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CASE REPORT

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# Dacryocystitis Caused by Lymphoproliferative Infiltration in the Course of Lymphocytic Lymphoma: Case Report

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**ABSTRACT**

**Background:** Lacrimal drainage system lymphomas are rare, accounting for less than 10% of lacrimal sac tumors. They often appear as a secondary involvement within the confines of systemic lymphoproliferative disorders, therefore detailed ophthalmological examination and auxiliary testing is necessary to have an accurate diagnosis. **Case report:** We present the case of a 72-year-old woman with a medical history of chronic lymphocytic leukemia and small lymphocytic lymphoma. She presented to the ophthalmology clinic with a painful, discharging swelling in the right lacrimal sac area. Magnetic resonance imaging revealed a mass in the right nasolacrimal duct. A right external dacryocystorhinostomy was performed. The histological findings led to a diagnosis of small lymphocytic lymphoma. **Conclusions:** Special attention is required in cases of known systemic hematological disorders that associate with nasolacrimal duct obstruction or epiphora. Quick recognition and a full history with multidisciplinary clinical and diagnostic workup are fundamental to plan the treatment.

**Keywords:** dacryocystitis, lymphoproliferative disease, lymphoma, lacrimal sac

## INTRODUCTION

Lacrimal drainage system (LDS) pathologies, especially chronic dacryocystitis, represent 3% of cases that present to ophthalmologic clinic consults.[1]

The cardinal symptoms that draw the patients' attention are: epiphora, lacrimal duct obstruction, accumulation of mucous secretion and desquamated cells, leading to a subsequent infection caused by the spread of microorganisms from the surrounding areas (conjunctiva, nasal cavity).[1,2]

Chronic dacryocystitis can be caused by specific inflammation, traumatic damage, mechanical obstruction, and neoplasms. However, in the majority of

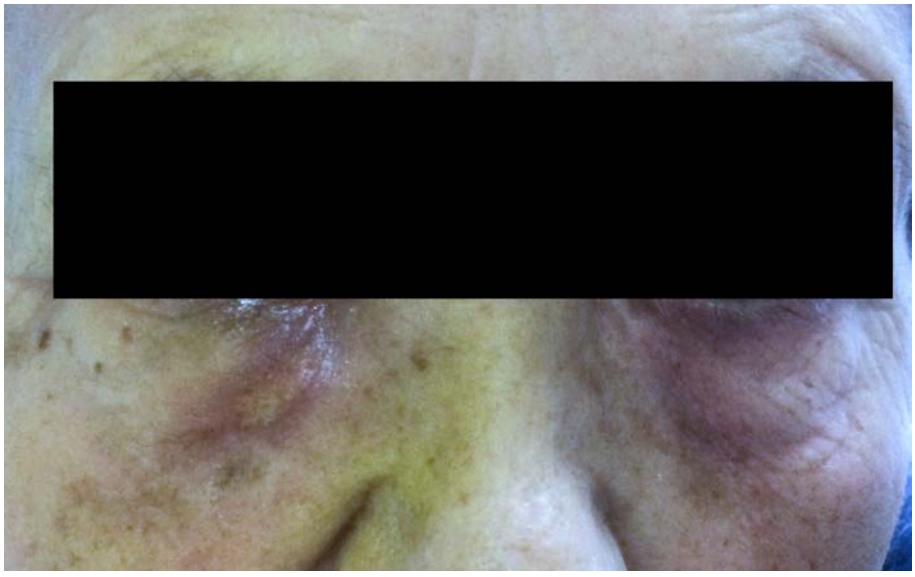

**FIGURE 1.** The patient's appearance at admission

cases, the etiology of dacryostenosis is idiopathic. It supposedly develops secondarily to an ascending inflammation from the nasal cavity and adjacent sinuses.[3]

Even though uncommon, malignancies can appear as obstructive neoplasms; therefore, a detailed ophthalmological examination and imaging are essential for a correct diagnosis. Lacrimal sac neoplasms appear on computed tomography as orbital tumor masses with or without local bone invasion.[4,5] Dacryocystorhinostomy is fundamental for confirming the diagnosis and planning the adjuvant therapy.[6]

Based on histopathological examination, 90% of lacrimal sac tumors are of epithelial origin, while lymphomas are very uncommon, representing less than 10% of lacrimal sac tumors.

LDS lymphomas usually appear as a secondary involvement within a systemic lymphoproliferative disorder.[5]

## CASE REPORT

A 72-year-old woman, with a medical history of chronic lymphocytic leukemia (CLL) and small lymphocytic lymphoma (SLL) since July 2015, presented to the ophthalmology clinic with a painful, discharging swelling in the right lacrimal sac area.

The initial diagnosis was established after the histopathological examination of an enlarged laterocervical lymph node, in which the immunohistochemistry revealed cells positive for CD20, CD79a, CD23, CD5, and BCL-2.

At admission, her ophthalmologic status was:

- best-corrected visual acuity: right eye (RE) 5/9, left eye (LE) 5/12;
- intraocular pressure (Goldmann tonometry): both eyes (BE) 19 mmHg;
- anterior segment evaluation: cortical cataract in BE, and the LE presented with a painful hard mass over the right lacrimal sac, edema, and erythema with nasolacrimal duct obstruction (Figure 1).

Differential diagnoses that were taken into account included acute blepharitis, bacterial conjunctivitis, basal cell carcinoma, chalazion, dermoid cyst, sinus tumors, obstruction of the nasolacrimal duct, and orbital cellulitis, which were all excluded by the patient's history of acute dacryocystitis. She was previously treated for acute dacryocystitis with systemic antibiotics that led to the subsiding symptoms.

She was diagnosed with recurrent right-sided dacryocystitis and cortical cataract in both eyes, and underwent systemic and topical antibiotic, as well as non-steroid anti-inflammatory treatment.

The patient was referred for additional ancillary testing and was later listed for a right external dacryocystorhinostomy.

Magnetic resonance imaging (MRI) revealed a 36/19 mm mass in the right nasolacrimal duct, without bone erosion and multiple laterocervical adenopathies.

Preoperatively, the lacrimal sac was noted to be inflamed and a biopsy was collected.

The histopathological examination revealed diffuse infiltration of small round lymphocytes, with round nucleus. On immunohistochemistry, the tumor cells were positive for CD20, CD5, CD23, and BCL-2, and negative for CD10 and cyclin-D1. In some areas, the cells were positive for

CD68. The proliferation index Ki-67 was 20%; thus, the histological findings led to a diagnosis of small lymphocytic lymphoma.

Taking into account the histological findings of the right-side nasolacrimal tumor, it was considered that there was a transformation of chronic lymphocytic leukemia into small lymphocytic lymphoma, hence leading to initiation of a treatment with vincristine, cyclophosphamide, and epirubicin, together with anti-inflammatory drugs.

All tissue and data collection complied with the guidelines of the Helsinki Declaration, and the patient signed an informed consent form before the admission and surgery (which specified our right to use her personal data and picture).

## DISCUSSIONS

Non-Hodgkin lymphomas, such as CLL and SLL, are characterized by the presence of small B-lymphocytes that express CD5 and CD23 as cell surface antigens.[4] The difference between the two is that CLL is associated with a leukemic phase, while SLL with a nodal or solid phase. Because of their similarities, the World Health Organization classified the two malignancies as manifestations of the same disease and combined the two into one category.[7]

Obstruction of the LDS caused by neoplasms are classified into: primary LDS tumors (papilloma/squamous cell carcinoma); primary tumors of tissues surrounding the lacrimal system that invade this system (commonly eyelid carcinoma, capillary hemangioma, osteoma, and lymphoma); and tumors that metastasize in the nasolacrimal region.[8]

The lacrimal duct tissue is rich in mucosa-associated lymph tissue (MALT) and plays a significant role in lymphocyte recirculation, forming a functional unit with the cornea, the lacrimal gland, and the nasal mucosa. Therefore, unsurprisingly, it gives place for the development of primary or secondary hematologic malignancies.[5] However, according to Krishna *et al.*, less than 70 cases were described of secondary involvement.[6,9,10]

In Romania, during a sixteen-year period (between 1999 and 2015), 18 cases of chronic dacryocystitis were described by Costea *et al.*, from which 11.11% presented nodular lymphocytic inflammation.[2]

Despite the fact that it is an uncommon disease, considering that our patient had CLL in her medical history, and her dacryocystitis was recurrent, particular attention was required.

Consistent inflammation can mask pathologic processes; thus, biopsy and histopathological examination of the nasolacrimal tumor are essential in revealing the lacrimal sac pathology.[11,12]

Chronic dacryocystitis should be differentiated from MALT lymphoma of the lacrimal sac. While the chronic inflammation presents a diffuse infiltration pattern, lymphomas of the nasolacrimal duct show small to medium-sized round lymphocytes with round nuclei. Furthermore, if the immunohistochemical examination shows positivity of tumor cells for CD20, CD5, CD23, and BCL-2 antibodies (like in our case), lymphoma diagnosis can be established.[13,14] All lymphoproliferative lesions require an open biopsy for obtaining an adequate tissue specimen, essential in establishing the diagnosis.

Because of its locally invasive and poten¬tially life-threatening nature, quick recognition and a multi¬disciplinary treatment plan is urgent to cure and diminish the metastatic risk.

Lacrimal duct obstruction does not disappear permanently in most described cases, despite the regression of lymphomas due to therapy.[9] The same way, metastases can still appear years after remission, thus long-term follow-up is needed.[4,8,15,16]

## CONCLUSIONS

Taking into account what was mentioned above, we have to pay special attention to cases of known systemic hematological disorders that associate with nasolacrimal duct obstruction or epiphora. Biopsy during surgery provides further instructions in diagnosing clinically suspected or unexpected neoplasms. Histopathological examination is undoubtedly fundamental for an adequate treatment. Dacryocystorhinostomy and adjuvant chemotherapy is not enough for a successful remedy. Long-term follow-up and routinely asking for symptoms of epiphora is required to identify recurrences or metastases.

## CONFLICT OF INTEREST

The authors declare no conflict of interest.

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