lens with fusion of vestigial lens remnants with the retrolental fibrovascular membrane.[3] Despite these atypical features, a diagnosis of PFV was supported by the central dragging of ciliary processes, the spokelike pattern of persistent fetal vessels within the plaque, and the hyaloid stalk extending from the plaque to the optic disk. Since PFV is an important consideration in the differential diagnosis of leukocoria, it is critical to be familiar not only with typical clinical features but also with atypical features, as demonstrated by our case.

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Primary lacrimal sac diffuse large B-cell lymphoma in a child

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We report the case of a 13-year-old boy who presented with a 2-month history of left eyelid swelling, ecchymosis, and epiphora. Magnetic resonance imaging revealed a lobulated lesion in the region of the left lacrimal sac extending to the left nasolacrimal duct. Biopsy revealed diffuse large B-cell lymphoma of the lacrimal sac. Chemotherapy was initiated, consisting of rituximab, methotrexate, cytarabine, doxorubicin, cyclophosphamide, and vincristine. The lesion resolved within weeks of treatment, and the patient remained disease free at 1 year. Primary orbital lymphoma is rare in children; primary diffuse large B-cell lymphoma of the lacrimal sac in a child has not been reported previously.

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Case Report

A 13-year-old white boy was referred by his pediatrician to an oculoplastics surgeon with a chief complaint of 2 months' progressive left lower eyelid swelling, ecchymosis, and epiphora (Figure 1). At the time of presentation, he was being treated with antibiotics for right ear otitis media and pneumonia. He had no significant past medical history and no past ocular history. There were no prior episodes of swelling, epiphora, or epistaxis. There was no history of trauma. Patient and family denied any recent weight loss, changes in appetite, fatigue, and night sweats.

On ophthalmological examination, his uncorrected visual acuity was 20/20 bilaterally. A palpable mass in the area of the left lacrimal sac and the left nasolacrimal duct was noted. The left lower eyelid was ecchymotic and edematous. Pupils were round and reactive, with no afferent pupillary defect. Extraocular movements were full. There was no proptosis and no resistance to retropulsion. Intraocular pressure was within normal limits in each eye. The rest of the slit-lamp and dilated examination was within normal limits. Physical examination was unremarkable for any palpable lymphadenopathy or organomegaly.

Magnetic resonance imaging (MRI) revealed a lobulated lesion molded to the bony space in the medial left orbit and extending to the left nasolacrimal duct, measuring 1.5 cm × 1.8 cm × 1.5 cm, with enhancement and mild diffusion restriction (Figure 2), initially concerning for rhabdomyosarcoma.

An incisional biopsy was performed, the results of which suggested a lymphoproliferative process, and a decision was made not to fully excise the lesion. Histopathologic examination showed sheets of tumor cells, medium to large in size, with irregular nuclei. No "starry-sky" appearance or necrosis was seen (Figure 3A). Immunohistochemistry staining showed CD20-positive, CD45-positive, and CD3-negative cells. Additionally, the tumor cells were strongly MUM-1 positive (Figure 3B). A diagnosis of diffuse large B-cell lymphoma (DLBCL) was made.

Systemic evaluation, including bone marrow biopsy, computerized tomography (CT) scan of the neck and chest, MRI of the abdomen and pelvis, and positron emission tomography/CT imaging revealed no other systemic evidence of lymphoma. Chemotherapy was initiated according to Children's Oncology Group Protocol ANHL01P1, group B, consisting of rituximab, methotrexate, cytarabine, doxorubicin, cyclophosphamide, and vincristine. Within weeks of treatment the lesion had resolved.

Discussion

Lymphoma around the eye typically originates from the conjunctiva, eyelid, or orbit.[1] Lacrimal sac lymphoma is unusual, with a median age of onset of 51 years old.[1]

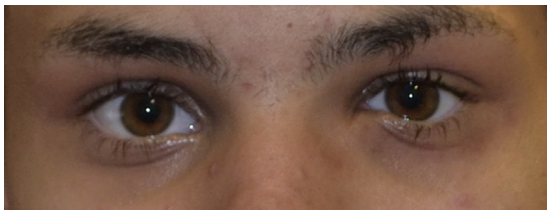

FIG 1. Clinical photograph of a 13-year-old boy with a 2-month history of progressive epiphora, ecchymosis, and swelling of the left lower eyelid.

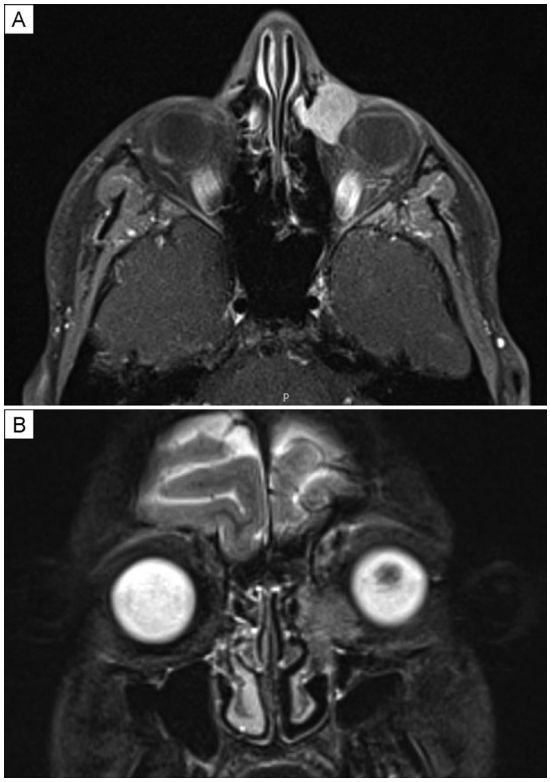

FIG 2. Magnetic resonance imaging showing lobulated lesion in the medial left orbit extending to the left nasolacrimal duct (A, axial; B, coronal).

Pediatric lacrimal sac lymphoma is exceedingly rare.[1-5] In a review of 250 pediatric orbital lesions published in 1986, Shields and colleagues[6] found only 6 orbital lymphoma cases, none of which were in the lacrimal sac. We identified only 4 pediatric cases of lymphoma of the lacrimal sac in the literature.[1-3,5]

Lacrimal sac lymphoma may present similarly to nasolacrimal duct obstruction.[1,2] Patients may have epiphora or painless swelling of the lacrimal sac area. It may present as a dacryocystitis.[1,2] Imaging is helpful in diagnosing a lacrimal sac tumor, with MRI preferred, given its better tumor definition and characterization of the cystic versus solid nature of the mass.[1,2] Histopathological examination is always required for definitive diagnosis.[1-3]

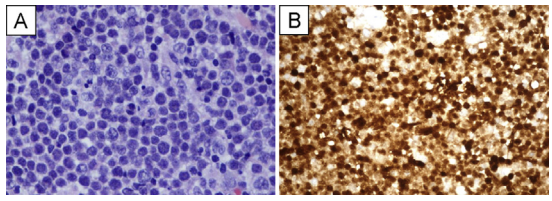

FIG 3. Sheets of tumor cells, medium to large in size with irregular nuclear. A, No "starry-sky" or necrosis is visible (hematoxylin and eosin; original magnification × 400). B, Tumor cells are strongly MUM-1 positive (original magnification × 400).

Primary orbital lymphoma is usually a low-grade mucosa-associated lymphoid tissue (MALT) lymphoma, more commonly found in adults, with a median age of presentation of 60 years.[4] Most common subtypes of primary orbital lymphoma include MALT (50%-80%) and follicular (about 20%).[4] Less common types include mantle cell lymphoma, lymphoplasmacytic lymphoma, anaplastic large cell lymphoma, T-cell lymphoma, and DLBCL.[4]

Of the 4 pediatric lacrimal sac lymphomas in the literature, 1 was a poorly differentiated lacrimal sac lymphoma in a 10-year-old treated with external beam radiation in 1970 and without recurrence at 11 months.[3] The second case was a MALT lymphoma in a 10-year-old treated with excision and chemotherapy and without recurrence at 30 months.[1] The third case started as a reactive lymphoid hyperplasia in a 9-year-old, which, after excision, recurred as a DLBCL and was subsequently treated with chemotherapy; the patient was in complete remission a year later.[2] The fourth case was a MALT lymphoma in a 10-year-old treated with radiation. To our knowledge, ours is the only case of lacrimal sac lymphoma in a child that started as a primary DLBCL.[5]

Given the sparsity of cases, the management remains controversial but generally includes surgery for diagnostic purposes, external beam radiotherapy for local disease, and chemotherapy for more aggressive subtypes.[1,2] At 1 year's follow-up, the patient was doing well, with no residual lesion in the lacrimal sac, nasolacrimal duct, or elsewhere. He remains asymptomatic, with no residual tearing. In older children who present with an acquired, nontraumatic nasolacrimal duct obstruction, tumors of the lacrimal sac and nasolacrimal duct should be considered in the differential diagnosis.

Literature Search

A MEDLINE search of the literature (1970-present), with no restriction, was conducted on October 21, 2018, using the following terms: *lacrimal sac lymphoma*, *nasolacrimal lymphoma*, *orbital lymphoma*, *pediatric lacrimal sac lymphoma*, *pediatric orbital lymphoma*, *orbital masses in children*, *lacrimal sac lymphoma in a child*, and *pediatric orbital masses*.

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Oral doxycycline and azithromycin in the management of recurrent conjunctival dehiscence following glaucoma drainage implantation in a child

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Aqueous drainage devices play an important role in the treatment of refractory glaucomas; however, they can be associated with early and late postoperative complications, including conjunctival dehiscence, which must be repaired surgically. Recurrent dehiscence despite surgical repair is uncommon and can be sight threatening. We discuss possible causes and management options of recurrent conjunctival dehiscence in a 2-year-old and the role of oral doxycycline and azithromycin in its management.

Case Report

A 2-year-old boy with anterior segment dysgenesis and secondary glaucoma was referred to L V Prasad Eye Institute for keratoplasty. The child had undergone combined trabeculotomy with trabeculectomy in both the eyes 1 year previously at a local hospital and was currently on a single glaucoma medication. Examination under anesthesia revealed a flat bleb, grade 3 corneal haze with central corneal scar in both eyes, and peripheral iridocorneal adhesions. Horizontal corneal diameter was 10.5 mm in the right eye and 10 mm in the left eye. Intraocular pressure (IOP) was 12 mm Hg in the right eye and 14 mm Hg in the left eye by Perkins tonometry. The child underwent penetrating keratoplasty in each eye 3 months apart and, 4 months later, cataract surgery in both eyes. After keratoplasty and cataract surgery, the IOP increased to 34 mm Hg in the right eye and 28 mm Hg in the left eye. There was an increase in disk cupping from 0.3 to 0.6 in both eyes. Maximal medical therapy and repeat trabeculectomy with low-dose (0.02%) mitomycin-C failed to control IOP; hence, surgery with Aurolab Aqueous Drainage Implant (AADI; Aurolab, Tamil Nadu, India), an indigenous nonvalved implant (similar to Baerveldt), was planned in the right eye. After limbal-based conjunctival incision, the implant was placed in the inferonasal quadrant and fixed to the underlying sclera. The tube was ligated using two 6-0 polyglactin 910 sutures and inserted into the anterior chamber through a 3 mm tunnel, and the tube was covered using a scleral patch graft. The conjunctiva was closed in a continuous fashion using 8-0 polyglactin 910 suture on a tapered round-bodied needle.

Five days after AADI implantation, the child presented emergently with a whitish appearance in the right eye noticed since that morning. Examination under anesthesia revealed an 8 × 4 mm conjunctival dehiscence with broken polyglactin 910 suture. The underlying scleral patch graft was exposed; there was no leak or tube exposure (Figure 1A). There was no history of ocular trauma or eye rubbing. Due to a large conjunctival defect, resuturing was planned. Conjunctival margins were freshened and sutured using 8-0 polyglactin 910 in a continuous fashion; additional interrupted 10-0 nylon sutures were applied (Figure 1B). Topical medications were continued, and the parents were instructed to shield the eye, to avoid retraction of the lower lid for eye drop instillation and an elbow-splint to prevent eye rubbing.

Ten days after resuturing, the conjunctival dehiscence recurred at the same location. Repeat conjunctival closure was performed using 8-0 polyglactin 910, and additional 10-0 nylon sutures were used to anchor the conjunctiva to the underlying scleral patch graft. Because of recurrent dehiscence and possible altered wound healing response, we started the patient on systemic immunomodulators. Based on our experience with oral doxycycline (tetracycline antibiotic) in adults with conjunctival retraction and wound gape following implant surgery,[1] we planned to use it in this child, although its pediatric use is controversial in view of its dental side effects in children. After consulting the patient's pediatrician and with informed consent of the parents, oral doxycycline and azithromycin

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