clinically it presents as brownish to red tender nodules over the legs in elderly females which can be bilateral. Sometimes it affects the trunk as the primary site.²

The pathogenesis of PCDLBCL is unknown. It is postulated that a lymphoproliferative response secondary to antigenic stimuli in the dermis plays an important role. Infections like *Borrelia burgdorferi* may lead to the development of DLBCL.¹

Histopathologically, PCDLBCL-LT is characterized by a Grenz zone separating the epidermis and the dermis and a dense and diffuse infiltrate throughout the dermis and subcutaneous tissue. The infiltrate contains large B-cells with roundish nuclei, prominent nucleoli and open chromatin, resembling centroblasts and immunoblasts, which are arranged in confluent sheets with few anaplastic cells.³ Immunohistochemistry of PCDLBCL LT is positive for CD20, MUM1 protein and BCL 2 and negative for CD3 and TdT.⁴ Our case was positive for CD20, MUM1 protein with focal BCL6 expression.

Multiple skin lesions (other body sites apart from the legs) disseminated lesion on the legs and an age above 75 years are poor prognostic factors while variables such as gender, BCL 2, MUM1 protein expression, serum LDH levels and duration of lesions have no effect on survival.⁵ As it is an aggressive type of lymphoma, it requires effective treatment such as anthracycline containing chemotherapy along with rituximab, depending upon the age and comorbidities. Ibrutinib and lenalidomide are

more efficacious in management of relapsing and refractory cases of PCDBCL-LT.⁶

It is important to identify such cases early and work in conjunction with an oncologist for proper management.

CONCLUSION

PCDLBCL-LT is a rare type of PCBCL which can be diagnosed on histopathological supplemented by immunohistochemical. It is however atypical to find BCL-6 positivity on immunohistochemistry and one needs to be aware of this while doing a clinicopathological correlation. As it is an aggressive type of lymphoma, it requires effective treatment such as anthracycline containing chemotherapy along with rituximab, depending upon the age and comorbidities. Ibrutinib and lenalidomide are more efficacious in management of relapsing and refractory cases of PCDBCL-LT.

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