CASE REPORT

Investigating a fast-growing nodule on the face: A case of primary cutaneous marginal zone lymphoma

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Abstract

Primary cutaneous B-cell lymphomas are non-Hodgkin lymphomas that present in the skin without evidence of extracutaneous involvement at diagnosis. There are three types of primary cutaneous B-cell lymphomas: primary cutaneous marginal zone lymphoma, primary cutaneous follicle center lymphoma, and primary cutaneous diffuse large B-cell lymphoma, leg-type.

Keywords: B-cell Lymphoma, Marginal Zone Lymphoma, Nodule

Introduction

Primary Cutaneous B-cell lymphoma comprises of three subtypes: Marginal Zone Lymphoma (MZL), Follicle Centre Cell Lymphoma (FCL) and Diffuse Large B-Cell Lymphoma (LBCL). Full staging investigations in patients with cutaneous B-cell lymphoma are essential to exclude the possibility of secondary cutaneous involvement with a nodal lymphoma.

Case Report

A 44-year-old female presented with a nodule on her right cheek since the past 20 days. It seemed to have progressively increased in size. There was no history of pain, trauma, insect bite or discharge from the lesion, and no present or past history of similar lesions anywhere else in the body. (Figure 1a) Differentials considered were granuloma faciale, insect bite hypersensitivity, pseudolymphoma, Hansens disease, Jessner's lymphocytic infiltrate and lymphoma. We subjected the patient to a skin biopsy and histopathological examination revealed a diffuse lymphocytic infiltrate throughout the dermis and subcutaneous tissue (Figure 2). Immunohistochemistry (IHC) markers such as CD 3 and CD 20 were found to be positive. CD 20 was positive in background of B cells. CD 3 was positive in atypical cells. IHC markers such as BCL 2, BCL 6, CD 10 were done and it was strongly positive for BCL 2, scattered positive for BCL 6 and negative for CD 10 (Figure 3). The patient was started on a course of 100 mg oral Thalidomide daily for a week and an intralesional steroid injection (Injection Triamcinolone 10 mg/ml) was given after that. The lesion appeared to flatten and improve and oral Thalidomide was continued (Figure 1b).

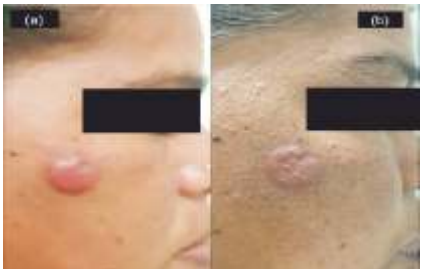

Figure 1a: Erythematous nodule on right cheek
Figure 1b: Flattening of the lesion post intralesional steroid injection

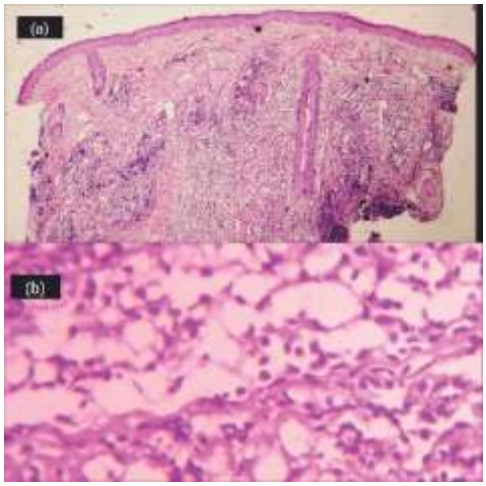

Figure 2a: Epidermal atrophy with the presence of inflammatory infiltrate (H&E, 10×)

Figure 2b: Inflammatory infiltrate consisting predominantly of lymphocytes (H&E, 40×)

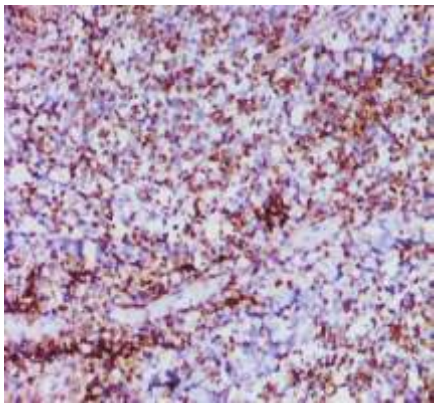

Figure 3: Strong positive BCL2 (Immuno-histochemistry)

Discussion

Primary Cutaneous B Cell Lymphomas (pCBCL) are a type of lymphoproliferative disorders characterized by a clonal proliferation of B-cells, primarily involving the skin [1]. They form only a minority of the non-Hodgkin lymphomas whose primary site is the skin. They can be classified as: Primary Cutaneous Marginal Zone B-Cell Lymphoma (PCMZL), Primary Cutaneous Follicle Centre Lymphoma (PCFCL), Primary Cutaneous Diffuse Large B-Cell Lymphoma (PCDLBCL), Leg Type (LT) and primary cutaneous diffuse large B-cell lymphoma and other [2]. The aetiology is unknown. Serologic evidence of prior infection with *Borrelia burgdorferi*, *Helicobacter pylori*, and *Epstein-Barr* virus has been found in some cases, especially in endemic areas. Presently, no other microorganism has been convincingly linked to the development of pCBCL. However, it should be noted that pCBCLs have been observed in patients with AIDS, and reversible pCBCLs have been observed in patients undergoing therapy with methotrexate, thus suggesting that immune dysfunction may play a role in the development of this disease. It affects both adult males and females, with a slight predominance in males. In pCBCL, low grade tumours are more common than high grade tumours and among the lowgrade tumours, marginal zone and follicular centre tumours are common which are characterised by an indolent clinical behaviour.

Marginal Zone Lymphoma

PCMZL is a low-grade malignant B-cell lymphoma that presents in the skin, with no evidence of extracutaneous localizations at the time of diagnosis. Previously, these lymphomas were referred to as primary cutaneous immunocytomas, but in recent years, the term primary cutaneous marginal zone B-cell lymphoma has been preferred. Clinically, patients present with recurrent pink–violet to red–brown papules, plaques and nodules localized preferentially on the extremities (upper more so than lower) or trunk. A small number of patients can show generalised lesions.

Skin lesions are usually asymptomatic and ulceration may rarely occur. Systemic signs and symptoms, such as fever, night sweats, weight loss and malaise are not present. The serum level of LDH is within normal limits. In certain cases, resolution of lesions may be accompanied by secondary anetoderma due to loss of elastic fibres in the area of the tumour infiltrate. The prognosis of PCMZL is excellent. The small neoplastic B cells have a Bcl-2+, Bcl-6−, CD10− phenotype, which facilitates differentiation from primary cutaneous follicle center lymphomas and cutaneous lymphoid hyperplasias (pseudolymphomas) [3].

Radiotherapy or surgical excision, topical drugs, intralesional therapies, immunochemotherapy have been found to be useful. Thalidomide, in particular, may act directly on tumour cells to induce apoptosis or cell cycle arrest, or indirectly, by inhibiting angiogenesis, altering immune cell cytokine secretion, enhancing T cell, natural killer cell, and dendritic cell activity, and inhibiting NF-κB activity [4].

Conclusion

This case of primary cutaneous marginal zone B-cell lymphoma has been reported in view of its rarity and acute presentation, and to serve as a reminder that immunohistochemistry plays a role in confirming a diagnosis of cutaneous lymphoma as it can mimic a number of other dermatological conditions both clinically and on histopathology. Primary cutaneous B-cell lymphoma being an indolent entity, entails the use of intralesional steroids as therapy which is highly beneficial, as evidenced in our case.

References

1. Selva R, Violetti SA, Delfino C, Grandi V, Cicchelli S, Tomasini C, *et al*. A literature revision in primary cutaneous B-cell lymphoma. *Indian J Dermatol* 2017;62(2):146-157.
2. Gupta P, Agarwal P, Ahuja A, Durga CK. Primary cutaneous non-Hodgkin's lymphoma, clinically mimicking a soft tissue sarcoma. *CytoJournal* 2018;15:2.
3. Hoefnagel JJ,Vermeer MH, Jansen PM, Flueren GJ, Meijer CJLM, Willemze R. Bcl-2, Bcl-6 and CD10 expression in cutaneous B-cell lymphoma: further support for a follicle center cell origin and differential diagnostic significance. *Br J Dermatol* 2003;149(6): 1183- 1191
4. Ji D, Li Q, Cao J, Guo Y, Lv F, Liu X,*et al*. Thalidomide enhanced the efficacy of CHOP chemotherapy in the treatment of diffuse large B cell lymphoma: A phase II study. *Oncotarget* 2016;7(22):33331.

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