both EyeWiki and AAO. Given the highly technical nature of the content published by EyeWiki and AAO, however, Wikipedia may be a more approachable yet still reliable source of information for patients and parents. Finally, the higher-scoring online sources could serve as a guide for further content development of other sources.

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Orbital conjunctival epithelial cyst mimicking cyst with microphthalmos

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Orbitopalpebral cysts are most commonly associated with microphthalmic eyes (microphthalmos with cyst). We report a 15-year-old girl with a large orbitopalpebral cyst in the absence of associated microphthalmos. The patient presented with a massive swelling involving the left lower eyelid overhanging the left cheek and occupying much of the left orbital cavity. The globe was not visible on clinical examination. No history of trauma or surgery preceding the development of the cyst was reported by the patient or her father. Imaging showed a small eyeball displaced superiorly along the anterior part of the orbital roof. On excision of the cyst, a normal-sized globe with a cataractous lens was noted. Histopathological examination revealed the cyst to be of conjunctival origin.

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Case Report

A 15-year-old girl presented to the outpatient clinic at the LV Prasad Eye Institute, Bhubaneswar, Odisha, with a massive swelling involving the left lower eyelid. There was no other ocular complaint. The eyelid swelling had been noted by her parents since the girl was about 2 years of age but was initially very small. There was no definite history of birth trauma. Major developmental milestones had been achieved normally and no other ocular or systemic developmental anomalies were detected on clinical examination. Progressive increase in the size of the swelling had been noted from around 4 years of age. No medical advice had been sought until the swelling had progressed to massive dimensions.

On clinical examination, the right eye had an unaided visual acuity of 20/20, and the anterior and posterior segments were unremarkable. On the left side, the swelling involved the left lower eyelid and overhung the left cheek, extending to the lateral edge of the ala of the nose (Figure 1A). The inferior fornix was stretched and completely exposed, showing signs of early keratinization. The swelling felt tense but was appreciated to be cystic. The overlying skin was indurated and covered with multiple superficial crusts giving it a leopard-spotted appearance. The left globe was not visible clinically. Computed tomography (CT) revealed a massive orbitopalpebral cyst

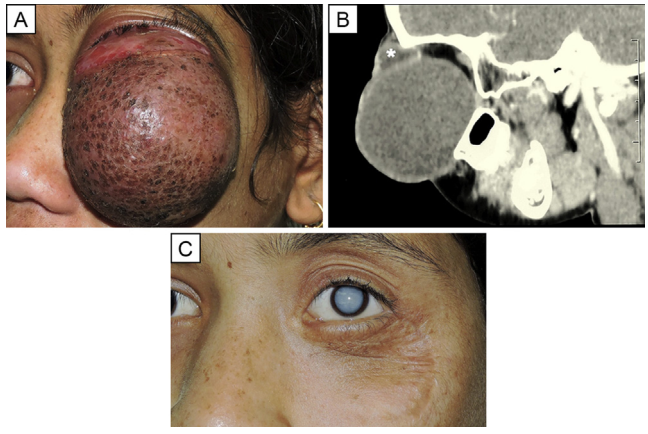

FIG 1. A, Clinical photograph showing a massive cystic swelling of the left lower eyelid overhanging the cheek. B, Computed tomography showing a large orbitopalpebral cyst with a superiorly compressed eyeball (asterisk). C, At 1-year's follow-up there was no recurrence of the cyst.

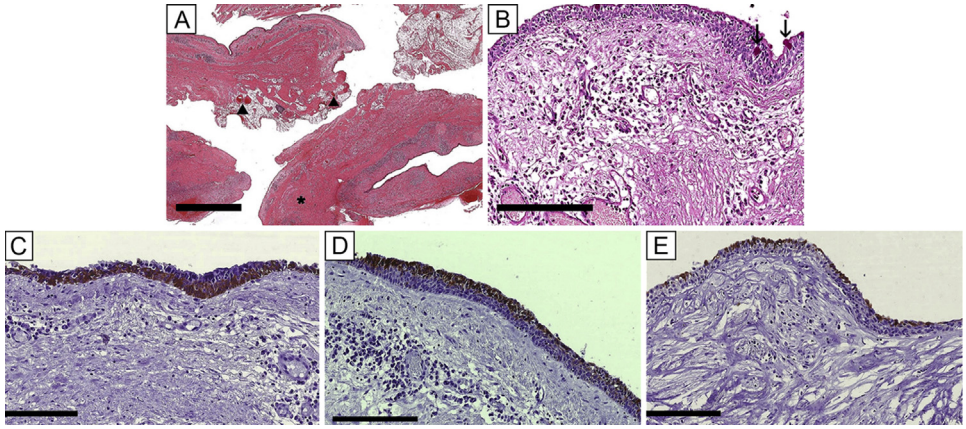

FIG 2. A, Histopathology revealed that the cyst wall contained fibromuscular to fibrofatty tissue (asterisk), with patchy aggregates of chronic lymphomononuclear and plasma cells and reactive lymphoid follicles; congested thin-walled dilated blood vessels (arrowheads) were also observed (hematoxylin-eosin, original magnification ×0.7 [bar = 3 mm]). B, Lining epithelium also showed dispersed goblet cells (arrows; periodic acid-Schiff, original magnification ×15.6 [bar = 200 μm]). C-E, Immunohistochemical staining showing an expression of CK5/14 in the basal cell layers (C) and CK7 in the superficial cell layers (D) of the epithelium; CK18 showed patchy expression in the goblet cells and superficial squamous metaplastic cells (E). Findings suggested characteristics of conjunctival lining epithelium (original magnification ×20 [bar = 200 μm] for all three images).

Table 1. Features of congenital cystic eye compared with microphthalmos with cyst and cyst without microphthalmos

Characteristic	Congenital cystic eye	Cyst with microphthalmos	Conjunctival epithelial cyst mimicking cyst with microphthalmos (current case)
Origin	Developmental	Developmental	Unknown
Embryonic stage of insult	2–7 mm	7–14 mm	Unknown, may not be developmental
Eyeball status	Absent	Microphthalmos	Normal
Cyst location within orbital cavity	More commonly central, less often upper eyelid	More commonly lower eyelid, attached to inferior part of the microphthalmic eyeball	Lower eyelid, no attachment to the eyeball
Histopathology	Primitive neuroglial tissue lining cystic cavity, complete absence of ectodermal elements	Inner neuroglial and outer fibrovascular layers lining cystic cavity, small globe with essentially normal structures	Conjunctival epithelium lined cystic cavity, normal sized globe with essentially normal structures

occupying nearly the entire left orbital cavity and a small globe abutting the superior orbital rim anteriorly (Figure 1B) giving the impression of microphthalmia. However, no evidence of any communication between the globe and the cyst could be identified on imaging. No bony erosion was evident on the CT scan. On the other hand, the inferolateral orbital bones had expanded because of pressure.

Cyst aspiration yielded 108 ml of clear straw-colored fluid and revealed an apparently normal-sized globe with a cataractous lens. The cyst was too large for effective sclerotherapy, and an excision was performed. The cyst wall was significantly thickened and fibrous but was easily separable from the eyeball at surgery. Postoperatively, the axial length of the left eyeball measured 21.06 mm and the structural details were found to be normal on B-scan ultrasonography. Microscopic examination of the excised lesion revealed cystic fragments lined by nonkeratinized stratified columnar to squamous epithelium with interspersed goblet cells. The wall comprised fibrous, fibromuscular to fibrofatty tissue with interspersed congested thin-walled dilated vessels (Figure 2A). Chronic lymphocytic mononuclear and plasma cell infiltration was noted as was the presence of reactive lymphoid follicles. The epithelial lining also showed dispersed goblet cells (Figure 2B). This suggested an origin of the cyst from conjunctival epithelial tissue. Immunohistochemical staining demonstrated expression of CK5/14 in the basal cell layers (Figure 2C) and CK7 in the superficial cell layers (Figure 2D) of the epithelium. CK18 showed patchy expression in the goblet cells and superficial squamous metaplastic cells (Figure 2E). CK17 did not show any expression. Morphological and immunohistochemical correlation thus confirmed the conjunctival origin of the cyst.

The patient's postoperative recovery was uneventful. One year after surgery there was no recurrence of the cyst (Figure 1C). On examination, visual acuity of the left eye was hand motions, with a briskly reactive pupil and no afferent pupillary defect. The

patient underwent uneventful cataract surgery, with anticipated dense deprivational amblyopia, but was lost to follow-up.

Discussion

Orbitopalpebral cysts have been classically described in association with microphthalmos or anophthalmos.[1] They are rare and account for 2% of orbital cystic lesions and <1% of orbital biopsies.[2] These cysts are more commonly unilateral than bilateral.[3] They may occur in isolation or in association with other developmental ocular or systemic anomalies.[3,4] Histopathologically they lack a lining epithelium and must be differentiated from other cystic lesions of the orbit without an epithelial lining, such as a cystic eye, microphthalmia with cystic teratoma, meningoencephaloceles, and ectopic brain tissue.[5] Of these, a cystic eye is likely to be a close masquerader.

Development of a cystic eye is believed to stem from the failure of invagination of the primary optic vesicle at the 2–7 mm stage of the embryo leading to a fluid-filled cavity lined by primitive neuroglial tissue and the absence of all other ocular structures that develop from the surface ectoderm.[5,6] Development of the cyst associated with microphthalmos arises from the failure of the fetal fissure to close at the 7–14 mm embryotic stage. In this case, evidence of development of ocular structures is usually present albeit with smaller dimensions, resulting in a microphthalmic globe.[5,6]

The cyst in the present case resembled neither of these entities histopathologically; rather, it mimicked a typical cyst with microphthalmos clinically and radiologically, although it apparently arose from conjunctival tissue. The presence of a thick fibrous cyst wall and evidence of chronic inflammation may suggest unidentified remote birth trauma or chronic infection or inflammation of undetermined cause, but the pathogenesis of the lesion remains unknown. Table 1 compares the findings in this case with typical features of congenital cystic eye and the cyst with microphthalmos reported in the literature.[5-8]

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Neonatal bilateral acute retinal necrosis in a neonate with a history of severe intrauterine growth restriction

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We present the case of a baby girl born at term with severe intrauterine growth restriction (IUGR) to a gravida 1 mother who was previously healthy and HIV negative. The newborn was evaluated by an ophthalmologist because of her history of IUGR and was diagnosed with intraretinal hemorrhages associated with areas of peripheral retinal necrosis at the posterior pole of both eyes. A diagnosis of acute retinal necrosis of presumed viral origin due to cytomegalovirus virus was considered, and the infant was started on and responded well to valganciclovir.

Acute retinal necrosis (ARN) is a retinopathy of viral etiology that is characterized by 360° of peripheral retinal necrosis, occlusive vasculitis, and vitritis.[1] It can compromise both retinal arteries and veins and cause optic neuropathy and retinal detachment. The annual incidence is estimated at 1 case per 2 million individuals,[2,3] occurring more frequently in adults than children; neonatal cases are exceptional.[4] It affects both sexes and any ethnicity or age group; it occurs in both immunocompetent and immunosuppressed patients.[2] Its etiology is mainly viral and can involve the Herpesviridae family, such as herpes simplex virus type 1 (HSV-1) and herpes simplex virus type 2 (HSV-2) varicella zoster virus (VZV), cytomegalovirus (CMV) or Epstein–Barr virus.[5]

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