WJPMR, 2025, 11(11), 268-270

Case Study

WORLD JOURNAL OF PHARMACEUTICAL AND MEDICAL RESEARCH

www.wjpmr.com

Impact Factor: 6.842

ISSN (O): 2455-3301

ISSN (P): 3051-2557

Coden USA: WJPMBB

WHEN THE SKIN HIDES A LYMPHOMA: LONG-TERM REMISSION OF A PRIMARY CUTANEOUS FOLLICLE CENTRE LYMPHOMA

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DOI: https://doi.org/10.5281/zenodo.17538876

How to cite this Article: Lbarrah Fatima Zahra*, Aghoutane Yossra, Benlachgar Naoufal, Haidouri Soukaina, Tazi Mezalek Zoubida. (2025). WHEN THE SKIN HIDES A LYMPHOMA: LONG-TERM REMISSION OF A PRIMARY CUTANEOUS FOLLICLE CENTRE LYMPHOMA. World Journal of Pharmaceutical and Medical Research, 11(11), 268–270.

Article Received on 07/10/2025 **Article Revised on 27/10/2025** **Article Published on 01/11/2025**

ABSTRACT

Background: Primary cutaneous follicle centre lymphoma (PCFCL) is an indolent subtype of B-cell lymphoma confined to the skin, without evidence of systemic involvement. It typically follows a benign course and responds well to local or systemic therapy. **Case presentation:** We report the case of a 57-year-old man who presented with a one-year history of vesicular plaques on the chest, which evolved into a large ulcerated lesion. Physical examination revealed a single ulcer on the trunk, without lymphadenopathy or systemic symptoms. Laboratory findings, including complete blood count and lactate dehydrogenase levels, were within normal limits. Serological tests for hepatitis B and C, HIV, and syphilis were negative. Histopathological examination of two skin biopsies demonstrated a diffuse and follicular proliferation of large lymphoid cells. Immunohistochemistry revealed positivity for CD20 and BCL6, and negativity for CD10, BCL2, MUM1, CD5, and EBV, consistent with the diagnosis of PCFCL. A CT scan showed extension to the anterior mediastinum, without other secondary localizations. The patient was treated with R-CHOP (rituximab, cyclophosphamide, doxorubicin, vincristine, prednisone), achieving complete remission. After three years of follow-up, the patient remains disease-free. **Conclusion:** This case illustrates the favourable prognosis of primary cutaneous follicle centre lymphoma, even in cases with mediastinal extension. R-CHOP therapy can lead to durable remission in patients with extensive disease.

KEYWORDS: Primary cutaneous follicle centre lymphoma, B-cell lymphoma, R-CHOP, chest wall, long-term remission.

INTRODUCTION

Primary cutaneous follicle centre lymphoma (PCFCL) is a rare, indolent B-cell lymphoma characterized by the proliferation of follicle centre cells confined to the skin. It accounts for approximately 10–20% of all primary cutaneous lymphomas and typically affects middle- aged or elderly patients. Clinically, PCFCL manifests as solitary or grouped plaques, nodules, or tumours, most often on the head, neck, or trunk. Unlike systemic follicular lymphoma, PCFCL has an excellent prognosis, with a 5-year survival rate exceeding 95%. Nevertheless, unusual presentations, such as ulceration or deep tissue extension, may raise diagnostic and therapeutic challenges. Here, we report a case of ulcerated PCFCL of the chest wall with anterior mediastinal extension, successfully treated with R-CHOP, and discuss its clinicopathological characteristics and management.

CASE PRESENTATION

A 57-year-old man presented with a one-year history of vesicular plaques on the anterior chest, which gradually evolved into a large ulcerated lesion. On admission, physical examination revealed a solitary ulcer on the trunk without palpable lymphadenopathy, hepatomegaly, or splenomegaly. The patient denied fever, night sweats, or weight loss.

Routine laboratory tests, including complete blood count, renal and hepatic panels, and LDH levels, were within normal limits. Serologic screening for hepatitis B and C viruses, HIV, and syphilis (VDRL, TPHA) was negative.

Histopathologic examination of two skin biopsies showed a diffuse and follicular proliferation of large lymphoid cells within the dermis. Immunohistochemistry demonstrated that the neoplastic cells were positive for

CD20 and BCL6, but negative for CD10, BCL2, MUM1, CD5, and EBV, consistent with primary cutaneous follicle centre lymphoma.

A CT scan revealed infiltration of the anterior chest wall with extension to the anterior mediastinum, without evidence of nodal or visceral involvement elsewhere.

The patient was treated with six cycles of R-CHOP (rituximab 375 mg/m², cyclophosphamide 750 mg/m², doxorubicin 50 mg/m², vincristine 1.4 mg/m², prednisone 100 mg for 5 days). Treatment was well tolerated, and the lesion completely regressed.

Radiologic evaluation confirmed complete remission at 18 months. After three years of follow-up, the patient remains in good clinical condition, with no signs of recurrence.

Figures 1 illustrate the clinical evolution of the lesions before, during, and after treatment, highlighting the favorable and sustained response to R-CHOP therapy.

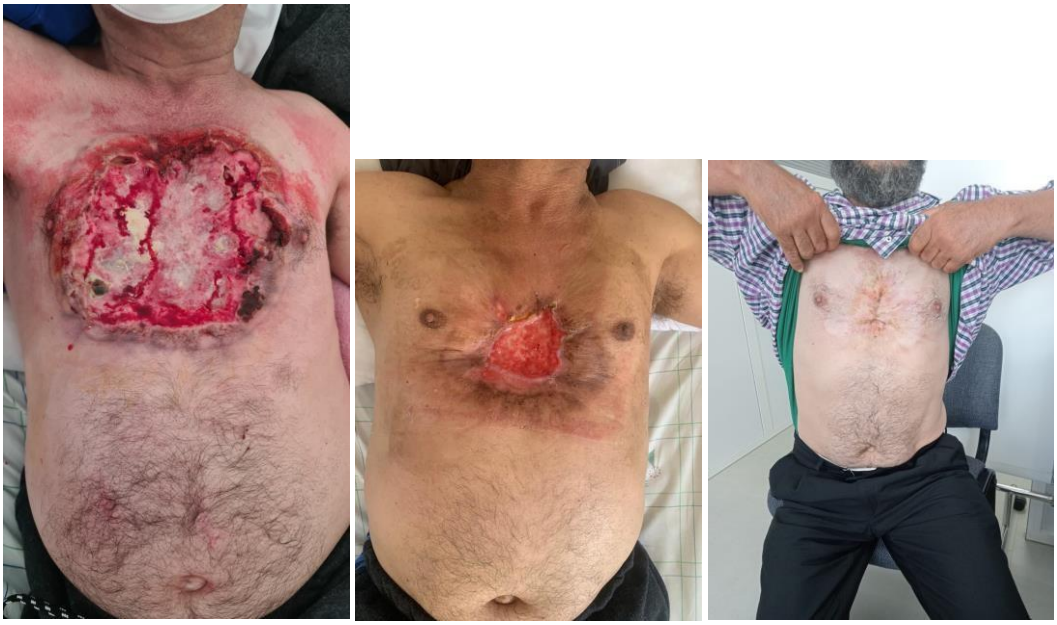

Figure 1: (A) Pre-treatment (ulcerated lesions). (B) During treatment. (C) Post-treatment (complete remission).

DISCUSSION

PCFCL represents a distinct clinical and pathological entity within the group of primary cutaneous B-cell lymphomas. It predominantly affects middle-aged men and typically presents with localized skin lesions on the scalp, forehead, or trunk. The disease is indolent and rarely disseminates to extracutaneous sites.

Histologically, PCFCL shows a follicular, diffuse, or mixed growth pattern composed of centrocytes and centroblasts. Immunophenotyping is essential for diagnosis: PCFCL cells usually express CD20, CD79a, and BCL6, and may express CD10. They typically lack BCL2 expression, which distinguishes PCFCL from systemic follicular lymphoma with secondary skin involvement.

Our patient's case was unusual due to the ulcerated presentation and mediastinal extension, features that are rarely observed in PCFCL. Despite this, the immunohistochemical profile supported a primary cutaneous origin. The absence of BCL2 and CD10 expression, together with the absence of systemic involvement on imaging, reinforced the diagnosis.

Therapeutic options depend on the disease extent. Localized PCFCL can often be managed with radiotherapy alone. However, for multifocal lesions or cases with extracutaneous extension, systemic therapy is indicated. Rituximab monotherapy or combination chemotherapy with R-CHOP has been associated with excellent outcomes. Our patient achieved complete remission after six cycles of R-CHOP, with a durable remission at three years, consistent with the favourable long-term prognosis described in the literature.

CONCLUSION

Primary cutaneous follicle centre lymphoma is a rare, indolent B-cell lymphoma with an excellent prognosis. Accurate histopathological and immunohistochemical evaluation is crucial to differentiate it from systemic follicular lymphoma. Even in cases with unusual presentations or local extension, systemic therapy with R-CHOP can achieve long-term remission.

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