cyst is a quick, easy, and minimally invasive procedure and hence often used as first-line management to provide immediate relief. However, it often offers only temporary benefit, with the cyst reforming within weeks.

The use of sclerosing agents in tandem with aspiration has shown good effect.[5,6] The sclerosing agent helps to bind the inner walls of the cyst, creating resistance to fluid reaccumulation. Some believe that because the cyst formation results from arrested closure of the fetal fissure (at approximately 4 weeks of fetal life at 8 mm stage), it may have an intracranial extension and that sclerosing agents should be avoided on that account.

Primary cyst excision is notoriously difficult to manage; the thin walls of the cyst can be easily ruptured at surgery, leading to loss of the surgical plane and eventually resulting in incomplete excision and cyst reformation. Some advocate removing the microphthalmic eye to ease cyst removal, cutting the optic nerve proximal to the microphthalmic cyst.[7] We prefer not to remove of the microphthalmic eye, because, we believe, its presence aids in orbital bony development.

Lesion inflation with fibrin glue to aid complete surgical excision has also been used with good effect in lymphangiomas,[8] anophthalmic socket cysts,[9] and conjunctival inclusion cysts.[10] In this case we drew on our previous experience using fibrin glue to completely excise orbital cystic lesions, such as lacrimal ductule cysts.[1]

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Persistent fetal vasculature presenting with axial elongation and platyphakia

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Leukocoria in children should always raise the concern for retinoblastoma. However, a variety of non-neoplastic conditions can also present with leukocoria, including persistent fetal vasculature (PFV), a nonhereditary, congenital anomaly caused by a failure of the fetal intraocular vasculature to regress during development. Classically PFV presents with features that make it easily distinguishable from retinoblastoma, including microphthalmia, retrolental fibrovascular membrane, central dragging of ciliary processes, and cataract. We present an atypical case of PFV in a 9-month-old boy who presented with the unusual features of axial myopia and platyphakia.

Case Report

A 9-month-old, full-term boy presented at the Bascom Palmer Ocular Oncology Service, Miami, for evaluation of leukocoria in the right eye noticed 2 weeks earlier by the mother. He had no prior ocular or medical history. The patient demonstrated mild developmental delay but no obvious systemic syndrome. Chromosomal testing revealed no copy number variants. On examination under anesthesia, corneal diameters were 12 mm in the right eye and 11 in the left eye. Intraocular pressure was 18 mm Hg in the right eye and 15 mm Hg in the left eye. The anterior chamber angle was open, with no detectable abnormality in both eyes. The remainder of the examination of the left eye was normal. Anterior segment examination of the right eye revealed that the crystalline lens was replaced by small, flat, whitish-gray, plaquelike patches anterior to a fibrovascular membrane with ciliary processes dragged centrally toward the plaque (Figure 1A).

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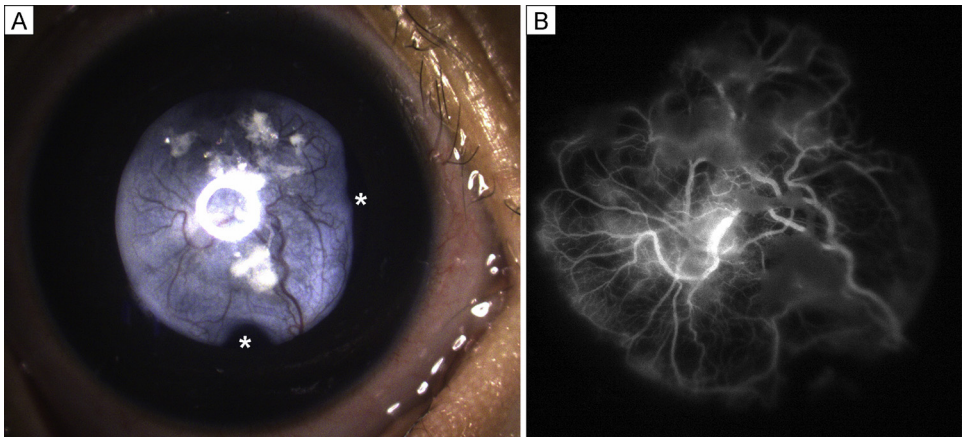

FIG 1. A, External photograph showing the retroiridal fibrovascular membrane, overlying crystalline lens remnants, and ciliary processes dragged centrally (asterisks). B, Fluorescein angiography showing a highly vascularized retroiridal membrane in a spokelike pattern consistent with persistent tunica vasculosa lentis, which is obscured in places by the intervening patches of residual lens material.

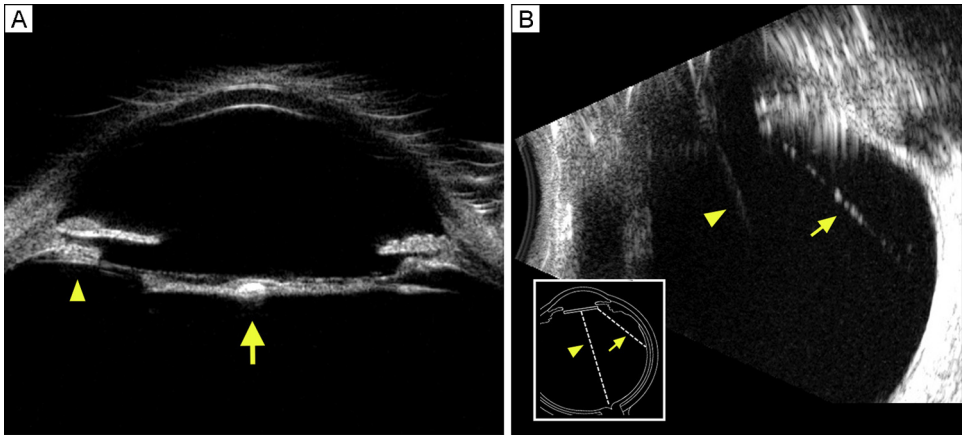

FIG 2. A, Anterior segment high-resolution B-scan ultrasonography showing highly reflective flat lens remnant (platyphakia) and underlying fibrovascular membrane (arrow); a central ultrasonic accentuation suggests the location of the insertion of the hyaloid artery remnant in the retrolental membrane (eg, Mittendorf dot). The ciliary processes are dragged centrally (arrowhead). B, Posterior B-scan ultrasonography showing the hyaloid artery remnant (arrowhead) and hyperechoic linear deposits that likely represent vestiges of the posterior tunica vasculosa lentis (arrow); illustration (inset) depicts ultrasound findings.

On A-scan ultrasonography, axial length of the right eye was 27 mm; of the left eye, 22.3 mm. Fluorescein angiography revealed marked vascularization of the membrane (Figure 1B) in a pattern compatible with the tunica vasculosa lentis. On anterior segment high-frequency B-scan ultrasonography, the lens remnant and membrane appeared as a flat, highly reflective retro-iridal plaque with ciliary processes stretched centrally (Figure 2A). There was no view of the posterior segment using indirect ophthalmoscopy. Posterior B-scan ultrasonography of the right eye revealed a delicate linear stalk extending from the plaque toward the optic disk consistent with a persistent remnant of the primary hyaloid artery.[1] Further, there was a linear arrangement of small mobile vitreous opacities extending from the peripheral edge of the plaque to the equator (Figure 2B), consistent with remnants of the vasa hyaloidea propria.[1] No solid mass was identified.

Discussion

Persistent fetal vasculature (PFV) is caused by a failure of one or more components of the fetal intraocular vasculature to regress during development, which can result in a broad range of clinical features, including microphthalmia, retrolental fibrovascular membrane, central dragging of ciliary processes, cataract, secondary angle closure glaucoma, hyaloid stalk, optic nerve dysplasia and hypoplasia, retinal folds and retinal detachment.[2-5] Although PFV is typically associated with microphthalmia, our case demonstrated an enlarged eye with axial elongation and mild corneal enlargement. Buphthalmos rarely has been described in association with PFV and may be caused by neonatal glaucoma,[1] although the intraocular pressure was normal in our case. Platyphakia (flat lens) is exceedingly rare in PFV and may represent absorption of the

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]