

A Rare Case of Primary Cutaneous Diffuse Large B- Cell Lymphoma- Leg Type with Testicular Infiltration in a Young Male

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Abstract: *Primary cutaneous B-cell lymphomas (PCBCLs) are a heterogenous group of rare clonal B-cell lymphoproliferative disorders with distinct clinicopathological features compared with nodal counterparts. It is less frequent than lymphomas of T-cells. Primary cutaneous diffuse large B-cell lymphoma-leg type (PCLBCL-LT) is a rare subtype which constitutes only 4%. It is predominantly seen in elderly people and has a female preponderance. We report a case of PCLBCL-LT with testicular infiltration occurring in a young male who presented with three painless skin swellings over thigh, chest and upper lip and with bilateral scrotal swelling.*

Keywords: skin, leg tumors, B-cell, lymphoma, testicular infiltration

1. Introduction

Primary cutaneous lymphoma refers to cutaneous T-cell lymphomas (CTCLs) and cutaneous B-cell lymphomas (CBCLs) that present in skin with no evidence of extra cutaneous disease at the time of diagnosis or even after 6 months of the diagnosis.^[1,2]

Primary cutaneous B-cell lymphoma are less frequent than cutaneous T-cell lymphomas with incidence rates varying considerably between European (ranges from 20-25% of all cutaneous lymphoma) and north American studies (ranges from 3.2 to 7.7%).^[3]

As per WHO-EORTC classification for primary B-cell cutaneous lymphomas, they are classified in five types: Marginal zone primary cutaneous B-cell lymphoma, centrofollicular primary cutaneous lymphoma, Diffuse large B-cell primary cutaneous lymphoma-leg type, Large b-cell primary cutaneous lymphoma-other type, intravascular large B-cell primary cutaneous lymphoma. The diagnosis is made by the clinicopathological correlation, immunohistochemical findings and molecular pathology.^[4,5]

The primary diffuse large B-cell cutaneous lymphoma-leg type (PDLBCL-LT) comprise 5-10% of the B-cell cutaneous lymphomas, often affecting the lower limbs and less frequently other areas. It shows relapses with extracutaneous dissemination in 10-20% cases.^[2] Visceral dissemination includes central nervous system, eye, testis, peripheral blood, bone marrow and paranasal sinuses.^[6]

2. Case Report

A 28 year male presented with non-ulcerated plaque over left thigh and two nodular swellings, one each over chest and upper lip since 3 months [figure 1]. Scrotal swelling was also present for the same duration [figure 2]. The lesions were slowly increasing in size. On examination, the left thigh

swelling was the largest, measuring around 6×5 cm in size, firm in consistency, non-tender and free from the underlying structures. Scrotal USG showed enlarged size of both the testis with bilateral epididymo-orchitis. His hematological profile was normal.

Fine-needle aspiration cytology was done from skin swellings which showed moderately cellular smears with discohesive atypical cells. Cells were pleomorphic with hyperchromatic coarse nucleus, indistinct nucleoli and moderate amount of cytoplasm in the background of lymphocytes and given as features highly suspicious of cutaneous lymphoma [figure 3]. FNAC from scrotal swelling also showed the same features [figure 4]. Later, Edge biopsy was performed from the thigh swelling and material was sent for histopathological examination which revealed dense diffuse non epidermotropic infiltration of atypical lymphoid cells involving variable proportions of centroblasts and immunoblast like cells and a clear zone between epidermis and dermis. These cells in dermis were round to oval with scant cytoplasm, pleomorphic vesicular nuclei and prominent nucleoli. Dispersed among these large cells were few small lymphoid cells [figure 5 and 6].

With the presumptive diagnosis of cutaneous lymphoma, Immunohistochemistry was carried out which revealed positivity for CD20 showing B-cell origin of tumor cells [figure 7 and 8]. They showed negativity for CD3 thus ruling out the possibility of T-cell lineage. Also the cells were negative for CD5 and CD10 which ruled out the possibility of other B-cell variants.

A final diagnosis of primary diffuse large B-cell lymphoma-leg type with testicular infiltration was made but patient refused treatment after diagnosis and he expired after 3 months.



Figure 1: non ulcerated plaque in left thigh and two nodular swellings over chest and upper lip



Figure 2: Scrotal Swelling

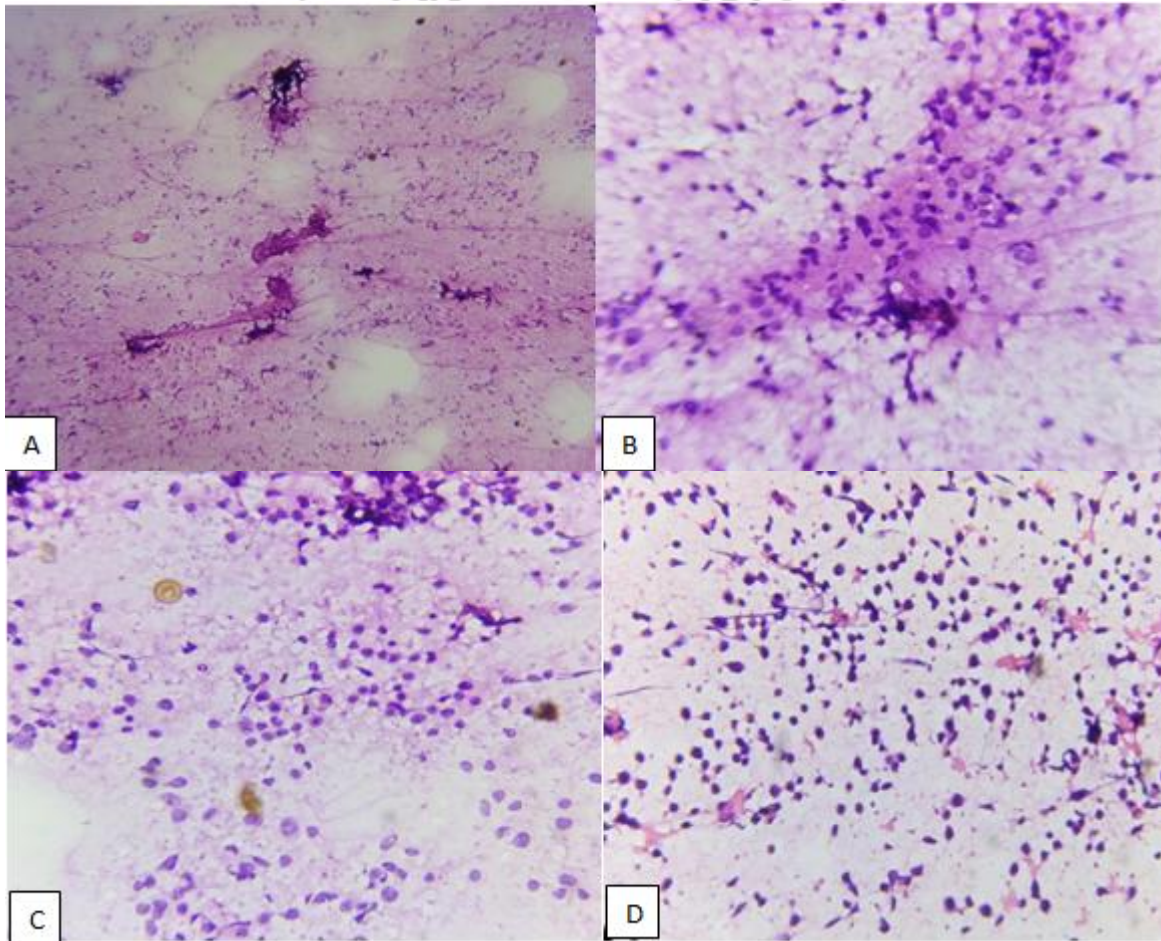


Figure 3: [A] FNAC from skin swellings showing moderately cellular smear with atypical cells (H&E,4×) [B], [C], [D] High power view showing pleomorphic cells with hyperchromatic dense nuclei, indistinct nucleoli and moderate amount of cytoplasm in the background of lymphocytes (H&E, 10×)

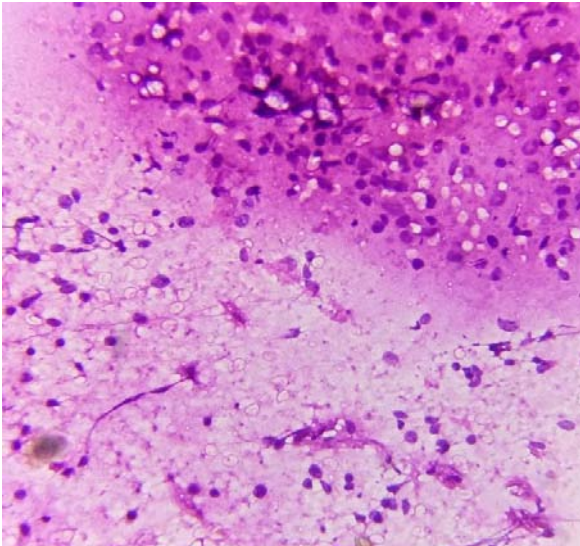


Figure 4: FNAC from scrotal swelling showing atypical cells in the background of lymphocytes (H&E, 10×)

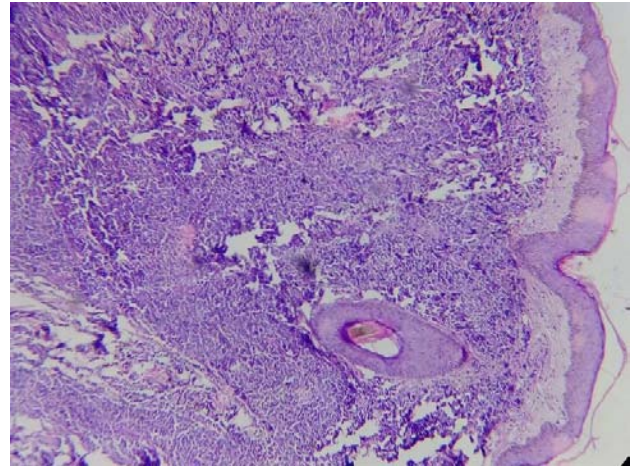


Figure 5: Low power view showing dense diffuse infiltration of tumor cells in papillary and reticular dermis sparing epidermis (non-epidermotropic) and clear zone between epidermis and dermis is seen

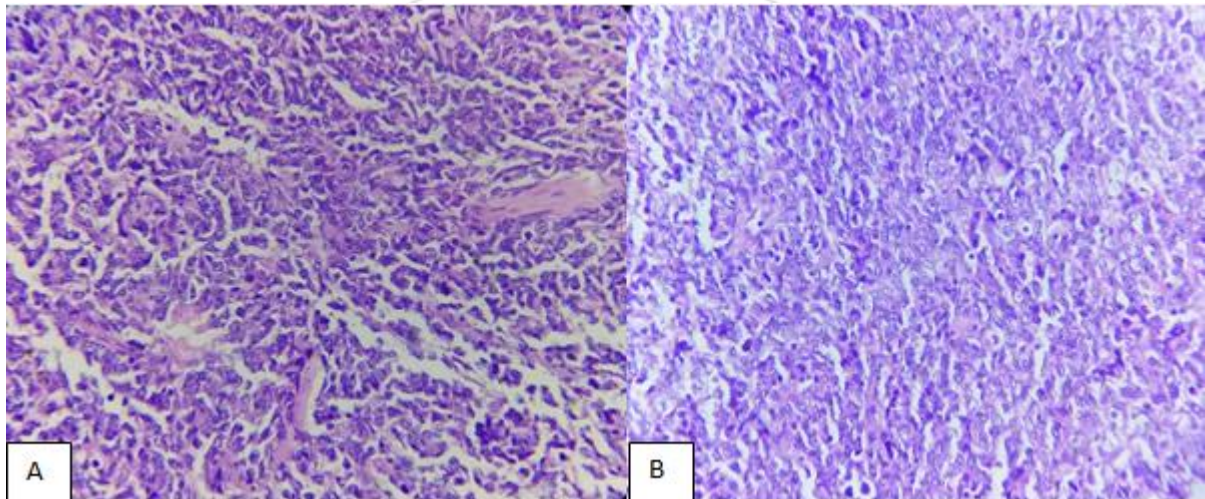


Figure 6: [A], [B] Round to oval cells with scant cytoplasm in the background of small lymphocytes (H&E, 10×) [C] Cells with vesicular nuclei and prominent nucleoli, few mitotic figures are also seen (H&E, 40×)

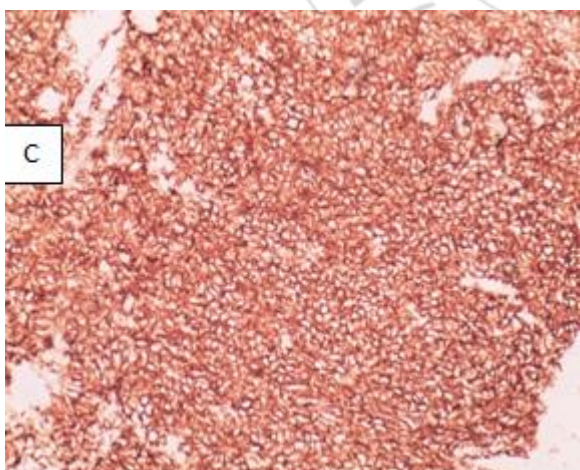


Figure 7: Immunohistochemistry showing CD20 positivity (cytoplasmic membrane) by tumor cells

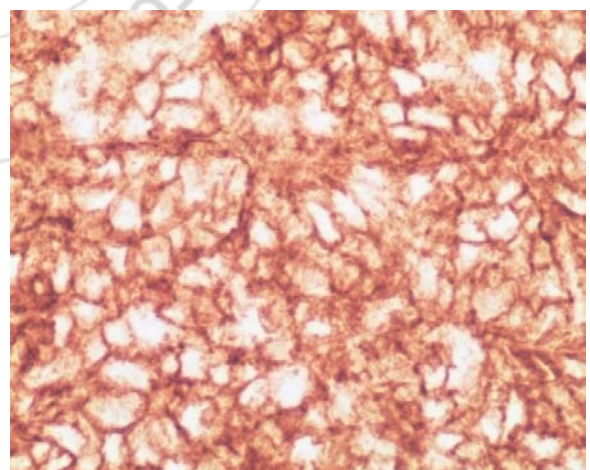


Figure 8: High power view of CD20 positivity

3. Discussion

Approximately 27% of non-Hodgkin lymphomas occur in extranodal sites, skin being second most common site.^[7] Sites other than the leg have also been reported. Incidence over trunk is found to be 7.5-13.3% of all PCLBCL-LT.^[11]

Primary cutaneous DLBCL of leg differs from non-cutaneous DLBCL occurring at the other sites. Patients with cutaneous DLBCL are more frequently females in an older age group presenting with a short duration of skin lesion and often with extra-cutaneous disease. Histopathologically cutaneous DLBCL are composed predominantly of round blasts often expressing bcl-2 protein cells as opposed to cleaved cells seen in non-cutaneous DLBCL.^[8] In cutaneous DLBCL patients have reduced disease free survival and a worse prognosis.^[8,9] Lesions may disappear completely but recurrences are commonly seen in this disease.^[8,10]

Our case is rarely reported in any literature because a young male patient is affected. Moreover the lesion was non-ulcerated and with testicular involvement which is an uncommon finding in PCLBCL-LT.

Prognosis of PCLBCL-LT depends on the features such as age of the patient, presence of multiple skin lesions (but not spread of single skin lesion), location on the leg, round cell morphology and disease duration.^[11,12] Many studies have supported the contention of the EORTC cutaneous lymphoma working groups in implicating the location on the leg as the most significant factor in predicting adverse prognosis in case of primary cutaneous DLBCL and hence has been classified as a separate entity.^[8,13] It has intermediate clinical behavior and 5 year survival rate is 41%.^[14]

Recent reviews suggest that PCLBCL-LT should be treated as systemic DLBCL with multiagent chemotherapy with or without rituximab.^[14] Nowadays R-CHOP with or without IFRT is considered as first line of treatment in PCLBCL-LT but the efficacy of this approach is still poorly documented.^[15]

Primary cutaneous diffuse large B-cell lymphoma-leg type has a typical clinicopathological features and has aggressive behavior. Surgical excision is highly effective in other cutaneous lymphomas, unlike this type.^[16,17,18] Hence it is important to be aware of this condition so that any suspicious lesions in the leg and other sites should be biopsied for early diagnosis and timely management of the patient.

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