ABSTRACT

Educational Objective: To describe the clinical, surgical, and pathologic findings in CLL/SLL of the nasolacrimal drainage system and ethmoid labyrinth.

Objectives: To underscore the significance of routine microscopic pathological review of all intranasal surgical specimens and describe the pathology of Chronic Lymphocytic Leukemia, Small Lymphocytic Lymphoma (CLL/SLL), involving the ethmoid bone and the nasolacrimal drainage system (NDS).

Study Design: Case report and literature review.

Methods: The clinical and pathological features of CLL/SLL involving the sinus and nasolacrimal duct mucosa and bone are described.

Results: Physical examination revealed tearing, increased tear lake, and poor eye discharge in the left eye. Nasal endoscopy showed a left septal spur and bilateral osteointernal complex crowding with mildly enlarged turbinates. Computed tomography of the sinuses demonstrated scattered sinus disease, a soft tissue prominence in the left medial canthus, and multiple enlarged lymph nodes throughout the neck bilaterally. The patient underwent a left endoscopic anterior ethmoidectomy and dacryocystorhinostomy. Routine surgical pathology of a sample of left sinus contents and septal cartilage revealed CLL/SLL, involving the upper respiratory tract mucosa and associated cancellous bone, confirmed by immunohistochemical studies.

Conclusions: Initial presentation of CLL/SLL within the NDS and ethmoid is rare and reflects a systemic lymphoreticular malignancy. While CLL/SLL is generally an indolent illness, early detection permits accurate clinical staging and appropriate therapy. Involvement of the ethmoid and nasolacrimal duct system with Infections such as CLL/SLL underscores the need for routine microscopic pathological review of all intranasal surgical specimens.

INTRODUCTION

Background:
- Epiphora, watering eye
- Chronic obstruction of the lacrimal drainage system (LDS) is common in the elderly and has a broad differential diagnosis, including:
  - granulomatous inflammation and infection, dermoid cyst, mucocele, lipoma, lymphangioma, squamous cell carcinoma, adenoid cystic carcinoma, hemangiomas, sarcomas, melanoma, lymphoma, and secondary involvement of lymphoreticular disorders.
- Neoplasms involving the LDS:
  - Uncommon, > 90% of epithelial origin.
  - In 2001, Lee-Wong et al published a series of 166 patients who underwent DCR for primary acquired nasolacrimal duct obstruction and found no neoplasms in 202 biopsy specimens.
  - In 2002, Yip et al published a review of all biopsies obtained during DCR performed at the Mayo Clinic (Rochester, MN) from 1969 to 1997 and found 11 of 381 patients to have lymphoid lymphomatous involvement.
- According to Yip et al:
  - The mucosa of the LDS serves as an "adherent" layer (mucosal-associated lymphoid tissue, or MALT).
  - MALT lymphomas involving the LDS may be primary or a "metastatic" lesion of a systemic lymphoma or leukemia.
- Lymphomatous involvement of the LDS is rare, and an uncommon cause of tearing in the elderly.

METHODS AND MATERIALS

Study Design: Case report and literature review.

Methods:
- The clinical and pathological features of CLL/SLL involving the sinus and nasolacrimal duct mucosa and bone are described.
- The patient underwent a septoplasty, left endoscopic anterior ethmoidectomy, and dacryocystorhinostomy (DCR).

RESULTS

Clinical Presentation:
- 63 year old woman presented with a one year history of left sided epiphora.
- Past medical history included chronic sinusitis, hypertension, and a remote history of a peripheral leukocytosis, for which she saw an oncologist who recommended therapy.

Physical Exam:
- Vision: 20/25 OD and 20/40 OS.
- No afferent pupillary defect.
- Extrinsic motility full.
- Tear lake increased on the left with poor eye discharge.
- Probing and irrigation on the left showed complete reflux with some mucous discharge.
- Nasal endoscopy revealed a left septal spur and bilateral osteo-internal complex crowding.

DISCUSSION

Chronic Lymphocytic Leukemia/Small Lymphocytic Lymphoma (CLL/SLL) of the lacrimal drainage system (LDS).

Subtle clinical and radiologic signs and symptoms:
- The most common presentation of Non-Hodgkins Lymphoma (NHL) involving the LDS is epiphora and either painless or painful medical canthal swelling. A palpable mass with prominent nuclei (prolymphocytes and paraimmunoblasts) can also be seen histologically. A lymphangiogram with CT evidence of a soft tissue mass in the left medial canthus region may expand into adjacent structures.
- This diagnosis can also be suggested by similar MRI findings.

REFERENCES
1. Please see handout.

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Diagnosis relies on confirmatory pathological analyses:
- CLL is a malignant monomorphic B cell proliferation of small, mature-appearing lymphocytes. A variable number of large cells with prominent nuclei (prolymphocytes and paraimmunoblasts) can also be seen histologically. An immunophenotypic expression of CD5, CD20, and CD22 (weak) positivity with CD23 and CD10 negativity support the diagnosis.
- A diagnosis of SLL is preferred in non-leukemic cases with tissue morphology and immunophenotype consistent with CLL/SLL.

CONCLUSIONS

- CLL of the LDS is rare and is an uncommon cause of epiphora and LDS obstruction.
- The radiologic findings of a soft tissue mass in the medial canthal region support the diagnosis of a lymphoproliferative disease involving the LDS.
- While CLL/SLL is generally an indolent disease, the prognosis and management when the LDS is involved is poorly defined.
- As more cases of CLL of the LDS are reported, and confirmed with immunohistochemical analyses, it may be possible to make more definitive conclusions regarding prognosis and optimal management.
- Although rare, involvement of the ethmoid labyrinth, nasal septum, and nasolacrimal duct system with diseases such as CLL/SLL underscores the need for routine microscopic pathological review of all intranasal and LDS surgical specimens.