margins and preserve healthy tissues; however, owing to the propensity for skin lesions (intraepithelial neoplasia) in sebaceous carcinoma, microscopic intraepithelial neoplasia may be present even with negative margins and may lead to local recurrences. It is interesting that our patient developed a late regional nodal metastasis without any evidence of local recurrence. One plausible explanation is that the primary sebaceous carcinoma in the caruncle was a very low-grade, slow-growing carcinoma that gained access to the lymphatic channels at the time of original diagnosis and before surgical removal of the ocular tumor and, because of its slow-growing nature, it took 11 years for the nodal metastasis to reach a size to be detectable on palpation by the patient.

Typical follow-up for a patient with periocular sebaceous carcinoma includes serial examination of the ocular surgical site, palpation of the regional lymph nodes, and imaging (eg, ultrasonography or computed tomography) of the regional lymph nodes for 5 years after resection of the primary tumor. Although it is unrealistic, on the basis of this single case, to recommend surveillance of patients with periocular sebaceous carcinoma beyond 5 years, it is important for clinicians and patients to be aware of the potential for late nodal metastasis of sebaceous carcinoma beyond the initial 5 years after diagnosis and treatment of the ocular tumor. Patients with ocular and periocular sebaceous carcinoma should be educated regarding the possible but rare incidence of late relapse in the regional lymph nodes and the location of lymph nodes at risk in the parotid and submandibular regions.

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Congenital Cystic Eye In Utero: Novel Prenatal Magnetic Resonance Imaging Findings

First described in 1939,1-3 congenital cystic eye is an exceedingly rare orbital malformation due to failure of optic vesicle invagination during embryogenesis.1 Approximately 30 to 40 cases have been reported. The malformation consists of anophthalmic orbit containing a fluid-filled cyst and, frequently, rudimentary ocular derivatives.2 Concomitant nonocular malformations including intracranial anomalies and systemic malformations are often found in association.1 Discovery is typically after birth. Diagnosis is based on clinical, imaging, and histopathological characteristics. Generally, surgical excision is performed, ameliorating the expanse cyst and facial dysmorphism.

Herein, we report a case of congenital cystic eye with left frontal dysplasia, colpocephaly, and agenesis of the corpus callosum and septum pellucidum discovered in utero via ultrasonography. Prenatal and postnatal magnetic resonance imaging (MRI) characteristics, clinical and histopathological findings, and surgical management are discussed. To our knowledge, no previous cases of congenital cystic eye observed via prenatal MRI have been reported to date.

Report of a Case | Prenatal ultrasonography of a 28-year-old woman at 23 weeks’ gestation disclosed a heterogeneous orbital mass. A noncontrasted abdominal MRI at 26 weeks’ gestation revealed a large left orbital cystic mass and agenesis of the corpus callosum and septum pellucidum. The mass was generally hypointense (dark) on T1-weighted images and hyperintense (bright) on T2-weighted images, similar to cerebrospinal fluid (Figure 1). Posteriorly, tissue with mixed signal intensity was observed on T2-weighted images, consistent with internal septations. No significant solid component or globe was identified.

The cyst did not significantly enlarge during the pregnancy. Examination of the female child revealed a protruding purplish orbital mass (Figure 2A), which transilluminated homogeneously. The eyelids were separated by normal-appearing conjunctiva. Palpation disclosed a spongelike consistency and no apparent globe.

Orbital MRI with and without contrast and brain MRI without contrast were obtained after birth (Figure 2B). The cyst was generally hypointense on T1-weighted images and hyperintense on T2-weighted images, similar to cerebrospinal fluid. The internal septations enhanced heterogeneously with gadolinium. Extraocular muscle was present inferiorly. Optic nerve merged with the posterior aspect of the cyst, but no globe was present. Absent corpus callosum and septum pellucidum, colpocephaly, left frontal lobe dysplasia, and a right dacryocystocele were also noted. Karyotype analysis in the form of an
oligonucleotide array comparative genome hybridization (DNAarray Oligo 180K) revealed no genetic aberrations (including SOX2 and PAX6 mutations).

Needle decompression and tarsorrhaphy were performed on day 5 after birth. The cyst revolumized despite multiple aspirations. Given the recalcitrant nature, surgery was planned. Excision with silicone implant placement was performed at age 1 month. Posteriorly, rudimentary muscle cone and hypoplastic optic nerve were identified. At age 1 year, a prosthesis was fashioned (Figure 2C).

Histopathological analysis of the cyst revealed a multiloculated structure predominantly composed of mature neuronal tissue (Figure 3). The walls were composed of connective tissue and the cavities were lined by ependymal epithelium. Foci of pigmented neuroepithelium and neural rosettes resembling primitive retinal pigment epithelium and retina were present, respectively. Skeletal muscle was noted at the periphery.

Discussion | Congenital cystic eye is the abortive product of partial or complete arrest in optic vesicle invagination during week 4 of embryogenesis.1,2 Total arrest between the 2- and 7-mm stages of embryogenesis results in the complete absence of surface ectodermal structures and dysplasia of neuroectodermal tissue.1,2 It is postulated that partial arrest results in abortive ectodermal or neuroectodermal products, as reports of rudimentary lens, iris, and cornea exist.1,2 Failure between the 7- and 14-mm stages results in the more common condition of microphthalmia with cyst.1,4

The etiology is unknown1 but is thought to be nonhereditary.1 The condition may be secondary to a maternal insult, but most cases are not associated with such an event.1,5 Interestingly, our patient’s mother had a self-limited prenatal fever but the timing (9 weeks’ gestation) was not coincident with the critical fourth week of embryogenesis. Results of chromosomal studies have been reportedly normal,2,5 as they were in our case.
The distinguishing feature of congenital cystic eye is an anophthalmic orbit associated with a cyst. Clinical findings are varied, but often distention of the upper eyelid with purplish discoloration is found. This is in contrast to microphthalmia with cyst, which typically distends the lower eyelid.\(^1,2,4\) The condition is usually unilateral, but bilateral cases exist.\(^1,2\) Congenital cystic eyes are usually fluid filled but can be solid.\(^1\) The cyst volume may be related to patency of a posterior stalk extending into the cranial cavity.\(^4\) Static cysts are postulated to be nonexpansile owing to this communication.\(^4\) Coexistent ocular and nonocular malformations are frequently found with congenital cystic eye,\(^1,5\) in particular, intracranial abnormalities such as midline deformities.\(^1,4,5\)

Despite that a unifying diagnosis for this uncommon collection of malformations does not exist, Pasquale et al\(^5\) suggested that the designation cranial ectodermopathy provides a generalized classification for most of these anomalies. We believe a separate consideration is that congenital cystic eye with midline malformations may represent a novel presentation in the septo-optic dysplasia sequence.

Neuroimaging is of marked utility in diagnosing this condition. Magnetic resonance imaging reveals a cystic mass that variably contains septations and solid components.\(^3,4\) The main cystic cavity is well defined, is homogeneous, and demonstrates low signal intensity on T1-weighted images and high signal intensity on T2-weighted images, similar to cerebrospinal fluid.\(^1,3,4\) Septations are isointense to muscle on T1-weighted images and heterogeneously hyperintense on T2-weighted images.\(^3\) Gadolinium may enhance the septations and solid tissue, greater than brain parenchyma.\(^3\) Vestigial extraocular muscle and optic nerve are variably observed.\(^3\) An orbital cyst with anophthalmia is noted, which uniquely distinguishes this entity from other diagnoses, including microphthalmia with cyst.

A varied histopathological appearance is noted owing to incomplete development of neuroectodermal elements. Robb and Anthony\(^6\) suggest that the malformation contains 5 principal elements: (1) central unilocular fluid-filled cavity, (2) neuroglial layer sometimes lined by nonpigmented epithelium, (3) foci of retinal pigment epithelium, (4) dense connective tissue.

![Hematoxylin-Eosin-Stained Histopathological Sections](image-url)

A, The cystic eye is composed primarily of neuroglial tissue. Connective tissue forms the walls, and the inner layer is lined by ependymal epithelium (original magnification ×2). B, Microvilli on the surface of the ependymal epithelium (original magnification ×60). C, Scattered neural rosettes resembling primitive retinal elements (original magnification ×20). D, Foci of pigmented neuroepithelium, similar to retinal pigment epithelium (original magnification ×20).
sue in concentric layers, and (5) external loose connective tissue with fibroadipose elements, muscle, and blood vessels. Our patient had each of these elements.

Conservative treatment includes observation with aspiration as needed. In recalcitrant cases not amenable to aspiration or if considerable facial dysmorphism exists, excision is conventionally undertaken.

In summary, congenital cystic eye is exceedingly rare. Diagnosis historically was based on physical and histopathological findings. However, newer imaging modalities are revealing characteristic findings of the condition at or even prior to birth, as in our case. Given the frequent association with intracranial abnormalities, including the possibility of septo-optic dysplasia, neuroimaging is warranted to screen for such aberrations. Finally, although not always required, treatment conventionally involves excision.

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Development of a Premacular Vitreous Pocket

The premacular vitreous pocket (PVP), or vitreoschisis cavity, is a liquefied vitreous cavity in front of the posterior retina that is characteristic of various macular diseases, including macular holes and diabetic maculopathy. The reason for the development of PVPs is unknown because of the difficulty observing the formed vitreous in vivo. India ink and the fluorescein staining technique have delineated the structure of the PVP in the vitreous cavity in human eyes at autopsy; however, the technique is limited because of the presence of artifacts during fixation of the fragile and mobile vitreous and postmortem changes. Optical coherence tomography has facilitated observation of the vitreous structures in vivo. Herein, we describe the development and fine details of PVPs in real time.

Methods | We retrospectively analyzed the posterior vitreous, retinas, and optic discs of 56 healthy eyes (39 patients; age range, 1-54 years) using swept-source optical coherence tomography (Topcon), which provides detailed images of the fine ocular structures. The scanning protocol used in this study was a single-line scan with 96 overlapping images and a radial scan with 32 overlapping images. Each line has a 12-mm transverse scanning length with 1024-pixel resolution. Eyes that appeared healthy were excluded if the patient had a family history of a hereditary vitreoretinal disease.

Results | A PVP (Figure 1C-F) was detected in all eyes of patients older than 10 years and in no eyes of patients younger than 2 years (Figure 1A). A crack in the formed vitreous (Figure 1B), considered to be a primitive structure of the PVP, developed first in eyes around age 2 years. Between ages 3 and