These 2 cases illustrate a complication of DLEK that we feel led to an undesirable visual outcome. Considering the potential causes for this condition, we feel that there is currently a lack of understanding of the ideal donor thickness and donor and host diameters. If irregularities secondary to the manual dissection of the corneal tissues, with resulting disparity in donor-host shape or diameters, are demonstrated to be a factor in future cases, one might argue for intentionally oversizing the recipient bed diameter. Alternatively, automated dissection, such as laser-assisted techniques, may ensure ideal lamellar thickness and size. We also query whether early treatment of future patients with these folds, whether by prolonged gas tamponade or repositioning of the donor button, might improve the postoperative optical performance of the cornea graft.

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**Pineal Cyst Simulating Pinealoblastoma in 11 Children With Retinoblastoma**

It is estimated that approximately 3% of all children with retinoblastoma and 5% to 15% of children with hereditary retinoblastoma will develop an intracranial neuroblastic malignancy, usually a pinealoblastoma (“trilateral retinoblastoma”).1,2 This malignancy typically appears in the first 5 years of life1,3 and is nearly always fatal when the patient has related symptoms.1,2 Therefore, children with retinoblastoma, especially those with hereditary retinoblastoma, are advised to have routine neuroimaging for the first 5 years of life with special attention to the pineal gland region.2,3 We recently encountered several retinoblastoma patients with a pineal cyst simulating pinealoblastoma, causing diagnostic and therapeutic confusion. For this reason, we reviewed our experience with this condition.

**Methods.** The medical records of 1400 patients with retinoblastoma managed at the Ocular Oncology Service (C.L.S., J.A.S.) of Wills Eye Hospital, Philadelphia, Pa, between 1975 and 2004 were retrospectively reviewed. Those patients recorded to have a pineal cyst on neuroimaging were identified and included in this study.

**Results.** Table 1 lists the clinical data of the patients. A pineal cyst was identified in 11 patients at a mean age of 4.5 years (median, 3 years; range, 6 months to 16 years). The mean age of the patient at the time of diagnosis of retinoblastoma was 13 months (median, 14 months; range, 0.35-30 months). All 11 patients were white, and 2 were male whereas 9 were female. The retinoblastoma was unilateral in 3 patients, bilateral in 8 patients, and trilateral in 0 patients. Family history for retinoblastoma was positive in 2 patients and negative in 9 patients. Of the 11 patients, 2 had bilateral familial retinoblastoma, 6 had bilateral sporadic retinoblastoma, and 3 had unilateral sporadic retinoblastoma.

**Table 1. Demographics and Tumor Features in 11 Retinoblastoma Patients With Pinal Cyst on Brain Scan**

<table>
<thead>
<tr>
<th>No./Sex/Age at Diagnosis of Retinoblastoma, mo</th>
<th>Race</th>
<th>Laterality Retinoblastoma (Eye)</th>
<th>Heredity Retinoblastoma</th>
<th>Age at Diagnosis of Pineal Cyst</th>
</tr>
</thead>
<tbody>
<tr>
<td>1/F/0.35</td>
<td>White</td>
<td>Bilateral (OU)</td>
<td>Familial</td>
<td>6 mo</td>
</tr>
<tr>
<td>2/F/4</td>
<td>White</td>
<td>Bilateral (OU)</td>
<td>Familial</td>
<td>14.5 mo</td>
</tr>
<tr>
<td>3/M/4</td>
<td>White</td>
<td>Unilateral (OD)</td>
<td>Sporadic</td>
<td>16 y</td>
</tr>
<tr>
<td>4/M/4.5</td>
<td>White</td>
<td>Unilateral (OS)</td>
<td>Sporadic</td>
<td>8 mo</td>
</tr>
<tr>
<td>5/F/6</td>
<td>White</td>
<td>Bilateral (OU)</td>
<td>Sporadic</td>
<td>16 mo</td>
</tr>
<tr>
<td>6/F/11.5</td>
<td>White</td>
<td>Bilateral (OU)</td>
<td>Sporadic</td>
<td>5 y</td>
</tr>
<tr>
<td>7/F/14</td>
<td>White</td>
<td>Bilateral (OU)</td>
<td>Sporadic</td>
<td>3 y</td>
</tr>
<tr>
<td>8/F/16</td>
<td>White</td>
<td>Bilateral (OU)</td>
<td>Sporadic</td>
<td>3 y</td>
</tr>
<tr>
<td>9/F/20.5</td>
<td>White</td>
<td>Bilateral (OU)</td>
<td>Sporadic</td>
<td>8 y</td>
</tr>
<tr>
<td>10/F/29</td>
<td>White</td>
<td>Bilateral (OU)</td>
<td>Sporadic</td>
<td>4.5 y</td>
</tr>
<tr>
<td>11/F/30</td>
<td>White</td>
<td>Unilateral (OD)</td>
<td>Sporadic</td>
<td>6.5 y</td>
</tr>
</tbody>
</table>
The pineal cyst appeared to be isointense to cerebrospinal fluid on T1-weighted images and displayed increased signal intensity on T2-weighted images (Figure). Sagittal plane images provided optimal visualization of the lesion. The pineal cyst diameter measured less than 10 mm in all cases except 1 case where the largest dimension was 12 mm. No instances of associated hydrocephalus or solid pineal tumor were noted. The range of follow-up time was 11 months to 25 years (mean, 7 years; median, 4 years). At a mean 7-year follow-up period, the pineal cyst was stable in all cases except 1, in which the lesion resolved over a period of 3 years.

**Comment.** Pineal region tumors and associated lesions compose less than 1% of central nervous system lesions. Nonneoplastic lesions such as pineal cysts are relatively common incidental benign lesions whose distinction becomes crucial for the patient for both prognostic and therapeutic reasons. Small (0.5–1 cm) pineal cysts are encountered in 25% to 40% of autopsy cases according to the literature. Current estimation of their prevalence based on MRI series ranges from 1.5% to 4.3% of the normal population. Many mechanisms have been proposed to explain the origin of pineal cysts. One theory suggests that pineal cysts are related to involution of the pineal gland secondary to ischemic glial degeneration. It has also been suggested that larger cysts are formed by coalescence of smaller ones, secondary to either hemorrhagic expansion or hormonal influences, thus explaining their more frequent occurrence in girls. The fact that pineal cysts are relatively rare in young children but are most often discovered in patients younger than 30 years of age suggests that pineal cysts may actually develop in late childhood or thereafter. The relatively young average age of our patients may be largely due to the young age of retinoblastoma patients who routinely undergo follow-up neuroimaging the first 5 years of life.

Despite their benign nature, pineal cysts have been reported to enlarge in size and cause symptoms of increased intracranial pressure, most commonly headache secondary to a mass effect. None of the patients in this series had symptoms, and this was probably related to the size of the cysts being generally less than 1 cm.

The fact that MRI can distinguish benign pineal cysts from pineal neoplasms is of obvious clinical importance. The presence of a small, well-circumscribed, round cystic lesion with smooth margins in the pineal gland, showing slightly increased signal of the cyst wall on T1-weighted images, is characteristic of a pineal cyst. The cyst fluid, best visualized on T2-weighted images, shows bright signal intensity, a feature seen with most cysts in the human body. Although we have been unable to find a previous report of a purely cystic pinealoblastoma, the MRI characteristics of pinealoblastoma differ in many aspects from a pineal cyst. Pinealoblastoma is described as a large, nodular, solid mass that typically shows diffuse enhancement after administration of contrast material. On the contrary, pineal cysts normally show a fine rim of cyst wall enhancement, but the cyst itself does not enhance after administration of contrast material. Fleige et al, however, reported that symptomatic pineal cysts may have an atypical MRI appearance, including large size and nodular, irregular enhancement, that often overlaps with the appearance of pineal neoplasms and may lead to an incorrect diagnosis.

Most of the pineal cysts in this study occurred in patients with bilateral retinoblastoma, who are at risk of developing midline brain tumors. However, the later age of appearance of the pineal cyst, the absence of symptoms, and the neuroradiologic appearance of the lesion made the diagnosis of a pinealoblastoma unlikely. Furthermore, follow-up MRI on a 2- to 4-month basis confirmed the lack of progression.

In conclusion, pineal cysts are infrequent midline intracranial lesions in the young population. They are usually small and asymptomatic and carry a good prognosis. Ophthalmologists should be aware of the presence of this benign lesion when performing routine neu-
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Photodynamic Therapy for Exudative Hamartoma in Tuberous Sclerosis

Tuberous sclerosis is a systemic disorder characterized by hamartomas in multiple organs, commonly including the skin and brain, as well as additional cardiac, renal, pulmonary, and ocular findings. Approximately 50% of cases with tuberous sclerosis have ocular involvement with unilateral or bilateral retinal astrocytic hamartomas. Although most retinal astrocytic hamartomas remain asymptomatic or gradually regress during life, some exceptional cases may develop symptomatic alterations by an enlarged tumor with leakage, macular edema, accumulating lipid exudates, serous retinal detachment, or vitreous hemorrhage. Persisting macular edema and lipid accumulation may cause permanent visual impairment. This article highlights the novel approach with photodynamic therapy.

Report of a Case. A young boy developed multiple petit mal seizures at age 2 years and was diagnosed with tuberous sclerosis based on typical cerebral lesions including subependymal, paraventricular astrocytomas as seen with computed tomography. He later experienced additional symptoms of the disease, including multiple slightly elevated, yellow-red, butterfly-shaped papules distributed on his face (sebaceous adenoma), multiple small nodules in the right kidney, and 2 discrete cardiac calcifications, all of which are tumors typical for tuberous sclerosis.

Annual ophthalmic examinations were initiated when the patient was aged 17 years. By then, his best-corrected visual acuity was 20/25 OD and 20/32 OS. Extensive fundus examination in the right eye disclosed a retinal type 3 hamartoma at the superotemporal arcade and 4 type 1 lesions. The type 3 hamartoma had a typical mulberry appearance and a peripheral semitranslucent rim of approximately 1 disc diameter. Three other astrocytic type 1 hamartomas were present at the inferior arcade of the fundus in the left eye.

At age 22 years, the patient had progressively blurred vision with metamorphopsia in his right eye; this was present for 2 weeks. On examination, his visual acuity was 20/80 OD, and it had remained 20/32 OS. Biomicroscopical analysis of his right eye displayed a normal optic disc with well-perfused retinal vessels. The previously described retinal type 3 hamartoma had remained unchanged. However, one of the type 1 hamartomas localized inferior to the type 3 hamartoma had changed its appearance. This lesion, still showing the characteristics of a retinal type 3 hamartoma, had increased in size and was surrounded by subretinal fluid accumulation with multiple small, whitish dots of lipid exudates extending close to the center of the macula.

Figure. A, Sagittal T1-weighted magnetic resonance image (MRI) without contrast shows a pineal cyst (arrow) of 1.2 cm in diameter in a child with retinoblastoma, which is isointense relative to cerebrospinal fluid (CSF) with a rim slightly hyperintense relative to CSF. B, Axial T2-weighted FLAIR (fluid-attenuated inversion recovery) image shows contents of cyst (arrow) hyperintense relative to CSF. C, Sagittal T1-weighted postgadolinium MRI shows no internal cyst enhancement (arrow).

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