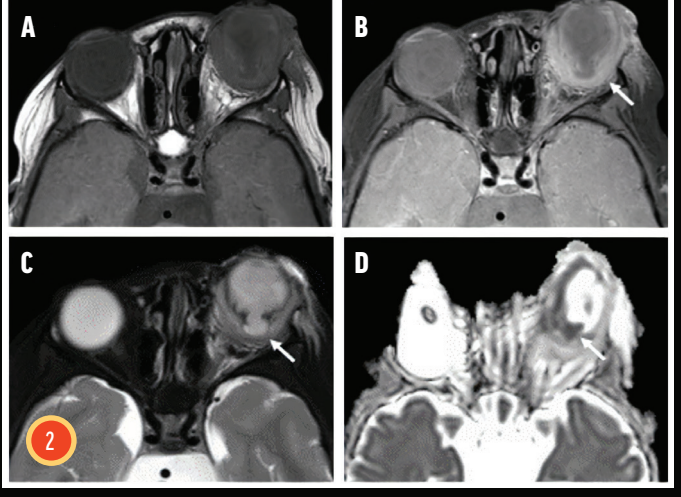

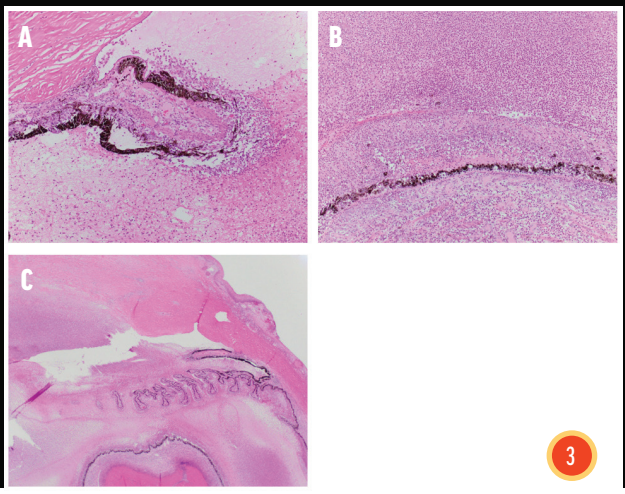

Figure 1. Clinical photograph of the patient upon initial presentation. The left eye demonstrates proptosis, complete corneal opacity, and a central corneal staphyloma with significant conjunctival chemosis and injection with periorbital edema.
Figure 2. An MRI of the orbits demonstrates abnormal soft tissue surrounding the left globe and infiltrating the retrobulbar fat (A). The abnormal soft tissue enhanced after the injection of contrast (B, white arrow). Axial T2 weighted images (C) show debris along the choroid and a focal outpouching along the posterior globe (white arrow) that raises concern of uveal prolapse. Diffusion-weighted imaging was performed. Apparent diffusion coefficient images (D) demonstrate hypointensity, indicating restricted diffusion of water molecules in the choroidal effusion, a finding that may indicate purulence.
Figure 3. A microscopic examination of the enucleated left eye demonstrates acute keratitis and necrosis involving the entire anterior and posterior chambers (A); uveal, retinal, and choroidal abscesses (B); and residual rudimentary iris and angle structures with ectropion uveae and neo-descemetization (C). All panels are stained with hematoxylin and eosin.

ENDOPHTHALMITIS IN A PATIENT WITH PRESUMED AXENFELD-RIEGER SYNDROME

The presentation was atypical in this nonverbal toddler.

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A developmentally delayed, nonverbal 3-year-old girl with severe anterior segment dysgenesis (ASD) presented to the emergency department with a 1-day history of redness in the left eye. A diagnosis of conjunctivitis was rendered, and the patient was discharged with moxifloxacin eye drops. She returned 1 day later with worsening periocular edema, conjunctival injection, chemosis, proptosis, and corneal exposure due to an inability to close the eye (Figure 1). Her parents reported no trauma or systemic complaints, but they stated that the patient had a cold earlier that week and appeared more agitated than usual.

PATIENT HISTORY

The patient was born with completely opacified corneas that had central areas of anterior staphyloma. Her sister was born with a similar corneal presentation. An examination of the mother's eyes found a traditional presentation of Axenfeld-Rieger syndrome (ARS), clear corneas, and no glaucoma.

The patient's exam was stable during a routine visit with the glaucoma service at another institution 5 months earlier. Routine B-scan ultrasound at that visit showed no retinal detachments or acute issues. Her IOP was stable at 15 mm Hg OU on a regimen of twice-daily timolol and latanoprost at bedtime.

DIAGNOSTIC IMAGING AND LABWORK

A computed tomography scan performed at the emergency department showed left-sided proptosis with preseptal and retro-orbital soft tissue fat stranding, scleral thickening, and choroidal enhancement but no significant sinusitis or orbital abscess. The patient was admitted for possible orbital cellulitis, and therapy with intravenous ampicillin-sulbactam and vancomycin was initiated. A laboratory workup showed a white blood cell (WBC) count of 26,700 per µL and C-reactive protein (CRP) of 4.41 mg/dL. A repeat computed tomography scan showed increased superficial inflammation in the soft tissue and no change in the appearance of the left globe.

The following morning, B-scan ultrasonography revealed a retinal detachment with disorganized ocular contents. Blood cultures and adenovirus testing were both negative; conjunctival cultures grew *Cutibacterium acnes* after a few days. The ampicillin-sulbactam was replaced with intravenous ceftriaxone and metronidazole, and therapy with dexamethasone was started at a dosage of 1 mg/kg every 8 hours. Erythromycin ophthalmic ointment was applied to the eye, and the eyelid was closed with Tegaderm (3M) to prevent further exposure of the globe. The differential diagnosis at this time included occult rupture with endophthalmitis or a severe inflammatory reaction.

FURTHER EXAMINATION

On the following day, the patient was transferred to the institution where she receives routine care. During an examination under anesthesia, the eyelids were erythematous and edematous, the globe was proptotic and exposed, the conjunctiva was deeply injected and chemotic, and the cornea was opaque. The patient did not appear to respond to light in either eye. The IOP was 45 mm Hg OD and 80 mm Hg OS. While she was sedated, repeat B-scan ultrasonography was performed and showed stable findings.

A tiny corneal ulcer without significant thinning was found near the limbus of the left eye that might have been related to corneal exposure. An MRI showed a stretched globe with multiple areas of outpouching and scleral thinning (Figure 2). Although a vitreous tap and an injection of vancomycin and ceftazidime were performed, the patient's systemic symptoms continued to worsen, and the WBC and CRP increased. A repeat MRI demonstrated posterior scleral thinning/staphylomatous changes and outpouchings that were consistent with endophthalmitis.

SURGICAL COURSE AND OUTCOME

Because the infectious source could not be controlled, an enucleation was performed, and a peripherally inserted central catheter was placed. A gross pathologic examination found a fragmented globe due to severe inflammatory adhesions present at surgery. Histopathology revealed a purulent inflammatory infiltrate, including acute keratitis and necrosis (Figure 3).

Postoperatively, the patient experienced a marked improvement in her symptoms, WBC, and CRP, and she resumed normal activities. Netarsudil 0.02% was added to the IOP-lowering drug regimen for the right eye. A repeat examination under anesthesia found an IOP of 20 to 22 mm Hg OD. Ultrasound biomicroscopy of the eye showed the iris to be plastered against the cornea and areas of ciliary body detachment, which would make the placement of a tube shunt unsafe. Extreme scleral thinning, moreover, was a contraindication for diode cyclophotocoagulation.

The patient is currently stable on the aforementioned conservative medical management.

DISCUSSION

The cause of the patient's endophthalmitis remains unknown. Based on her presentation, imaging, and pathology, a full-thickness injury to the abnormal cornea that led to a perforation and secondary endophthalmitis is the most likely explanation. The lack of an obvious laceration, however, raises the question of other possible causes.

The differential diagnosis includes ASD-associated uncontrolled glaucoma leading to scleral perforation and subsequent endophthalmitis, unwitnessed trauma with corneal perforation, and sterile phacoantigenic endophthalmitis. Spontaneous globe perforation due to scleral thinning secondary to chronically elevated IOP has been reported in patients with Marfan syndrome,[1] and trauma is common in children.[2,3] Although no history of trauma was provided, the patient is a young child with compromised vision, so trauma cannot be ruled out. Disorganization of the posterior chamber as typically seen in a globe rupture supports these two hypotheses.

Phacoantigenic endophthalmitis does not usually lead to such a profound orbital reaction but is included in the differential diagnosis because of the florid inflammatory granulomatous reaction surrounding the disrupted lens.

This case illustrates an uncommon manifestation of ASD/presumed ARS with a devastating outcome. The patient was diagnosed with congenital glaucoma; most cases of ARS-associated glaucoma develop in childhood or early adulthood. Further, although corneal thickening is common in ARS, total congenital opacification, as seen in this patient, is rare. ARS-associated glaucoma and buphthalmos are difficult to treat. Medication alone failed to reduce the patient's IOP owing to severe dysgenesis of the angle structures, which might have made her more susceptible to

corneal injury. There are few reports on the surgical management of ARS-glaucoma; multiple surgeries are usually required, and complications are common.[4,5] One case series found primary combined trabeculotomy and trabeculectomy to be safe and effective for controlling IOP while maintaining a satisfactory visual outcome in ARS-associated early-onset glaucoma (< 3 years of age).[6] Children who have less severe congenital abnormalities, however, usually achieve better outcomes.

In summary, published research on the optimal treatment of rare manifestations of ASD due to phenotypic heterogeneity is limited. Appropriate management of ASD exacerbations is necessary to preserve native eye structures and maximize visual potential. ■

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