An unusual presentation of diffuse large B-cell lymphoma: a case report of an ulcerative inguinal mass

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B-cell lymphomas represent most non-Hodgkin lymphomas (NHLs) arising within lymph nodes, and about 27% of patients have extranodal involvement. Primary cutaneous lymphoma is defined as malignant lymphoma limited to the skin at diagnosis. Diffuse large B-cell lymphoma (DLBCL) is the most common form of NHL, accounting for over one-third of all lymphomas. Primary cutaneous diffuse large B-cell lymphoma (PCDLBCL) is a type of non-Hodgkin's lymphoma with skin involvement as the first and only site of involvement. Primary cutaneous diffuse large B-cell lymphoma typically presents as a rapid-growing, red or bluish nodule or tumor on the legs, though around 10–15% of patients present with lesions elsewhere. This case report illustrates a rare manifestation of PCDLBCL presenting as a non-healing, rapidly progressive ulcer in the groin area diagnosed based on histopathology and immunohistochemical expression. The patient was treated successfully with systemic chemotherapy. This report could have implications for clinicians to consider the diagnosis of PCDLBCL in patients with unusual, non-healing, chronic ulcers, especially in the elderly, despite the anatomic site of the lesions.

Keywords: diffuse large B-cell lymphoma, lymphoma, ulcer, non-Hodgkin lymphoma, skin neoplasms

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INTRODUCTION

B-cell lymphomas represent the majority of non-Hodgkin lymphomas (NHLs) arising within lymph nodes. Approximately 27% NHLs have extranodal involvement, with the gastrointestinal tract and skin (18%) being the most prevalent sites [1]. Diffuse large B-cell lymphoma (DLBCL) is the most common form of NHL, accounting for over one-third of all lymphomas.

Primary cutaneous lymphoma is defined as malignant lymphoma limited to the skin at presentation after complete staging procedures. The main clinical subtypes include primary cutaneous follicle-center lymphoma (PCFCL), primary cutaneous diffuse large B-cell lymphoma, leg type (PCDLBCL-LT), and primary cutaneous marginal zone lymphoma (PCMZL) [2].

PCDLBCL usually manifests as papules, nodules, or deeply infiltrated plaques, but some unusual variants presenting as a chronic skin ulcer, verrucous plaque, multiple nodules, and a bluish-reddish multicolored pattern have also been described [3].

Independent of the site of involvement, any primary cutaneous large B-cell lymphoma (PCBCL) showing both BCL-2 (B-cell lymphoma 2) and MUM-1(multiple myeloma oncogene 1) expression

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is considered as PCDLBCL-LT. While the most common site of involvement is the distal aspect of one leg, the lesions are localized to other sites in 10 to 15% of patients [4].

This case report illustrates a rare manifestation of PCDLBCL presenting as a non-healing, rapidly progressive ulcer in the groin area.

CASE PRESENTATION

A 60-year-old woman presented to the Loghman Hakim Hospital with a new-onset, rapidly progressive ulcer in the right groin area that appeared three months earlier. The lesion first appeared as a small subcutaneous papule, enlarging over time to a large ulcerative mass with multiple purulent fistulas.

Physical examination revealed an ulcerated tumoral mass in the right inguinal fold measuring approximately 20 cm in diameter. The mass was erythematous, non-tender, and slightly warm, with multiple fistulas draining purulent discharge but without odor (Figure 1). She had previously received various antibiotics for this lesion without an adequate therapeutic effect. During these three months, she had 5 kg weight loss and generalized weakness, but she had no complaints of B symptoms. She had a past medical history of hypertension controlled with losartan and amlodipine for the last five years.

Laboratory assessments including complete blood count, serum creatinine, serum electrolytes, ferritin, lactate dehydrogenase (LDH), erythrocyte sediment ratio (ESR), liver function tests, urine analysis, blood culture, hepatitis markers, HIV (human immunodeficiency virus) antibody, venereal disease research laboratory test (VDRL), and rapid plasma regain (RPR) were normal.

To exclude infectious etiologies, especially sexually transmitted diseases presenting as an inguinal mass, a donovanosis smear and culture from the wound were performed. After obtaining negative results, an incisional biopsy was taken from the lesion.

Histological examination and hematoxylin and eosin staining demonstrated a tumoral tissue with diffuse infiltration throughout the dermis and neoplastic large lymphoid cells with round nuclei, prominent nucleoli, and high mitotic activity (Figure 2 a and b). Immunohistochemical (IHC) staining was positive for CD20, CD 79a, BCL-2, MUM-1, and Ki 67,

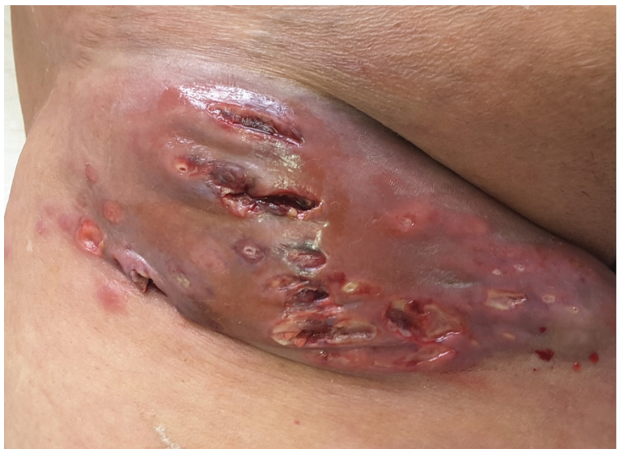
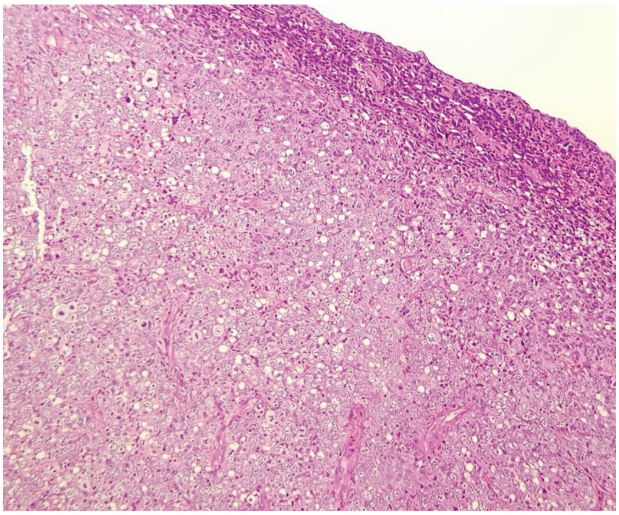
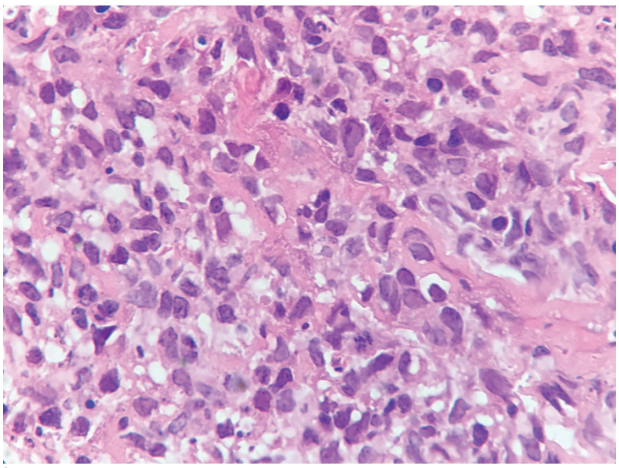


Figure 1. An ulcerated tumoral mass in the inguinal area.



a



b

Figure 2. Histological examination demonstrates a tumoral tissue with diffuse infiltration throughout the dermis and neoplastic large lymphoid cells with round nuclei, prominent nucleoli, and high mitotic activity (H&E stain, (a) 100×, (b) 1000×).

showing proliferative activity in 75% of tumoral cells in the hot spot area (Figure 3). According to these findings, the diagnosis of diffuse large B-cell lymphoma was made. Adequate staging procedures required to differentiate between PCBCL and systemic lymphomas secondarily involving the skin including bone marrow biopsy and computed tomography (CT) scan of the head, neck, chest, abdomen, and pelvis were performed, and no systemic involvement was detected. Accordingly, the diagnosis of PCBCL was confirmed.

The cutaneous lesion was treated by total excision with free histopathological margins. The patient was referred to an oncologist for chemotherapy. She underwent eight sessions of chemotherapy with the rituximab, cyclophosphamide, doxorubicin, vincristine, and prednisone regimen (R-CHOP), resulting in complete remission of the lesion without any relapse during a two-year follow-up.

Ethical considerations

Informed consent was obtained from the patient.

DISCUSSION

The prevalence of cutaneous lymphomas is 0.3 cases per 100,000 populations per year, of which 10–20% are cutaneous B-cell lymphomas [5]. PDLBCL is a type of non-Hodgkin's lymphoma with skin as the first and only site of involvement. This condition predominantly affects elderly patients (mean age:

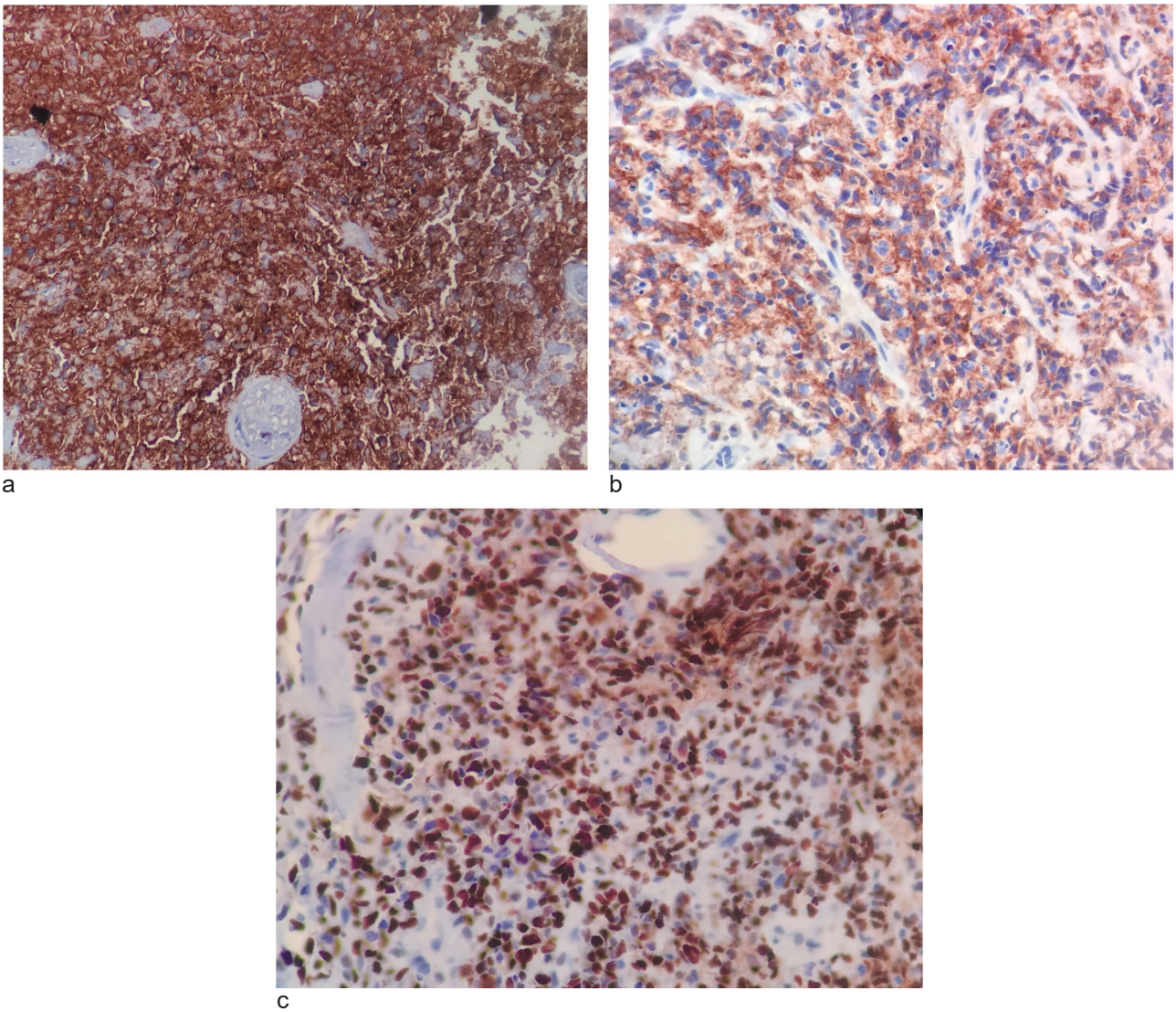


Figure 3. Immunohistochemical studies demonstrate that the cells were CD 20+ (a) and expressed BCL-2 (b) and MUM1 (c) (100×).

70.6) and is more frequent in females [2].

PDLBCL typically presents as a rapid-growing, red or bluish nodule or tumor on the legs; however, around 10–15% of patients present with lesions elsewhere. The differential diagnosis of a rapidly growing ulcerative mass in the groin area is very wide and includes donovanosis, syphilis, other STDs, capillary and lymphatic tumors or malformations, pyoderma gangrenosum, and most importantly, neoplastic etiologies like squamous cell carcinoma (SCC), melanoma, metastasis, and lymphomas [6,7].

Although PDLBCL manifesting as an inguinal ulcerative mass is a very rare presentation, it is important to keep it in mind due to its grave prognosis and the importance of early detection. The prognosis of these lymphomas is less favorable than that of other types of PCBCL, with an estimated 5-year survival rate of 40–50% [3].

The diagnosis is based on histological examination and IHC; characteristic histologic findings include diffuse infiltrative proliferation patterns predominantly consisting of large non-cleaved B-cells, with variable amounts of centroblast and immunoblast-like cells. The cancer cells are CD20+, CD79a+, BCL2+, FOX-P1+, MUM-1/IRF-4+ and CD10–.

After confirming the diagnosis, treatment needs to be initiated urgently. The recommended first-line therapeutic approach is R-CHOP (rituximab-cyclophosphamide, doxorubicin, vincristine, prednisone), sometimes along with adjuvant radiation therapy. More recently, targeted therapies such as bortezomib and ibrutinib have shown promising results [4].

CONCLUSION

Although PCDLBCL leg type usually presents as erythematous nodules on the distal lower extremities, it can have various forms of manifestations, such as this rare form presented in this report: an ulcerated mass in the inguinal fold. As the incidence of this disease is increasing with the aging of the population [8], clinicians must keep in mind its unusual presentations, leading to careful evaluation and proper investigations of the patient. A prompt, accurate diagnosis ensures the early initiation of treatment, which leads to a better prognosis. Hence, clinicians must consider the diagnosis of PCDLBCL in patients with unusual, non-healing, chronic ulcers (especially in the elderly), regardless of the anatomic site of the lesions.

Patient consent

Consent was obtained from the patient to publish clinical data and images in this case report.

Authors' Contribution

All authors contributed equally to this study. Soheila Nasiri and Niloufar Najar Nobari performed the research. Shirin Zaresharifi and Nooshin Zaresharifi wrote the paper. Soheila Nasiri revised the manuscript and supervised it. All authors read and approved the final manuscript.

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