REDISCOVERING THE PHYSICAL EXAM



Eyelid Mass Secondary to T-Cell Lymphoma in a Child: A Rare Presentation and Literature Review

healthy 3-year-old boy presented with a 3-week history of right upper eyelid redness and swelling, fever, anorexia, and decreased activity. Initially, he presented with a bump on his right upper eyelid that was diagnosed as a stye. He received erythromycin ointment, yet developed progressively worsening eyelid redness and swelling. At 2 weeks after onset, he received oral clindamycin for preseptal cellulitis. While on oral antibiotic therapy, the right upper eyelid symptoms persisted, and the patient developed decreased appetite and increased fatigue.

Physical examination revealed a fever of 102.3°F; right upper eyelid erythema, edema, ecchymosis, discharge, and ptosis (**Figure 1**); and anterior cervical lymphadenopathy. When lying supine for a computed tomography (CT) scan of the orbit without contrast, he developed respiratory distress. A subsequent chest radiograph revealed a large mediastinal mass and bilateral pleural effusions (**Figure 2**; available at www.jpeds.com), suggesting respiratory decompensation owing to positional airway compression.

Once stable, the patient underwent a CT scan of the chest and a CT scan of the orbit, which revealed a large anterior mediastinal mass compressing the trachea and a soft tissue mass in the upper eyelid (**Figures 3** and **4**; available at www.jpeds.com). These findings were concerning for malignancy. The patient underwent an ultrasound-guided percutaneous core biopsy of the anterior mediastinal mass. Immunohistochemistry and flow cytometry of the biopsy specimen revealed tumor cells consistent with aggressive T-cell lymphoma of medullary type T cells (**Figure 5**; available at www.jpeds.com).

We describe a case of aggressive secondary ocular adnexal lymphoma (OAL) of T-cell origin that presented as refractory preseptal cellulitis followed by acute respiratory decompensation.

Factors that guide diagnosis of swollen, erythematous eyelid mass lesions in this age group include pain, onset, identifiable mass vs diffuse eyelid swelling, ecchymosis, and systemic symptoms. Hordeola and styes are well-defined, painful masses often near the eyelid margin that typically resolve spontaneously and are not associated with fever. Preseptal and orbital cellulitis presents with diffuse edema, fever, and eye pain. It classically results from skin trauma, hordeola and styes, or sinusitis. While preseptal cellulitis is limited to the eyelid, orbital cellulitis is a deeper infection, which results in decreased vision, an afferent pupillary defect, proptosis, and limited ocular motility. Orbital tumors—primary, metastatic, and lymphomas—



Figure 1. External ocular photograph showing the right upper eyelid. Note the mild ecchymosis.

often have an insidious, painless onset of diffuse lid swelling associated with possible proptosis, ecchymosis, and systemic signs such as anorexia, fever, and fatigue.

OALs are lymphoproliferative tumors involving orbital structures such as the conjunctiva, eyelid, lacrimal gland, orbital soft tissue, and/or extraocular muscles.¹ The most frequently encountered histopathologic subtypes are low-grade B-cell lymphomas, such as marginal zone lymphomas and follicular lymphomas, followed by various high-grade B-cell lymphomas.² Only 1-3% of OALs are of T cell or natural killer cell origin.¹ Of these, T-cell OALs most commonly present as a manifestation of known systemic peripheral T-cell lymphoma.³ In a large retrospective study, only 1.4% of cases occurred in individuals under age 21 years.⁴ Only a few other cases of T-cell OALs have been reported in the pediatric population (**Table**; available at www.jpeds.com).⁵⁻⁹

T-cell OALs are known to be associated with poorer outcomes compared with the more common B-cell–derived OALs.¹⁰ Because more than one-half of OALs also encompass extraorbital involvement, a thorough systemic workup is imperative.² Our case highlights the importance of including OAL in the differential diagnosis for cases of presumed preseptal cellulitis or hordeolum that are refractory to treatment. Failure to recognize this disease entity can lead to rapid deterioration. ■

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Figure 2. Frontal chest X-ray showing a large mediastinal mass (*arrowheads*).

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Figure 4. Axial image from contrast-enhanced orbital CT scan showing an enhancing preseptal process (*arrows*) in the right upper eyelid without invasion into the globe or orbit.



Figure 5. Mediastinal tumor, core biopsy histological and immunophenotypic features. **A** and **B**, Histological examination of the mass shows sheets of predominantly medium-sized neoplastic cells with a moderate amount of basophilic cytoplasm, irregular nuclear contours, somewhat clumped chromatin, and inconspicuous nucleoli. (Original magnification: **A**, 10×; **B**, 40×). **C** and **D**, Immunophenotyping by immunohistochemistry reveals that neoplastic cells are positive for **C**, CD2, CD5, CD7, CD8, and CD4 (partial), and negative for **D**, CD1a, CD10, CD34, CD57, PAX-5, CD30, and ALK-1.

Table. Literature review of cases of OAL in the pediatric population				
Articles	Year	Type of OAL	Age, y	Outcome
Amit et al ⁶	2012	Primary	6	Death within 1.5 mo
Hassan and Elner ⁷	2005	Secondary, known peripheral T-cell lymphoma, subpanniculitic features	6	Responsive to local radiation and chemotherapy
Cruz et al ⁸	2004	Possibly primary, sinonasal subtype	11	Chemothrapy initiated; death from sepsis and central nervous system infection
Bakhshi and Sidhu ⁵	2008	Unknown	Unknown	Unknown