

Cutaneous follicular B-cell lymphoma presenting as a solitary erythematous papule on the nose

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Abstract Primary cutaneous follicle center lymphoma is an uncommon type of cutaneous lymphoma. Typically presenting as raised erythematous lesions on the head or trunk, it can clinically mimic many other conditions. When cutaneous lymphoma is identified, the presence of systemic follicular lymphoma must also be considered. We present and discuss a case of primary cutaneous follicle center lymphoma in a patient with a solitary erythematous papule on the nose.

Key words

Cutaneous lymphoma; lymphoma.

Introduction

Primary cutaneous lymphomas are a subset of non-Hodgkin lymphomas that are localized to the skin. While the vast majority of systemic non-Hodgkin lymphomas are caused by an abnormal proliferation of B-cells, only about 25% of primary cutaneous lymphomas originate from B-cells.¹ The prevalence of primary cutaneous B-cell lymphoma (PCBCL) has been estimated to be 4 per 1,000,000.¹ Incidence is highest among males, non-Hispanic whites, and in individuals greater than 50 years of age, but PCBCL can affect people of any age or gender. PCBCL may be classified into three major subtypes: primary cutaneous follicle center lymphoma (PCFCL), primary cutaneous marginal zone lymphoma, and primary cutaneous diffuse large B-cell lymphoma, leg type. PCFCL is the most common subtype of cutaneous B-cell lymphoma.² It presents as a solitary lesion or a group of localized

erythematous papules, plaques, or nodules arising most commonly on the trunk, face, and scalp.³ The lesions are typically asymptomatic, but may ulcerate, systemic symptoms rarely occur. Recurrence of the lesions is commonly seen but systemic spread to lymph nodes or internal organs is rare. We present an unusual case of primary cutaneous follicle center lymphoma manifesting as a solitary papule on the nose.

Case report

A 75-year-old gentleman presented to the dermatology clinic with a red lesion on his nasal tip. He complained of redness and persistence of the lesion for the past 6 months. He endorsed occasional irritation from the lesion but denied any itching, bleeding, or ulceration. He reported no personal or family history of skin cancer. Physical exam revealed a 1cm erythematous papule on the nasal tip. Dermoscopy showed a pearly papule with arborizing vessels. The rest of his full body examination was only notable for seborrheic keratoses. A shave biopsy of the nasal lesion was taken and sent for histological examination. Hematoxylin and eosin staining showed a dense proliferation of small- and

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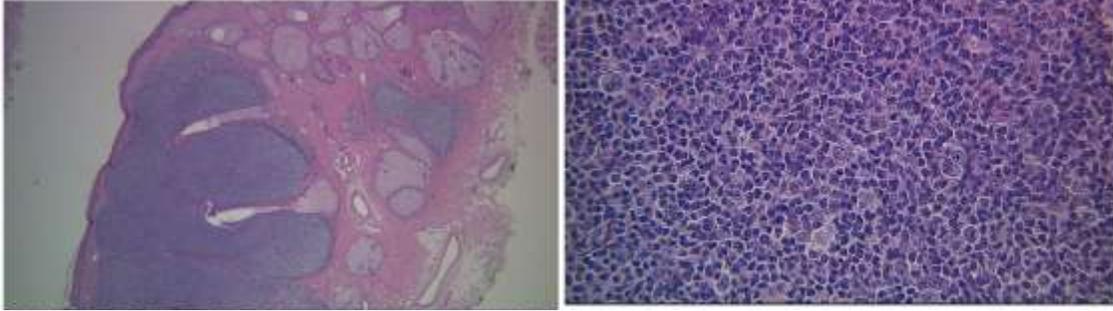


Figure 1 Initial shave biopsy demonstrated a dense proliferation of small- and medium-sized lymphoid cells with scattered plasma cells (H&E, original magnifications x10, x100).

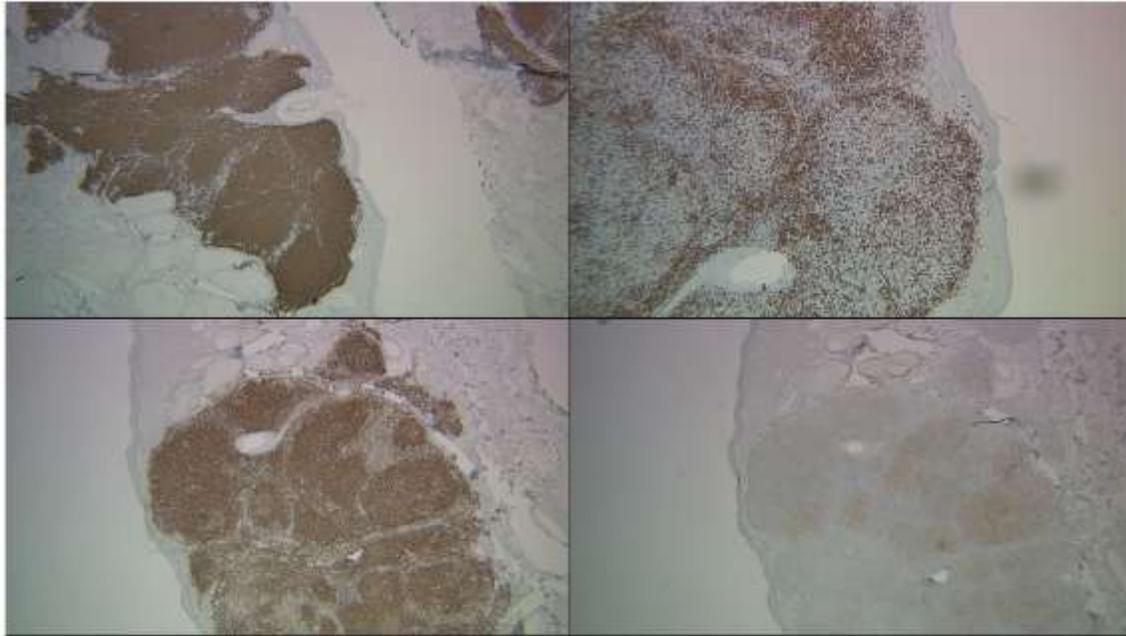


Figure 2 The lymphoid infiltrate expressed CD20 (upper left) with a subset of cells staining CD3 (upper right). There was a strong expression of Bcl-2 throughout this proliferation. There was also expression of Bcl-6. Expression of Bcl-2 (lower left) and Bcl-6 (lower right) were concerning features for follicle center lymphoma (IHC, original magnifications x10).

medium- sized lymphoid cells with scattered plasma cells (**Figure 1**). The lymphoid proliferation had a strong expression of Bcl-2 and Bcl-6 raising concern for cutaneous lymphoma. The patient was referred to surgical oncology and the lesion was excised. Pathologic examination confirmed the diagnosis. Immunohistochemical stains revealed the phenotype of the cells to be CD20+, CD3-, CD10+, Bcl-6+, Bcl-2+, MUM1-, CD43-, consistent with cutaneous follicular B-cell lymphoma (**Figure 2**). The patient underwent radical resection of his solitary cutaneous lesion

with 2 cm margins down to cartilage by surgical oncology with subsequent repair by plastic surgery. He also presented with thyroid gland enlargement and was found to have 3 nodules on the right lobe and 1 on the left. The patient is currently undergoing further evaluation.

Discussion

As raised erythematous lesions, PCFCL can clinically resemble many other inflammatory and neoplastic conditions. Thus, more extensive examination is required to adequately treat the

patient. Dermoscopic examination of PCFCL typically reveals white circles with a salmon-colored background with arborizing vessels and scale.⁴ Histopathological examination of PCFCL will show either a follicular or diffuse pattern of lymphocytic infiltration of the dermis, with sparing of the epidermis. Like other forms of B-cell lymphoma, PCFCL will stain positive for CD20, CD79a, and Pax5. PCFCL will also typically be positive for Bcl-6 but negative for Bcl-2, CD5, and CD43.⁵ CD10 is positive in the follicular pattern but usually negative when diffuse lymphocytic infiltration is seen. These markers separate PCFCL from primary cutaneous marginal zone lymphoma which normally are Bcl-2 positive and Bcl-6 negative. Thus, immunohistochemical phenotyping can be extremely useful in differentiating between the variants of cutaneous B-cell lymphomas and determining an appropriate course of treatment. The genetic markers for PCFCL can also provide insight to the presence of extracutaneous disease. As suggested by the lack of staining with Bcl-2, PCFCL typically does not possess the t(14,18) translocation involving the Bcl-2 locus that is classically associated with systemic follicular lymphomas.¹ Despite these patterns, histopathological examination alone cannot determine whether a biopsied sample is from a primary skin lesion or secondary cutaneous manifestation of systemic disease.⁶ Therefore, full body imaging must be performed to rule out the presence of follicular lymphoma elsewhere in the body. In confirmed cases of cutaneous follicle center lymphoma, management begins with tumor staging, including a history, physical exam, and imaging the chest, abdomen, and pelvis.⁶ If systemic involvement is detected, hemato-oncologic management is required as well. For patients with no signs of systemic disease and a solitary cutaneous lesion, surgical excision and low-dose radiation therapy have been utilized with good success. Intralesional corticosteroids or rituximab have also been

used.^{7,8} Radiation or observation are appropriate options for managing patients with multiple, localized lesions. For more extensive cutaneous disease, patients can be effectively treated with systemic rituximab.⁹ PCFCL presenting as a solitary papule on the nose, as seen in our patient, has only been reported twice in the literature.^{2,10} In these documented cases, the lesions were initially thought to be basal cell carcinomas prior to biopsy. Complete surgical excision resulted in resolution of the patients' symptoms and subsequent evaluation found no evidence of systemic lesions.⁴

Conclusion

Despite low incidence of PCFCL, it is important to recognize the clinical and histological characteristics of PCFCL when considering solitary erythematous lesions on the face. Solitary PCFCL carries a good prognosis and is easily treated. However, systemic involvement of disease poses a serious threat and appropriate imaging and staging is needed.

References

1. Wilcox RA. Cutaneous B-cell lymphomas: 2019 update on diagnosis, risk stratification, and management. *Am J Hematol.* 2018;**93**:1427-30.
2. Maruccia M, Di Taranto G, Ruggieri M, *et al.* Unusual presentation of primary cutaneous follicle center B-cell Lymphoma (PC-FCL) of the nose. *J Plast Reconstr Aesthet Surg.* 2017;**70**(9):e19-e21.
3. Hope CB, Pincus LB. Primary Cutaneous B-cell Lymphomas. *Clin Lab Med.* 2017;**37**(3):547-74.
4. Mascolo M, Piccolo V, Argenziano G, *et al.* Dermoscopy Pattern, Histopathology and Immunopheno type of Primary Cutaneous B-Cell Lymphoma Presenting as a Solitary Skin Nodule. *Dermatology.* 2016;**232**(2):203-237.
5. Wang HY, Zu Y. Diagnostic Algorithm of Common Mature B-Cell Lymphomas by Immunohistochemistry. *Arch Pathol Lab Med.* 2017;**141**(9):1236-46.

6. Kim YH, Willemze R, Pimpinelli N, *et al*. TNM classification system for primary cutaneous lymphomas other than mycosis fungoides and Sezary syndrome: a proposal of the International Society for Cutaneous Lymphomas (ISCL) and the cutaneous lymphoma task force of the European Organization of Research and Treatment of cancer (EORTC). *Blood*. 2007;**110**:479-84.
7. Penate Y, Hernandez-Machin B, Perez-Mendez LI, *et al*. Intralesional rituximab in the treatment of indolent primary cutaneous B-cell lymphomas: an epidemiological observational multicentre study. The Spanish Working Group on Cutaneous Lymphoma. *Br J Dermatol*. 2012;**167**:174-9.
8. Perry A, Vincent BJ, Parker SR. Intralesional corticosteroid therapy for primary cutaneous B-cell lymphoma. *Br J Dermatol*. 2010;**163**:223-5.
9. Senff NJ, Noordijk EM, Kim YH, *et al*. European Organization for Research and Treatment of Cancer and International Society for Cutaneous Lymphoma consensus recommendations for the management of cutaneous B-cell lymphomas. *Blood*. 2008;**112**:1600-9.
10. Wanczinsnki MI, Pinto CA, Trevisan F, *et al*. Primary cutaneous follicle center lymphoma simulating basal-cell carcinoma on the nasal ala. *An Bras Dermatol*. 2015;**90(3)**:111-4.