RESEARCH LETTERS

Cavernous Hemangioma of the Iris

Vascular tumors and malformations of the iris are rare. We report an iris cavernous hemangioma associated with recurrent hyphemas and elevated intraocular pressure.

Report of a Case. A 55-year-old man was referred for an iris lesion in the right eye. Eighteen years earlier he had a spontaneous hyphema that led to detection of the lesion. During the ensuing 2 decades, he had 30 spontaneous hyphemas, each associated with transient elevated intraocular pressure. One month prior to referral, a hyphema with a pressure of 55 mm Hg was treated successfully with topical medication. Visual acuities were 20/40 OD and 20/20 OS. Intraocular pressures were 12 mm Hg OD and 16 mm Hg OS. Slitlamp biomicroscopy showed a few scattered erythrocytes in the aqueous of the right eye but no hyphema. Mild pigment dispersion was present on the iris surface and transillumination revealed a circular pattern of radial slitlike defects in both irides.

The main finding was a lobulated, reddish-blue temporal iris mass measuring 3 mm in diameter (Figure 1). Ultrasound biomicroscopy showed a multiloculated mass, and fluorescein angiography demonstrated early hypofluorescence of the lesion with mild hyperfluorescence in the superior aspects of the vascular spaces. The clinical diagnosis was iris cavernous hemangioma, but vascular iris melanoma could not be excluded. The mass was removed by sector iridectomy. Histopathologic examination revealed a benign tumor composed of large blood-filled vessels lined by thin endothelial cells, compatible with a cavernous hemangioma (Figure 2). Scattered pigmented macrophages were present outside the vascular channels. The adjacent iris stroma near the pupil appeared focally fibrotic and hyalinized. The visual acuity remained 20/40 OD after surgery.

Comment. There are several vascular malformations and tumors that occur in the retina and choroid, but such lesions in the iris are rare. Ferry reviewed sections of cases submitted to the Armed Forces Institute of Pathology prior to 1972 and found that most “hemangiomas” had been misdiagnosed histopathologically and proved to be highly vascular melanomas, juvenile xanthogranuloma, and other lesions. He suggested that iris vascular tumors were extremely rare, and he even questioned their existence. However, based on our clinical experience, there are convincing cases of iris capillary hemangioma, cavernous hemangioma, racemose hemangioma, and varix.

Iris capillary hemangioma can occur in children with congenital periocular cutaneous capillary hemangioma. It tends to regress spontaneously with regression of the concurrent cutaneous hemangioma. Iris racemose hemangioma is an anomalous arteriovenous malformation. In a recent report of 14 cases, these vascular malformations were divided into simple and complex types. Unlike the retinal racemose hemangioma, it is not associated with Wyburn-Mason syndrome. Iris varix occurs as a red-blue mass that lacks a distinct blood supply and is generally hypofluorescent with fluorescein angiography.

Iris cavernous hemangioma can have either of 2 clinical variations. One is typically very small and located near the pupillary border. It often is difficult to visualize and can cause spontaneous hyphema, particularly after cataract surgery. The other type, like the case reported here, appears as a clearly visible mass in the iris stroma. Either type can rarely be seen as part of a syndrome with similar lesions in brain, kidney, and skin.

Figure 1. Clinical appearance of iris cavernous hemangioma showing a red-blue circumscribed iris stromal mass.

Figure 2. Low-magnification photomicrograph showing cavernous venous channels with intraluminal erythrocytes (hematoxylin-eosin, original magnification ×20).
Our patient had mild bilateral pigment dispersion syndrome. We speculate that the unilateral episodes of elevated intraocular pressure were due to the recurrent hyphemas and not to the pigment dispersion. Some iris melanomas can also be highly vascular and simulate a hemangioma. However, the diagnosis of cavernous hemangioma should be suspected when the entire lesion is composed of large venous channels without a solid melanocytic component.

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Peripheral Iridoplasty Efficacy in Refractory Topiramate-Associated Bilateral Acute Angle-Closure Glaucoma

Simultaneous bilateral acute angle-closure glaucoma (BAACG) is a rare and potentially blinding disease. From 2001 through 2007, there have been about 100 reports of topiramate-associated secondary BAACG, establishing topiramate as a leading cause of this condition in patients younger than 40 years. In 83 cases of bilateral and 3 cases of unilateral topiramate-associated acute secondary angle-closure glaucoma, 7 patients sustained permanent vision loss. Topiramate (Topamax; Ortho-McNeil Neurologics, Titusville, New Jersey) was approved by the Food and Drug Administration to prevent seizures in 1996 and migraine headaches in 2004, but it is also being used off-label for depression and bipolar disorders, neuropathic pain, and weight reduction. In 2001, a warning was added to the package insert describing a rare syndrome consisting of ciliochoroidal effusion, forward displacement of the lens-iris diaphragm, marked anterior chamber shallowing, acute myopia, and secondary angle-closure glaucoma. This process is usually reversible if topiramate is discontinued and ocular hypotensive therapy is instituted. In eyes unresponsive to these measures, there are no definitive treatment recommendations to avoid high-risk fistulizing surgery. Because pupillary block is not present in topiramate-associated BAACG, peripheral iridectomy is ineffective.

We describe 4 patients (8 eyes) who were effectively treated with argon laser peripheral iridoplasty (ALPI) for topiramate-associated BAACG unresponsive to ocular hypotensive therapy and topiramate discontinuation. To our knowledge and according to a MEDLINE literature review performed in March 2008, there are no published reports on the use of ALPI for this condition.

Report of Cases. Table 1 and Table 2 contain patient demographics, rationale and duration of topiramate treatment, clinical parameters of topiramate-associated BAACG before and after ocular hypotensive therapy, peripheral iridoplasty treatment and effect, and clinical parameters of topiramate-associated BAACG after peripheral iridoplasty. Because bilateral complete angle closure developed and persisted in 4 white women despite medical therapy and topiramate discontinuation, ALPI (300- or 500-µm spot size, 0.5-second duration, 200- to 400-mW power range) was performed (by A.H.Z.) in all of the 8 eyes (Figure). Within 30 minutes of ALPI, the intraocular pressure was markedly reduced and the peripheral anterior chamber had significantly deepened in all of the eyes. In an effort to prevent a subsequent pupillary block, all of the eyes underwent a laser iridotomy later the same day. Five months after iridoplasty, the intraocular pressure was normal in all of the eyes in the absence of ocular medications; only 4 eyes had mild, residual peripheral anterior synechiae.

Comment. Argon laser peripheral iridoplasty is a relatively easy, safe, and effective treatment for refractory topiramate-associated BAACG. To our knowledge, this case series represents the first report describing the use of ALPI for this condition. The ALPI technique is a well-recognized procedure used to treat angle closure from mechanisms other than pupillary block. Although this report is limited by its small sample size and lack of a control eye, refractory topiramate-associated BAACG should be added to the list of indications for ALPI.

A unique finding in this case series was progressive central anterior chamber shallowing even after ALPI eliminated appositional angle closure. While the mechanism is unclear, we believe that the topiramate-associated