A 78-year-old man with primary cutaneous large B-cell lymphoma-leg type: a case report and literature review

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Primary cutaneous diffuse large B-cell lymphoma-leg type (PCDLBCL-LT) is a rare malignant disease seen in older adults, especially women. In this case report, we discuss a 78-year-old man who developed erythematous indurated plaques on his left shin for about three months. The patient did not report pruritus, weight loss, night sweats, fever, or chills. There was no lymphadenopathy, splenomegaly, or hepatomegaly on the physical examination. A local tissue biopsy was taken from the plaques, confirming the diagnosis of PCDLBCL-LT via immunohistochemistry. The patient was referred to an oncologist to begin additional evaluation and treatment. According to the literature, chemotherapy with or without adjuvant radiotherapy is the first treatment choice for PCDLBCL-LT. Monotherapy with rituximab could be considered in some patients with this condition, but the disease may relapse in a short period.

Keywords: B-cell lymphoma, malignancy, skin disease

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INTRODUCTION

Primary cutaneous diffuse large B-cell lymphoma is a rare extranodal lymphoproliferative dermatology condition that rarely disseminates systematically. The subtypes of this condition are cutaneous B-cell lymphoma, primary cutaneous diffuse large B-cell lymphoma-leg type (PCDLBCL-LT), primary cutaneous follicle center lymphoma (PCFCL), primary cutaneous marginal zone lymphoma (PCMZL), and primary cutaneous diffuse large B-cell lymphoma-other (PCDLBCL-other); among these, PCDLBCL-LT is the most rare subtype [1,2]. These conditions are more common in women than men, with a median age of 76. The nature of PCDLBCL-LT is aggressive, and the prognosis is poor [3,4]. Patients with PCDLBCL-LT typically present with rapidly red to purple nodules or tumors that affect the lower extremities. The disease diagnosis depends on a local skin biopsy and immunohistochemical analysis. The histopathologic findings of PCDLBCL-LT include confluent sheets of large cells with round nuclei, with strong expression of Bcl-2. Furthermore, staining for the MUM-1

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protein is mostly positive [5–7].

To our knowledge, PCDLBCL-LT has rarely been reported worldwide, especially in Iran. This manuscript describes an older adult with PCDLBCL-LT referring to the Dermatology Clinic of Shahid Faghihi Hospital, Shiraz, Iran, as the main center for dermatology conditions in south Iran.

CASE PRESENTATION

A 78-year-old man was referred to the Dermatology Clinic of Shahid Faghihi Hospital, Shiraz, Southern Iran, with erythematous indurated plaques on his left shin for about three months. The initial lesions were asymptomatic red papules on the anterior side of his left lower extremity that rapidly expanded. The patient reported no weight loss, pruritus, night sweats, fever, or chills. Erythematous focally indurated infiltrative plaques with irregular borders were found on the anterior side of the left leg during the patient's physical examination (Figure 1). Moreover, the physical examination showed no palpable superficial lymph nodes, liver or spleen enlargement, or organ abnormalities. A local skin biopsy was taken from the lesion. Histological findings revealed a non-germinal central type lymphoma, most probably primary cutaneous diffuse large B-cell lymphoma-leg type (PCDLBCL-LT). The diagnosis was confirmed by immunohistochemistry (IHC), and the patient was referred to an oncologist for additional evaluation and treatment.

Ethical consideration

The patient signed an informed consent form allowing us to anonymously publish his history, physical examination, laboratory and histopathology findings.

DISCUSSION

This case report discusses an older male adult with PCDLBCL-LT—a rare dermatologic condition worldwide. This disease explains up to 2.6% of primary cutaneous non-Hodgkin lymphomas, predominantly affecting elderly women [5]. The etiology of PCDLBCL-LT is unknown, but evidence claims that a lymphoproliferative response to antigenic stimuli in the cutis might have a main role in disease development [8]. Human Herpes Virus-6 (HHV-6), Human Herpes Virus (HHV-8) [9], Epstein–Barr virus [10], *Borrelia burgdorferi* infection [8], and systemic methotrexate are considered as inciting factors [11].

In PCDLBCL-LT, the distal part of the lower extremities is most commonly affected. It may be unilateral or bilateral. This condition is expressed by the rapid growth of red or purple-red plaques, nodules, or tumors, which are sometimes ulcerative [3,4,6]. They also can spread to extracutaneous organs such as the central nervous system, bones, kidney, liver, spleen, pancreas, testicles, breasts, pelvic organs, and the brachial plexus nerves [12–15].

Patients with PCDLBCL-LT commonly have no symptoms except mild pruritus. Pathological and immunohistochemical tests are required to diagnose this disease. There is a monotonous population of large B cells with round nuclei, prominent nucleoli, and coarse chromatin in diffuse non-epidermotropic infiltrates that resemble centroblasts or immunoblasts, usually extending into the dermis and subcutaneous tissues and separated from the epidermis by a Grenz zone [1,16,17]. PCDLBCL-LT is determined by the positivity of MUM-1, CD20, and high Bcl-2 expression [3,18]. They often express other B-cell markers (CD19, CD22, CD79a), with a lack of CD5,

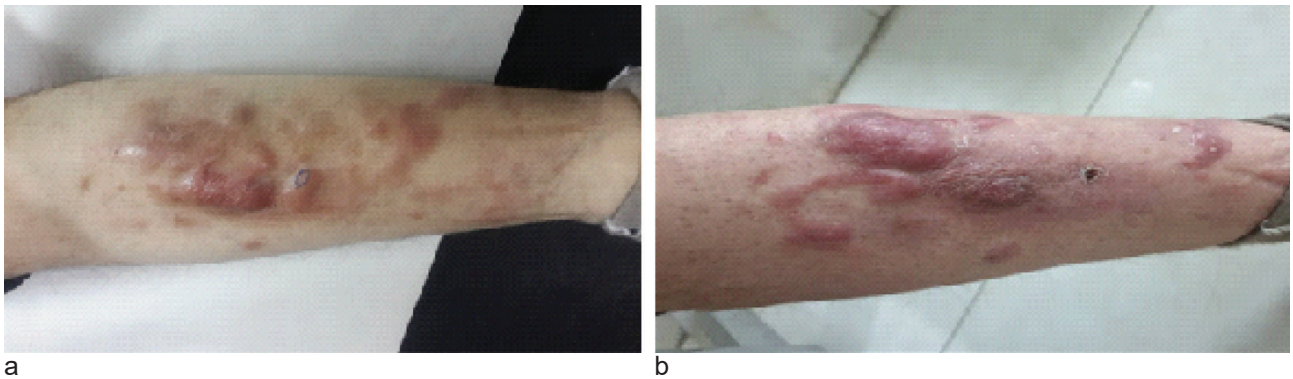

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Figure 1. Macroscopic view of the lesions of a 78-year-old man with primary cutaneous diffuse large B-cell lymphoma-leg type (PCDLBCL-LT): (a) erythematous indurated plaques on the anterior aspect of the left shin; (b) eighteen days later.

CD10, and t(14;18) (q32;q21) translocation [19]. Genetic analysis has found that NF-κB pathway-activating mutations (including CD79B, CARD11) are seen in PCDLBCL-LT. Moreover, the MYD88 mutation is the most prevalent mutation in this disease [4,18,20]. A number of poor prognostic factors contribute to a poor prognosis, including old age, multiple lesions, MYD88 mutations, advanced T stages, and leg location [2,6,7]. This condition rarely remits spontaneously [21].

The first-choice treatment for PCDLBCL-LT is chemotherapy (doxorubicin, vincristine, cyclophosphamide, prednisone, and rituximab) with or without radiotherapy [22–24]. In the case of a solitary lesion, excision or radiotherapy should be considered [24]. Additionally, monotherapy with rituximab is possible, but a relapse may occur quickly [10].

CONCLUSION

Although PCDLBCL-LT is a rare malignant dermatologic condition in men and may not be considered in the first-line differential diagnoses of dermatologists, further evaluations should not be neglected for those who come with a chief complaint of erythematous indurated plaques in their legs.

Authors contributions

Substantial contributions to the conception, design of the work, acquisition, and interpretation of data for the work: NS, MBS, FSA, MMP; Drafting the work: MBS, MMP; Revising it critically for important intellectual content: NS, FSA, MMP; Final approval of the version to be published: NS, MBS, FSA, MMP; All authors agreed to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved.

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