Keratoacanthoma is a benign tumor that shows spontaneous regression but often resembles well-differentiated squamous cell carcinoma histopathologically. Like squamous cell carcinoma, keratoacanthoma may show cellular atypia, mitosis, horn pearls, and focally infiltrative growth. In the present case, the initial microscopic section did not show a keratotic plug, and the first diagnosis was squamous cell carcinoma. However, additional sections cut from deeper in the tissue block revealed the central crater. Considering this architecture and the clinical course, the pathologic diagnosis was revised to keratoacanthoma. Specifically, features of rapid growth and a central crater are essential to diagnosis of keratoacanthoma.

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An Unusual Case of Leukocoria: Heterotopic Brain Arising From the Retina

A newborn infant was seen who had leukocoria, total retinal detachment, and a noncalcified mass in the posterior retina. Retinoblastoma was considered a possibility, and enucleation was performed. Pathologic examination findings revealed heterotopic brain tissue arising from the retina to lead us to conclude that posterior segment heterotopic brain tissue is a rare choristomatous lesion that can present as leukocoria.

Report of a Case. An 8-day-old white male was noted to have leukocoria in the right eye on routine examination at his pediatrician’s office. He was referred immediately for ophthalmologic examination. There was no family history of retinoblastoma or childhood eye disease.

On examination, the right eye minimally responded to light. The left eye responded normally to light with a brisk blinking reflex. A right afferent pupillary defect was present. Slitlamp examination of the right eye revealed a thick retrolental white plaque (Figure 1). The horizontal corneal diameter OD was 9.5 mm in comparison with 10 mm OS. The lens in the right eye was subluxated anteriorly and superotemporally. Fundus examination of this eye revealed a funnel-shaped retinal detachment with a fibrous membrane spanning the anterior part of the funnel just posterior to the lens. A large chalky...
white mass was visible through the retrolental plaque in the temporal aspect of the eye. This mass appeared to be breaking through the retina in several places. Two-dimensional ultrasound of the right eye demonstrated a total retinal detachment with a posterior noncalcified subretinal mass and a posterior colobomatous defect. The left eye was normal.

A head and orbit computed tomographic scan demonstrated a well-demarcated, noncalcified posterior intraocular mass with a posterior staphyloma in the right eye (Figure 2). Though atypical, retinoblastoma was considered a distinct possibility and enucleation was performed.

Pathologic Findings. On gross examination, the eye measured 19.0 × 17.5 × 17.5 mm with an 8-mm optic nerve. The cornea was globoid and the anterior chamber was formed. The pupil was round, and the lens was subluxated superotemporally. A posterior staphyloma was located between the 10- and 2-o’clock positions near the optic nerve. On transillumination of the sclera, there was a shadow from the 10- to the 3-o’clock position extending from 2 mm from the limbus to 15 mm posterior to the limbus. The eye was opened horizontally. The clear lens was slightly subluxated anteriorly and superotemporally. The retina was completely detached with a large subretinal white-tan, gelatinous, and homogeneous mass (Figure 3). The mass filled the posterior staphyloma without gross extrascleral extension. Obvious necrosis and calcifications were not seen.

Microscopically, the anterior segment was unremarkable with the exception of the subluxated lens. The mass in the posterior pole arose from the inner layers of the retina (Figure 4A) and focally occupied the choroid. The lesion was composed of mature neuroglial tissue with areas closely resembling brain tissue containing oligodendrocytes, neurons, and white matter (Figure 4B). There were some capillaries and foci of focally calcified pigment epithelium (Figure 4C). The choroid and retinal pigment epithelium were disrupted temporally where the staphyloma was identified. Here the lesion was more gliotic and disorganized, filling the choroid. No increased mitotic activity, necrosis, atypia, or increased vascularity was seen. The retina was centrally detached in a funnel shape and contained retinal rosettes and disorganized architecture. In the retrolenticular area, the retina merged with a gliovascular membrane that extended toward the pars plana (Figure 4D). The lesion involved the optic nerve head; however, the optic nerve beyond the lamina cribrosa was free of neuroglial proliferation. The cross section of the optic nerve at the surgical margin demonstrated a defect in the dura with focal invagination of adipose tissue into the subarachnoid space. The final diagnosis was microphthalmos with neuroglial proliferation consistent with brain choristoma (heterotopic brain) and subluxated lens, keratoglobus, posterior staphyloma, and lipomatosis of the optic nerve.

Comment. The collective findings in this case are those of a congenital lesion, most consistent with a brain choristoma. The lesion comprises mature neurons, glial elements, mature vessels, and foci of pigmented epithelium. The presence of oligodendrocytes, which are not part of the normal histologic composition of the retina, and the architecture of the main mass that closely resembles brain tissue are supportive of the clas-
sification of the lesion as a choristoma rather than a hamartoma. Ganglioneuroma, an entity that was included in the differential diagnosis, usually demonstrates disorganized architecture and displays nerve bundles and neurons. In this case, the architectural organization was that of organized neuron layers and of vessels, and pigment epithelium. Grossly, the consistency and color of the tissue were more consistent with brain tissue than ganglioneuroma.

Hamartomatous lesions of the optic nerve head have been described as containing disorganized neuroglial tissue admixed with vessels and hyperplasia of retinal pigment epithelium. Neuroglial hamartomas of the choroid are disorganized neuroglial proliferations occupying the anterior choroid. The presence of microphthalmos, subluxated lens, keratoglobus, posterior staphyloma with probable choroidal coloboma, and lipomatosis of the optic nerve with changes consistent with coloboma of the optic nerve suggests that this lesion is part of an embryologic defect probably similar to that occurring in microphthalmos with cyst but with milder changes. The wall of a colobomatous cyst consists of thin sclera lined by rudimentary tissue of neuroectodermal origin occasionally incorporating hyperplastic glial tissue. Like other colobomatous malformations, microphthalmos with cyst may occur either as an isolated congenital defect or in association with a variety of intracranial or systemic anomalies. Our patient did not have evidence of other intracranial or systemic anomalies or phakomatoses, though he was noted to be developmentally delayed at his most recent follow-up evaluation.

Heterotopic brain or brain choristoma is an unusual lesion described in the head and neck region that manifests no demonstrable connection to the cranial vault and does not demonstrate behavior of a true neoplasm. Histologically, it contains mature glial elements and sometimes contains ependyma and choroid plexus-like structures. Choristomatic brain tissue found in the oropharynx or nasopharynx may cause airway obstruction. Ectopic brain tissue in the orbit is rare, and only a few reports of this have been published in the literature. The proposed mechanisms are (1) herniation of normal embryonic neuroepithelium, subsequent sequestration, and then parallel development; (2) transformation of dysplastic neural rests with aberrant differentiation potential, or (3) preferential development of 1 germ layer in a teratoma.

Figure 4. A, Photomicrograph of the retina overlying the lesion (arrow) shows disorganization of the outer layers from where the proliferation of the neuroglial lesion (NGL) appears to arise. Notice the striking resemblance of the NGL to normal brain histologic features (hematoxylin-eosin, original magnification ×10). B, Higher magnification of part A shows oligodendrocytes (black arrow) admixed with neurons (white arrow) (hematoxylin-eosin, original magnification ×40). C, Foci of retinal pigment epithelium (arrow) and calcifications (arrowhead) were also found in the central and posterior areas of the NGL (hematoxylin-eosin, original magnification ×40). D, The retrolental membrane comprised a disorganized glial membrane (GM) that was attached to the pars plana of the ciliary body and pulled anteriorly toward the pars plicata (left to the glial membrane) (hematoxylin-eosin, original magnification ×10).
To summarize, this is the first reported case, to our knowledge, of choristomatous brain tissue in the retina. In this case, because the eye was blind and the diagnosis was in question, the most prudent method of management was to proceed with enucleation. Heterotopic brain choristoma, though extremely rare, should be added to the expanding differential diagnosis of leukocoria.

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Ocular Hypotonus Secondary to Spontaneously Ruptured Posterior Staphyloma

We describe a woman with ocular hypotonus secondary to a spontaneously ruptured posterior staphyloma. The staphyloma was effectively treated with a retrobulbar injection of autologous blood. A high-resolution 3-T magnetic resonance imaging (MRI) scan was useful in demonstrating retrobulbar fluid. We hypothesize that the injection of retrobulbar blood prompted the formation of a fibrotic scar that sealed the filtering site.

Report of a Case. A 39-year-old woman with high myopia was referred for further management of hypotonus of the right eye that had been present for 1 month. Her ocular history was significant for high myopia treated with bilateral radial keratotomy 10-years prior to this episode and a choroidal neovascular membrane in the right eye that was treated with photocoagulation 4 years prior to this episode.

The patient’s current episode involved a history of acute loss of peripheral vision in the right eye while straining at the stool. She reported no pain with this episode but stated that her eye subjectively felt “soft and squishy.” An examination was performed 2 days later by an ophthalmologist who found the intraocular pressure (IOP) to be 2 mm Hg OD. No leaks were found in the keratotomy scars. Other than the submacular scar, the examination findings were normal. There was no history of retrobulbar injection. The hypotonus with blurring persisted prompting her to seek care by another ophthalmologist. He diagnosed presumed leaks in her keratotomy scars. Sutures were placed in these scars, but the hypotonus and blurred vision persisted despite no identifiable anterior leaks. The patient also reported an intermittently present “pocket of fluid” in the right inferior conjunctival cul-de-sac that persisted after the suturing.

Our examination recorded an uncorrected visual acuity of 4/400 OD and 20/40 OS. Intraocular pressures by application tonometry were 4 mm Hg OD and 12 mm Hg OS. The right pupil was pharmacologically fixed and dilated without an afferent pupillary defect. Confrontational fields and rotations were normal. Slitlamp examination results were significant for radial corneal scars in both eyes that were negative for leaks by Seidel test with and without mild digital pressure on the globe. Five sutures were in place across 2 of the right cornea scars. The anterior chambers were deep and clear. Gonioscopy in the right eye showed the ciliary body band to be 360° without evidence of a cyclodialysis cleft. Funduscopy examination showed a swollen optic nerve head and choroidal folds consistent with hypotonus. A scar in the right macula consistent with the history of photocoagulation was also noted (Figure 1). Temporal to the right macula was a posterior staphyloma with a full-thickness choroidal defect (Figure 2). The temporal aspect of this defect revealed mobile sclera that would flap or un-