Primary cutaneous B-cell lymphoma located on the face and hard palate

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ABSTRACT

Introduction: Primary cutaneous B-cell lymphomas represent primary cutaneous lymphomas. The World Health Organization-European Organization for Research and Treatment of Cancer defines primary cutaneous B-cell limphomas as malignant lymphomas that are confined to the skin at presentation after complete staging procedures.

Case report: We report a case of a woman with a cutaneous nodular B-cell lymphoma of the face, with very slow evolution that could easily be misdiagnosed. The patient had two non-ulcerated nodules on the left part of the face. A histological examination on both pieces following surgical excision showed a diffuse infiltrate of atypical B cells. The patient was subsequently directed to the Oncology Department for further investigation, but she died 24 hours afterwards due to one of the acute possible complications of the disease: pulmonary embolism.

Conclusion: Cutaneous B-cell lymphoma is an unusual and rare skin neoplasm with a great range of clinical presentations and this is an alarm sign for apparent common skin lesion.

KEY WORDS: B-cell lymphoma, non-Hodgkin lymphoma, face and hard palate lymphoma

INTRODUCTION

Primary cutaneous T-cell and B-cell lymphomas represent primary cutaneous lymphomas. PCL are rare forms (2%) of non-Hodgkin's lymphomas with an annual incidence of 0.3–1 per 100,000 [1].

Primary cutaneous B-cell lymphomas (PCBCLs) differ clinically and have a different prognosis when compared to histopathologically similar systemic lymphomas. The World Health Organization-European Organization for Research and Treatment of Cancer defines PCBCLs as malignant lymphomas that are confined to the skin at presentation after complete staging procedures [2, 3]. PCDLBCL-LT presents as rapidly growing red or blue-red tumors on one or both legs and often occurs in elderly women.

Histopathologic features of these lymphomas include a non-epidermotropic, diffuse infiltrate of centroblasts and immunoblasts in the dermis. Large cells usually predominate over small B cells and there are very few reactive T cells, mostly around the vasculature [4].

CASE REPORT

A 54-year old woman was referred to our Department, for a one year history of two bluish-red nodular lesions, of about 4–5 cm of diameters, located on the left side of the face. The lesions were asymptomatic, with very slow growth in size, non-ulcerated and with no submandibular adenopathy. On physical examination the two nodules were not adherent to subcutis. The patient was in good general health, and she described the appearance of a small ulceration on the hard palate, one year ago, in parallel with the nodules on the face.

In the recent past, she has been several times addressed to Otolaryngology Department, where she was diagnosed with aphtous ulceration and nodular fibroma of the face. She underwent many laboratory investigations and topical treatments that aimed treating a potential inflammatory condition, but with no obvious improvement.

Both lesions from the face were excised and a small punch-biopsy was taken from the lesion on the hard palate. The histopatological examination of the three lesions came with the same result: diffuse large B-cell lymphomas.

The histological examination revealed diffuse non-epidermotropic infiltrates predominantly composed of lymphocytes, with perivascular, periadnexial disposition and some involvement of nervous fibers. The malignant cell type consisted of small and medium lymphocytes, some monocitoid-like or plasmocytoid-like, with atypical nuclei and frequent mitosis. The epidermis was normal without signs of invasion (Figure 1).

Immunohistochemistry of the biopsies (Figure 2) established the final diagnosis of cutaneous B cell lymphoma (CD20 positive and Ki67 positive). Multiple immunohistochemical markers were used in the diagnosis of lymphoma in order to determine the best therapeutic strategy. Representative images of the results are shown in Figure 2 and summarized in Table 1.

After being diagnosed with primary cutaneous B-cell lymphoma localized on the face and hard palate, the patient was immediately referred to the Oncology Department for further investigations, staging and treatment. Unfortunately 24 hours after hospitalization she died of pulmonary embolism.

TABLE 1.
The battery of markers used to diagnose and determine the prognosis of lymphoma for the case described, and their results.

Marker	Result
Citokeratin (CKc)	negative
S100	negative
CD68	negative
LCA	negative
Vimentin (VM)	**positive** – focally
UCHL1	negative
CD5	negative
CD20	**positive**
CD10	negative
CD15	negative
Ki67	**positive** on nuclear level in 46% of tumoral cells

DISCUSSION

The primary cutaneous B-cell lymphoma represents cutaneous B-cell lymphoma (CBCL). CBCL are also: primary cutaneous marginal zone B-cell lymphoma; primary cutaneous follicle center lymphoma; primary cutaneous diffuselarge B-cell lymphoma, leg type; primary cutaneous diffuse large B-cell lymphoma, other (intravascular large B-cell lymphoma).

The primary cutaneous B-cell lymphoma is an uncommon type of cutaneous lymphoma accounting for 20–25% of all cutaneous lymphomas [5–7]. Within this low percentage of B-cell lymphoma, the dominant type is the diffuse large B-cell lymphomas that usually occurs in elderly patients and are typically located in the lower legs [2, 8].

FIGURE 1.
Haematoxilin-eosin staining of tissue taken by biopsy from two separate lesions. A – biopsy taken from the ulcerated palatal lesion showing intense limphocytic infiltrate in the muscle tissue with multiple areas of necrosis (black arrows); B – biopsy from the non-ulcerated nodular lesion located on the face, characterized by massive lymphocytic invasion. Scale bar: 10 µm.

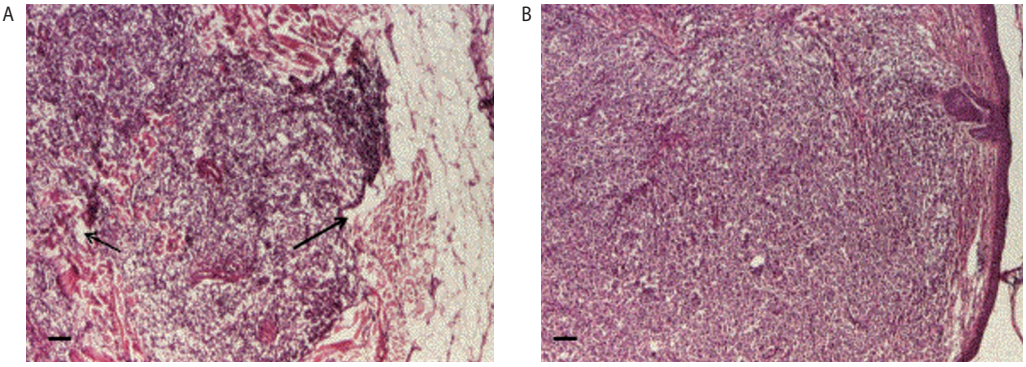
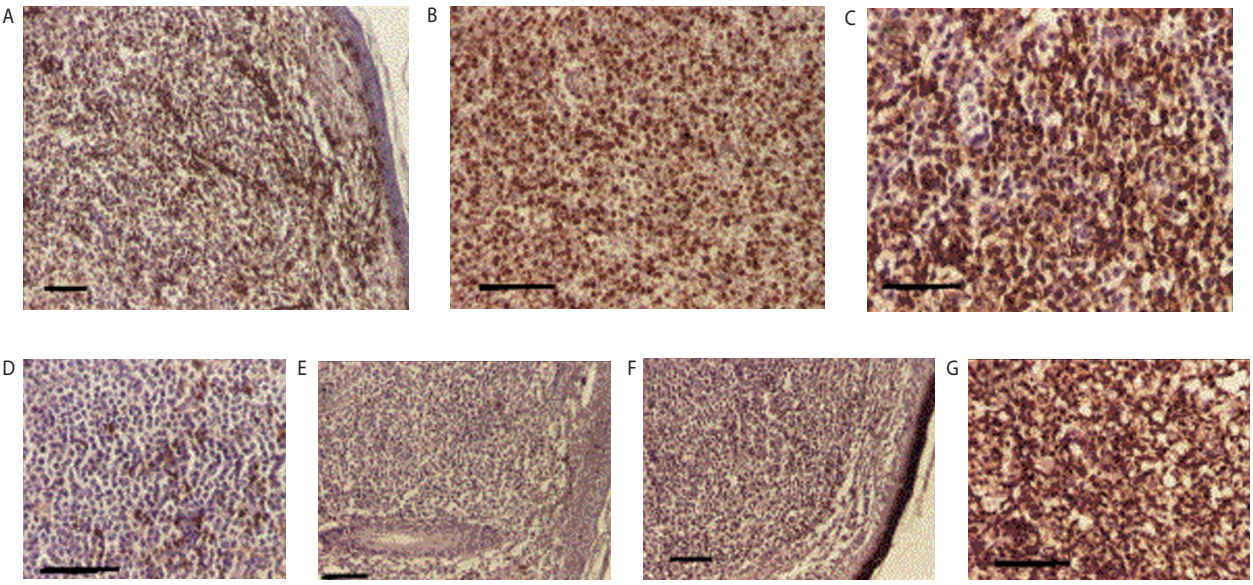


FIGURE 2.
Representative histological images for the markers used to determine the diagnosis and prognostic of the disease. A – vimentin, focally positive; B – CD20, positive; C – Ki67, positive; D – CKc, negative; E – LCA, negative; F – UCHL, negative; G – CD5, negative. Scale bar: 100 µm.



Most lymphomas are substantially defined by their immune profile. As such, the case described was characterized based on characteristic lymphocytic markers. Thus, the CD20 marker as well as vimentin – a marker of mesenchyme-derived cells confirmed the type of lymphoma. Ki67, a sensitive marker of proliferation and therefore of tumor aggressiveness, was also positive in the case subject (Figure 2).

The patient was a relatively young woman, with three lesions on the face and hard palate, with a one-year period of an apparent slow evolution and afulminate ending. However, the characteristic of this case was an apparent benign skin disorder that had very few of the typical signs associated to a malignant nature of the disease.

Oliveira A, et al. reported of a man, 80 years of age, with aprimary cutaneous marginal B-cell lymphoma of the chin [9].

Authors from India presented a 20-year-old woman presented with multiple painless nodular swellings on the skin of the extremities and face, without any systemic symptoms. Biopsy with immunohistochemistry revealed a diagnosis of precursor B-cell lymphoblastic lymphoma. There was no extracutaneous site of involvement. The patient denied chemotherapy and was subsequently lost to follow-up. She presented with symptomatic dis-

seminated disease 18 months later and rapidly succumbed to her illness [10].

Unfortunately, after such a slow onset of the disease, the evolution was very rapid through pulmonary embolism.

CONCLUSION

Cutaneous B-cell lymphoma is an unusual and rare skin neoplasm with a great range of clinical presentations and this is an alarm sign for apparent common skin lesion.

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