Cutaneous follicle center lymphoma of the eyelids: Unusual location

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INTRODUCTION

Primary cutaneous follicle center lymphoma is the most common primary cutaneous B-cell lymphoma. It is an indolent lymphoma, which develops from B cells in the germinal center without evidence of systemic or lymph node involvement at the time of diagnosis. Its incidence increases with age. The median age at diagnosis is 60 years.

In this article, we report the case of a patient with cutaneous follicle center lymphoma that initially manifested as eyelid nodules.

CASE REPORT

An 86-year-old patient with hypertension and ischemic heart disease and taking angiotensin–converting-enzyme inhibitors, β-blockers, antiplatelet drugs, and statins consulted for painless palpebral swellings progressing for 2 years. The swellings were gradually increasing in size to the point of obstructing the patient's sight.

Clinical examination revealed 6 erythematous nodules that were hard, bilateral, subcutaneous, symmetric, superficially fixed but mobile deeper down, in the 4 eyelids but also in both cheeks. The rest of the clinical examination was normal; notably, no adenopathy was observed (Fig 1, *A* and *B*).

A punch biopsy was performed on the lower portion of the left eyelid. On histologic examination, a malignant lymphoid tumor infiltration with cytonuclear atypia and the formation of germinal centers was found. The immunophenotypic study revealed positivity for anti-CD20, anti-CD10, and anti-B-cell lymphoma 6 antibody markers, whereas follicular center cells were negative for anti-B-cell lymphoma-2 antibodies. This histologic image and this immunohistochemical profile were compatible with a cutaneous follicle center lymphoma (Figs 2 and 3).

An extension assessment was carried out. Magnetic resonance imaging confirmed the lesion processes found clinically with extension toward the extraconical orbital fat and medially toward the tear ducts, sheathing the internal rectus muscle bilaterally, associating a bilateral superficial jugal process respecting the maxillary sinuses.

A complete biologic workup was performed, including a blood cell count and flow cytometry, which were normal, as well as a bone-marrow biopsy, which exhibited no abnormality, ruling out the possibility of medullary localization of follicle center lymphoma.

DISCUSSION

Primary cutaneous follicle center lymphoma accounts for approximately 10% to 20% of all cutaneous lymphomas and 50% of primary cutaneous B lymphomas.[1]

Clinically, it usually presents as a solitary erythematous or purplish papule but also sometimes presents as multiple papules, plaques or tumors localized on the forehead, neck and upper portion of the back, usually observed in elderly patients.

Histopathologically, primary cutaneous follicle center lymphomas are characterized by a pattern of follicular, diffuse or mixed growth, which is made of

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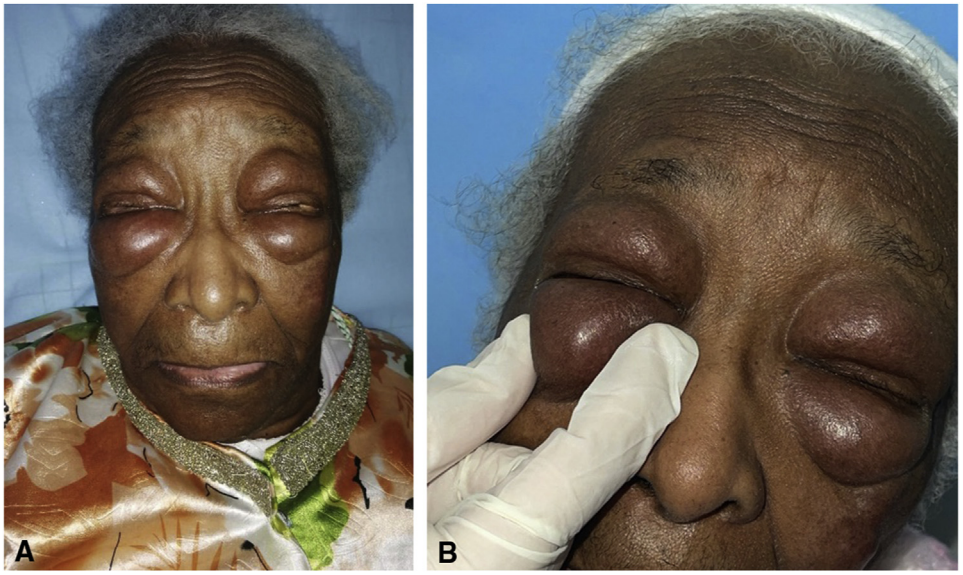

Fig 1. A, Bilateral and symmetric palpebral erythematosus nodules. **B,** Palpable nodule of the lower portion of the right eyelid.

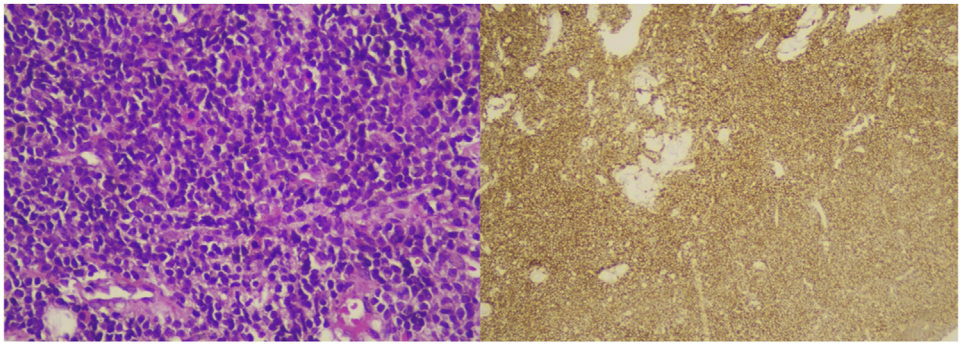

Fig 2. Lymphoid tumor infiltrate made of small-to-medium—sized cells. Rounded and notched nuclei with fine, granular chromatin without visible nucleoli were observed. Immunohistochemical study with anti-CD20^{+} antibody labeling (Hematoxylin-eosin stain).

large centrocytes and centroblasts derived from B cells of the germinal center.

Primary cutaneous follicle center lymphoma neoplastic lymphocytes express B-cell markers (CD19, CD20, CD22, CD79a, PAX5) and at least one follicular center marker, which is usually *BCL6* or, less frequently, CD10. *BCL2* staining is generally negative. Ki-67 may show a lower proliferation index than usual in reactive follicles.[2]

In this case, the patient had a very atypical involvement of the skin, with bilateral and symmetric eyelid nodules limiting the palpebral opening. The diagnosis of cutaneous follicle center lymphoma was confirmed on 2 skin biopsies with an extension assessment ruling out systemic or lymph node involvement.

Polychemotherapy including rituximab, cyclophosphamide, vincristine, and prednisone has been proposed as far as radiotherapy, which is normally indicated in localized forms, is contraindicated due to the lesion's localization, but the patient refused any treatment.

Lymphoma of the eyelid remains a poorly studied topic; only about 10 cases have been reported, with no large studies or reviews in the recent literature.

Conclusion

The recognition of the different presentations of cutaneous B-cell lymphoma, whether classic, atypical, or rare types is important for the clinician in order to detect B-cell lymphoma as early as possible, since the prognosis is better if the disease is

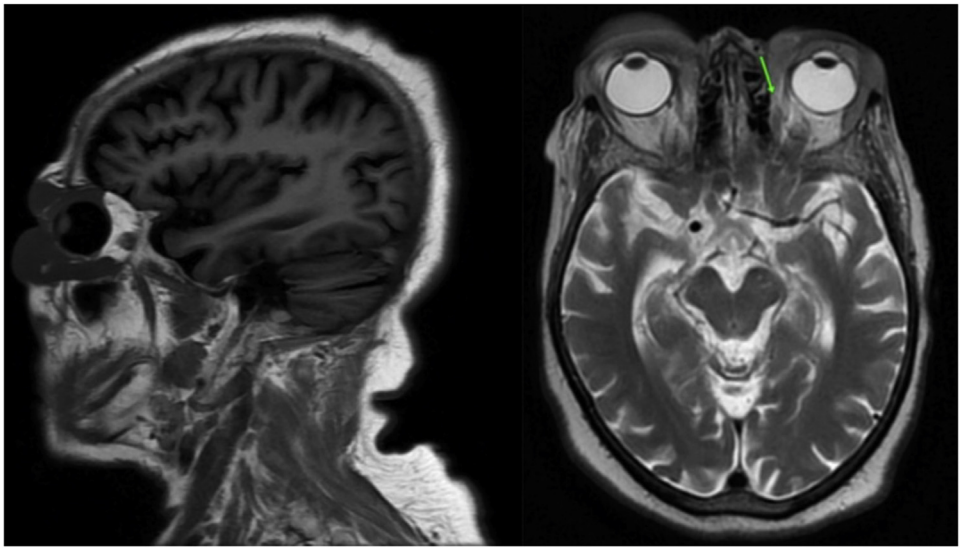

Fig 3. Magnetic resonance imaging of the facial mass: transverse and sagittal section of the lesion processes.

adequately managed at an early stage. Multiple and repeated skin biopsies should not be avoided to look for signs of B-cell lymphoma using histology and immunohistochemistry.

Once the diagnosis has been made, it is important to distinguish a cutaneous follicle center lymphoma from a secondary cutaneous localization of a systemic follicular B lymphoma, by staging, including a computed tomography scan, a positron emission tomography scan, and a bone-marrow biopsy.

Conflicts of interest

None disclosed.

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