maintenance with resulting short telomere that are also associated with exudative retinopathies, including dyskeratosis congenita, Revesz syndrome, Hoyeraal-Hreidarsson syndrome, and cerebroretinal microangiopathy with calcifications and cysts.[1,2] Dyskeratosis congenita is the only one of these with telomere abnormalities in which the patient has normal development and neurologic function; the others syndromes are often associated with neurologic abnormalities and developmental delays.[3]

Dsykeratosis congenita can be associated with many different mutations that can cause autosomal dominant, autosomal recessive, or X-linked inheritance.[3,4] Clinically the patient presents with the diagnostic triad of dysplastic fingernails and/or toenails, lacy reticular pigmentation of the skin, and oral leukoplakia.[5] There are multiple other manifestations of the disease, including dental, gastrointestinal, skeletal, neurological, genitourinary, pulmonary, and bone marrow aplasia.[5] Patients with dyskeratosis congenita are at increased risk for progressive bone marrow failure, myelodysplastic syndrome, acute myelogenous leukemia, solid tumors, and pulmonary fibrosis.[3] Ocular manifestations of dyskeratosis congenital include obstruction of the lacrimal drainage system, entropion, keratoconjunctivitis, and retina abnormalities.[5] Many retinal abnormalities have been described, including retinal vasculopathy with and without vitreous hemorrhage,[4,6] exudative vitreoretinopathy,[7] proliferative retinopathy,[8] retinal detachment,[5] Coats-like disease,[9] bilateral retinal hemorrhages,[10] retinal pigment epithelial changes, and retinal neovascularization.[5] Our patient had an exudative retinopathy complicated by vitreous hemorrhage, which could have been secondary to neovascularization or other retinal vasculopathies that were exacerbated by her low platelet count.

This case is unusual because the child already underwent extensive ablation of her peripheral retina to treat her ROP and still developed the exudative retinopathy described in dyskeratosis congenita. Diseases of the telomere such as dyskeratosis congenita should be considered when a child presents with peripheral ischemic or telangiectatic retinal disease.

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Fibrin glue–assisted excision of a large recurrent microphthalmic cyst

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Microphthalmic cysts are rare. Although small cysts can be left in situ to promote orbital expansion, large cysts require drainage or surgical excision. Complete surgical excision is notoriously difficult, and incomplete excision may result in cyst reformation. We describe a novel method of using fibrin glue to aid successful complete removal of a large recurrent microphthalmic cyst in a 6-year-old child who previously had multiple drainage and surgical attempts.

Case Report

A microphthalmic right eye was noted at birth in an otherwise healthy boy. Ocular ultrasound confirmed the diagnosis and that the eye had no visual potential. To

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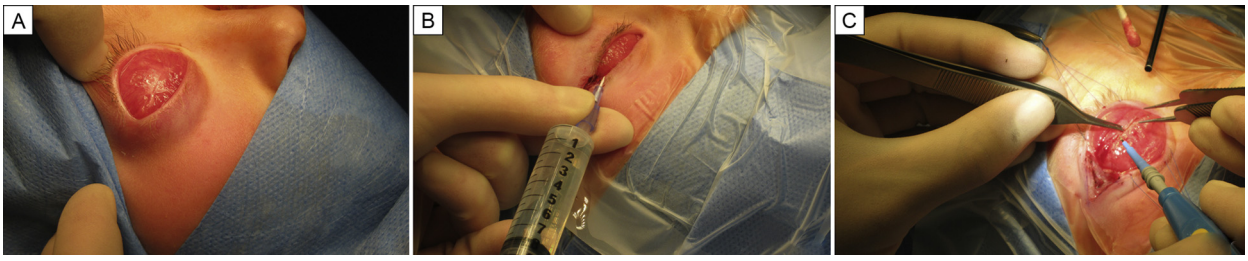

FIG 1. A, Preoperative right proptosis and orbital volume. B, Aspiration of full volume of cystic contents (8 mL). C, Stay sutures in place, start of cyst dissection.

promote socket expansion he was treated with gel expanders and ocular prosthesis. At 4 years of age a dermis fat graft was surgically placed over the microphthalmic eye to provide long-term socket volume.

A year later he presented to the Royal Hallamshire Hospital, Sheffield, with socket fullness, presumed to be from dermis fat growth, and proptosis, causing discomfort. Surgical debulking of the dermis fat graft was undertaken, but intraoperatively fluid leakage was noted. The artificial eye initially fitted well, but within 2 months the proptosis recurred. Ultrasound imaging suggested a cystic orbital lesion, and magnetic resonance imaging confirmed a large cyst associated with the microphthalmic eye, located under the dermis fat graft.

The cyst was drained of 8 mL of straw-colored fluid. Response to the procedure was initially good, but within a few months the cyst was refilling and proptosis recurred. A multidisciplinary team decided to attempt drainage and use fibrin glue to collapse and scar the cyst. Under computed tomography guidance, the cyst was drained, and a contrast agent was injected into the cyst that demonstrated the large size of the cyst and confirmed that there was no intracranial extension. Fibrin glue (2 mL) was injected into the cyst to encourage cyst adhesion. Unfortunately, within 2 weeks the cyst had completely reformed and proptosis recurred.

Given our previous experience in using fibrin glue to excise completely a lacrimal cyst,[1] we decided to attempt using fibrin glue as a scaffold to excise this microphthalmic cyst. Figure 1A shows the extent of preoperative proptosis and orbital volume prior to drainage and fibrin glue scaffold. Under general anesthesia, a lateral canthotomy was performed to gain adequate access to the cyst. The cyst was completely aspirated (8 mL) using a 22-gauge intravenous cannula (the sharp needle was removed after entry to provide a soft tip) attached to a 10 mL syringe (Figure 1B). The cyst cavity was then refilled with about the same volume (8 mL) of fibrin glue, keeping the cannula in situ. The cannula was then removed and the entry point closed with a cotton bud to allow the fibrin glue to set for 3 minutes. The glue provided a scaffold by adhering to the inner surface of the cyst, giving it a stable volume and defining the covering of the cyst. Afterward 5-0 silk stay sutures were applied to the upper and lower eyelids, providing adequate additional exposure and reducing pressure on the cyst. The midline conjunctiva was opened horizontally and then separated from the underlying dermis fat graft up to all fornices.

The dermis fat graft was dissected off from the anterior surface of the now well-set cyst. Careful dissection around the cyst was then undertaken. The cyst cavity epithelial lining was clearly identifiable against the fibrin glue, providing a well-defined surgical plane. During dissection, even if there was any microtrauma to the cyst lining, the cyst did not collapse and avoided any hindrance to further dissection. The cyst was completely dissected off and separated from the underlying microphthalmic eye, which was left in situ. Conjunctiva and lateral canthus were then closed with 7-0 and 6-0 polyglactin 910 sutures, respectively.

The child had an uneventful recovery, with an excellent surgical result and resolution of proptosis and socket fullness. At the 1-year follow-up there was no evidence of cyst recurrence or proptosis. The child is comfortable with a well-fitting and cosmetically acceptable prosthesis.

Discussion

Microphthalmia is estimated to have an incidence of up to 1.5 per 10,000.[2] Management of microphthalmic eyes involves maximizing and maintaining vision where possible, but where no visual potential exists, the aim is to achieve long-term comfort and cosmesis by promoting socket expansion. Current treatment options for socket expansion include hydrophilic expanders, prosthesis, dermis fat grafts or orbital implants in severe cases.[3]

The association of cysts with microphthalmia or anophthalmia is known but is epidemiologically thought to be uncommon. McLean and colleauges,[4] reviewing orbital cysts associated with microphthalmos and anophthalmos over a 28-year period, found 6 bilateral cases in 34 patients.

Microphthalmic cysts are thought to help promote orbital expansion; hence, small-sized cysts are usually left in place,[3,4] with an artificial eye prosthesis applied over the cyst. Large cysts can cause complications, such as prolapse of prosthesis due to proptosis, excessive socket expansion, and discomfort or pain. In such large cysts, drainage or surgical excision is performed. Drainage of

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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