

Primary bilateral and symmetric MALT lymphoma of the lacrimal sac mimicking chronic dacryocystitis

Linfoma MALT primário, bilateral e simétrico do saco lacrimal simulando dacriocistite crônica

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ABSTRACT

We report a case of primary bilateral mucosa associated lymphoid tissue (MALT) lymphoma of the lacrimal sac. MALT lymphoma is a subtype of Non-Hodgkin's Lymphoma of the ocular adnexa. When the primary site of the lymphoma is the lacrimal sac, it mimics chronic dacryocystitis. This may delay diagnosis, with potentially lethal results.

Keywords: Lacrimal apparatus diseases; Lymphoma, b-cell, marginal zone; Biopsy; Orbit/pathology; Case report

RESUMO

Descrevemos um caso de um linfoma MALT bilateral, simétrico e primário de saco lacrimal. O linfoma MALT é um subtipo do Linfoma Não-Hodgkin dos anexos oculares. Quando o local primário do linfoma é o saco lacrimal, ele pode simular uma dacriocistite crônica. Essa situação pode atrasar o diagnóstico e ter consequências fatais.

Descritores: Doenças do aparelho lacrimal; Linfoma de zona marginal tipo células B; Biopsia; Órbita/patologia; Relatos de casos

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INTRODUCTION

Lymphomas of mucosa associated lymphoid tissue (MALT) are extranodal marginal zone B cell tumors. They are the most common subtype of Non-Hodgkin's Lymphoma of the ocular adnexa. MALT lymphomas account for 5-10% of phenotype B, occurring at various sites, including the digestive tract, lungs, ocular adnexa, skin, thyroid, and breast.⁽¹⁻⁴⁾

Lymphoid neoplasms comprise 10–15% of all tumors in the ocular region. Lymphomas of the eye may originate in the periorbital and/or the orbital region (primary lymphoma), or may appear in this region as a result of systemic spread (secondary lymphoma).⁽⁵⁾ Here we report a rare case of a bilateral and symmetric MALT lymphoma of the lacrimal sac, mimicking chronic dacryocystitis.

CASE REPORT

A 62-year-old man presented with complaints of epiphora and masses in the region of both lacrimal sacs. The masses had grown slowly and gradually over the course of one year. He was diagnosed as chronic dacryocystitis and dacryocystorhinostomy was suggested at another hospital. At our institution, physical examination revealed bilateral, fibroelastic masses in the region of the lacrimal sacs that were painless to palpation. The masses extended beyond the limits of the medial canthal tendon (Figure 1A). There was no reflux upon lacrimal sac compression. Syringing elicited reflux from the opposite canaliculus; probing revealed a soft stop. Magnetic resonance imaging revealed bilateral symmetrical tumors in each lacrimal fossa. There was hyperintensity on T1 and T2-weighting, compatible with epithelial tumor or lymphoma (Figure 1B and 1C). An incisional biopsy was performed on both lesions (Figure 1D). Pathology revealed diffuse infiltration of small round lymphocytes, compatible with MALT lymphoma (Figure 2). On immunohistochemical examination, the lesions were positive for CD20 and B-cell-specific activator protein (DAK-Pax-5). The patient was evaluated by clinical oncologists, it was performed whole body scintigraphy who found no alternative primary site for the tumors. Chemotherapy with rituximab and chlorambucil was initiated, with favorable results.

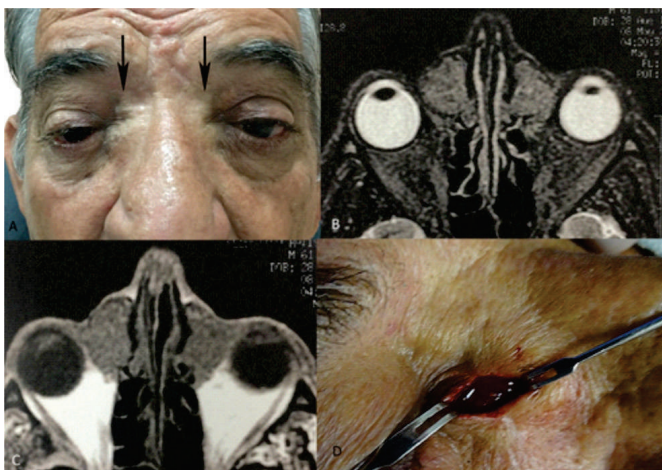


Figure 1: A: The masses beyond the limits of the medial canthal tendon (black arrows). B and C: Bilateral hyperintensity on T1 and T2-weighting. D: incisional biopsy

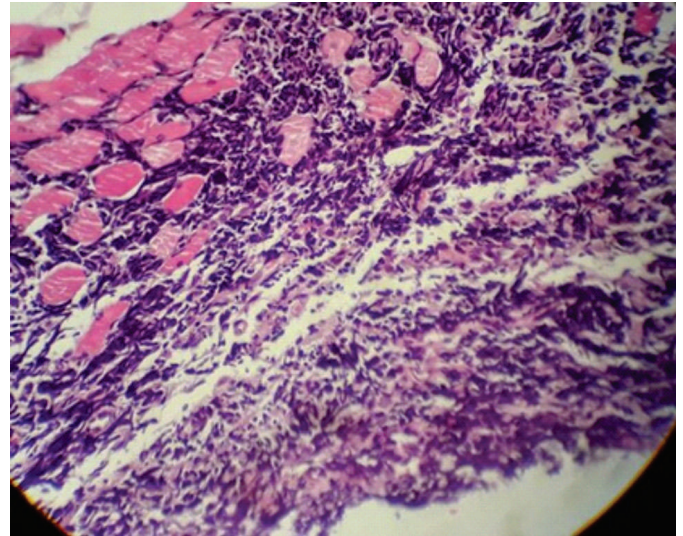


Figure 2: Diffuse infiltration of small round lymphocytes, compatible with MALT lymphoma

DISCUSSION

The majority of lymphomas in the ocular region are extranodal marginal zone B-cell lymphomas of the mucosa associated lymphoid tissue type (MALT lymphomas), according to the Revised European-American Lymphoma and the World Health Organization classification system.^(2,3,5)

MALT lymphomas may arise from the lacrimal sac, since this structure contains lymphoid tissue as part of the mucosa-associated lymphoid system. These lymphomas are the most common subtype, comprising one third of all primary lacrimal sac lymphomas.⁽⁶⁾

The pathogenesis of MALT lymphoma depends on the biology and function of resident lymphocytes and on lymphocyte homing to the particular site.⁽⁴⁾ Depending on the location of the tumor, clinical manifestations may include conjunctival salmon patches, ptosis, or lid swelling. With orbital involvement, there may be painless proptosis and diplopia. Epiphora may be caused by infiltration of the lacrimal sac.⁽⁵⁾ Lymphomas arising in the lacrimal sac are uncommon. In the literature, fewer than 50 cases of primary lacrimal sac lymphoma have been described over a period of 30 years.⁽⁵⁾ The symptoms can mimic those of chronic dacryocystitis, as in our case.^(1,4-7)

Epiphora with a swollen lacrimal sac wall is a diagnostic sign of lacrimal sac tumors, as well as of tumors located above the medial canthal tendon. However, in lacrimal sac MALT lymphoma, epiphora precedes tumor palpation by several months. Therefore, epiphora is a nonspecific symptom that is also present in acquired nasolacrimal duct obstruction.^(6,7) Chronic dacryocystitis sometimes demonstrates a nodular lymphoid infiltration pattern in subepithelial and intraepithelial areas, mimicking MALT lymphoma. However, chronic dacryocystitis generally shows a diffuse infiltration pattern. Increased goblet cell density and hyperplastic mucosal glands in cases with chronic dacryocystitis are also helpful for making a differential diagnosis.⁽⁶⁾

Mild MALT lymphoma of the lacrimal sac is characterized by chronic inflammation. It causes secondary obstruction of the nasolacrimal duct with non-specific clinical features that

cannot differentiate it from primary obstruction. Clinical suspicion of lacrimal sac lymphoma must be followed-up with imaging: dacryocystography, computed tomography, or magnetic resonance imaging. The definitive diagnosis is always made by histopathological examination. ⁽¹⁾

Abdelkhalek et al. ⁽¹⁾ described a similar case of bilateral MALT lymphoma of the lacrimal sac, but without palpable mass. Their patient presented with right-sided swelling of the inner canthal region, associated with an epiphora, evolving for three months. This was followed by left-sided symptoms one month later. Orbital computed tomography revealed a bilateral heterogeneous mass arising from the lacrimal sac, without infiltration of nearby structures or bone erosion. Biopsy with pathological examination and immunocytochemistry confirmed a MALT lymphoma.

Radiotherapy, chemotherapy and immunotherapy are effective treatments for lacrimal sac MALT lymphoma. Surgery should be limited to biopsy in order to avoid complications. A precise preoperative diagnosis is necessary for proper surgical planning. ^(1,6,8)

MALT lymphomas of the lacrimal sac are rare but not exceptional tumors. They may transform into more aggressive subtypes, including diffuse large B cell lymphoma. Therefore, early diagnosis of lacrimal sac lymphomas is essential. ⁽³⁾ In the context of any suspicious clinical sign, a biopsy should be performed. ⁽¹⁾ Our motivation in reporting this rare case of bilateral MALT lymphoma of the lacrimal sac is to help ophthalmologists recognize a disease that can become severe if left untreated, but which has a relatively good prognosis if treated early.

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