Keratoprosthesis implantation has become a viable treatment option for complex ocular diseases who are at high risk for graft failure. Therefore, allograft rejection does not occur with the keratoprosthesis. Another significant advantage is that the keratoprosthesis maintains a spherical anterior shape. Furthermore, the Boston type I keratoprosthesis, a polymethyl methacrylate device, can be custom made such that it can correct for refractive errors as well as aphakia. These advantages are particularly important in cases of pediatric PK. Immunologic graft rejection in children can be insidious and often eludes parents’ detection. Maintaining a clear visual axis while minimizing the induced astigmatism in the immediate postoperative period may minimize the development of amblyopia in this age group. The goal in cases of significant bilateral congenital corneal opacity is to minimize the amount of time that a child is not seeing clearly until he or she nears the end of the amblyopic age range. Theoretically, keratoprosthesis implantation may provide faster and superior visual results as compared with conventional donor corneal transplantation. The device can possibly be removed and replaced by a PK when the child is out of the amblyopic age range.

Corneal blindness remains the leading cause of pediatric blindness globally. In particular, infectious keratitis is the most common cause of corneal scarring in developing countries. Importantly, in the majority of these cases, the occurrence of corneal opacification is associated with poverty in areas devoid of eye-banking and transplantation-quality corneas. Regrettably, no significant developments in pediatric corneal transplantation have been achieved in recent years, and the results of pediatric PK remain poor even in developed countries. Although the postoperative complications may be severe enough to limit the use of currently available devices, we believe that the keratoprosthesis might have a role in the management of corneal blindness in carefully selected children with complex ocular diseases who are at high risk for graft failure.

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Intravascular B-Cell Lymphoma (Angiotropic Lymphoma) With Choroidal Involvement

Intravascular large B-cell lymphoma is a rare form of extranodal lymphoma characterized by the presence of large lymphoma cells in the lumen of small blood vessels, involving multiple organs. The diagnosis is usually made at the time of autopsy. We report a patient with intravascular large B-cell lymphoma who had vision loss from choroidal involvement, which improved with systemic therapy.

Report of a Case. A 44-year-old white man was initially seen with bilateral...
blurred vision, Coombs-negative hemolytic anemia, and a maculopapular skin rash on both upper arms. The patient’s best-corrected visual acuity (Snellen) was OD 20/40 −2 and OS 20/50. There were bilateral serous detachments involving the macula (Figure 1A). A small, superficial intraretinal hemorrhage was noted along the inferotemporal vascular arcade in the right eye, and peripheral flame-shaped intraretinal hemorrhages were present in both eyes. Fluorescein and indocyanine green angiography demonstrated irregular filling of the choroid early with bilateral pinpoint areas of fluorescence later (1 day) (Figure 1C). B-scan ultrasonography showed thickening of the choroid bilaterally (Figure 1B).

The patient had a normocytic anemia with an increased reticulocyte count, lactate dehydrogenase level, and bilirubin concentration. A computed tomographic scan of his chest and abdomen showed hazy ground-glass infiltrates throughout the lungs (Figure 2A), splenomegaly, and

Figure 1. A, Color fundus photograph of the right eye revealing serous detachments and intraretinal heme along the inferotemporal arcade. B, B-scan ultrasound of the right globe at initial examination. There is marked choroidal thickening and elevation of the serous detachments. C, Late-frame fluorescein angiogram (1 day) of the right eye revealing punctate hyperfluorescence surrounding the localized serous detachments, which accumulate fluorescein dye. D, Indocyanine green angiogram of the right eye revealing punctate hyperfluorescent lesions of the choroid. E and F, Late frames of the right eye (1 week after treatment) demonstrating resolution of previously noted abnormalities.

Figure 2. A, Granular, ground-glass appearance of patient’s computed tomographic scan of the chest. B, Hematoxylin-eosin stain of lung biopsy specimen (original magnification ×40) demonstrating large lymphoid cells within the vasculature of individual alveoli. C, Immunohistochemical stain (original magnification ×40) of lung biopsy specimen showing a positive stain for the CD20 marker in brown.
diffuse edema within the subcutaneous tissues of the thorax and abdomen. A biopsy specimen of his skin lesions showed nonspecific inflammatory cell infiltration of the dermis. A bone marrow biopsy specimen revealed hypercellularity with trilineage hyperplasia consistent with a peripheral destructive process. The blood smear demonstrated several atypical lymphocytes with convoluted nuclei. Cerebrospinal fluid examination results were normal, while a lung biopsy specimen revealed large, atypical CD20-positive and CD3-negative lymphocytes filling the lumen of the capillaries in the alveolar septae (Figure 2B and C). These findings were diagnostic of malignant intravascular lymphoma of B-cell phenotype (intravascular lymphomatosis).

A regimen of hyper-CVAD (cycle 1: cyclophosphamide, vincristine, doxorubicin, and dexamethasone) chemotherapy, intrathecal methotrexate and cytarabine, and methylprednisolone was initiated 10 days after initial examination. A week later, there was a marked improvement in visual acuity (20/20 OU) and resolution of serous detachments and choroidal thickening, shown by repeat fluorescein angiography and indocyanine green angiography (Figure 1E and F). The patient continued to do well 2 months after starting therapy.

Comment. Intravascular large B-cell lymphoma is a rare form of extranodal malignant lymphoma in which the lymphoid cells are principally found within small vessels without extensive involvement of bone marrow or lymphoid tissue. Aberrant expression or deletion of certain cell adhesion molecules critical for lymphocyte trafficking and transvascular migration (CD29 and CD54) contributes to the tumor’s intravascular and disseminated pattern of distribution. The tumor expresses CD20 and other B-cell markers and does not express the T-cell marker CD3. At diagnosis, the lymphoma typically involves various organs, including the central nervous system, skin, lung, kidneys, spleen, adrenal glands. Other types of lymphoma may separately coexist with intravascular large B-cell lymphoma in the same patient. Hematologic abnormalities, like microangiopathic and autoimmune hemolytic anemia, as in our case, are often associated with this disease process. The prognosis is generally very poor because of the usually delayed diagnosis. Intravascular large B-cell lymphoma can also involve the small vessels of the choroid. The case presented herein adds the eye as another end organ involved by this rare malignancy.

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Unilateral Eyelid Swelling and Ptosis Caused by Dural Arteriovenous Fistula in an Infant

Patients with potentially life-threatening intracranial vascular abnormalities may first come to an ophthalmologist with signs or symptoms that include visual disturbances, bruis, papilledema, or periorbital congestion commonly coexisting with other neurologic and cardiovascular problems. In this report, we discuss a girl seen first at 6 months of age with the rare finding of isolated, unilateral eyelid ptosis and fullness due to a dural arteriovenous fistula.

Report of a Case. A female child was noted at birth to have right upper eyelid swelling and ptosis. Although they were believed to be related to birth trauma, there was little change in her eyelid appearance over the next few months, prompting referral to a pediatric ophthalmologist (B.J.K.) when the girl was 6 months old. Fullness of the right upper eyelid was evident (Figure 1), but no palpable mass or orbital bruit was appreciated. The remainder of her ophthalmic examination, including eyelid function, motility, and sweep visual evoked potential testing, had normal findings for both eyes. The child had otherwise enjoyed good health and achieved appropriate developmental milestones.

Magnetic resonance imaging revealed a dural arteriovenous fistula with massive engorgement and dilation of the dural venous sinuses, including the distal superior sagittal sinus, torcular herophili, and right transverse sinus (Figure 2A). Cerebral angiography identified multiple arterial feeding vessels, the most prominent of which were derived from the occipital arteries bilaterally (Figure 2B). Also noted were bilateral dilated superior ophthalmic veins due to shunting of cerebral venous blood to the external periorbital veins. There was no evidence of intracranial mass effect, infarct, hemorrhage, or other parenchymal abnormalities.

With the poor natural history of dural arteriovenous fistula in children, we decided to attempt endovascular embolization of the lesion to prevent long-standing cerebral venous hypertension and potentially irreversible brain injury. A preoperative cardiovascular evaluation, including echocardiogram, revealed only a slightly enlarged left atrium due to mildly increased cardiac output. At 7 months of age, the patient underwent initial endovascular treatment using a combined transve-